Intrathoracic meningocele

Its development and association with neurofibromatosis

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An antero-lateral meningocele is a protrusion of the spinal meninges through an intervertebral foramen. It contains an extension of the subarachnoid space, filled with cerebrospinal fluid, and this gives rise to a paravertebral cystic swelling, usually spherical in shape. Theoretically such a swelling could occur in the neck, or in the thoracic, abdominal, or pelvic cavities, but practically all the recorded cases (and we have discovered only 66 in literature) have occurred in the thoracic cavity and are there referred to as intrathoracic meningoceles.

A feature of the abnormality is that nearly two-thirds of the recorded cases have occurred in association with neurofibromatosis. It has thus been of especial interest to neurological and thoracic surgeons because of the differential diagnosis from dumb-bell neurofibroma—that is, a neurofibroma beginning on a spinal nerve root, enlarging in the vertebral canal, extending out through the intervertebral foramen and enlarging again in the paravertebral space. Indeed the original observation by Pohl (1933) concerned a 47-year-old woman with von Recklinghausen's disease who was mistakenly thought to have an intrathoracic neurofibroma. The majority of the recorded cases have come into the hands of thoracic surgeons, and the increasing practice of mass radiography of the chest may be expected to provide more cases in the future.

Because of their rarity, little is known about the natural history of intrathoracic meningoceles and, that being the case, the indications for treatment and the most effective methods of treatment are by no means clear. We have not encountered a comprehensive review of the literature.

We present below a report of four cases seen in the Department of Neurological Surgery, Radcliffe Infirmary, Oxford, during the past 18 years, together with a review of the literature, a discussion of the

various theories of aetiology, and suggestions as to treatment.

CASE REPORTS

CASE 1 A.G. (R.I. No. 96373), a 66-year-old retired barman, with lifelong evidence of neurofibromatosis, presented in January 1949 with a year's history of progressive paraparesis. On examination he had an upper thoracic scoliosis and a moderate spastic paraparesis. There was a relative sensory deficit, most marked to pain and temperature stimulation, with an upper level at the 4th thoracic dermatome. Radiographs (Fig. 1) showed the gross scoliosis, convex to the left, at the apex of which there was a rounded opacity filling the upper pole of the thoracic cavity on that side. Lateral radiograph confirmed this position and also revealed gross scalloping of the posterior surfaces of the bodies of the 2nd, 3rd, and 4th thoracic vertebrae.

Lumbar puncture produced slightly yellow fluid, which proved to contain 100 mg/100 ml. protein, at a pressure of 50 mm water. Queckenstedt's manoeuvre produced a rise of only 55 mm in 10 seconds and the return to normal took 45 seconds—that is, there was thought to be a partial manometric block. Lipiodol myelography showed an incomplete arrest at the 6th thoracic vertebra and none of the contrast medium passed out of the vertebral canal. The presumptive diagnosis was compression of the spinal cord by dumb-bell neurofibroma.

At operation on 14 January 1949, the laminae of the 2nd, 3rd, and 4th thoracic vertebrae were found to be abnormally thin. When they were excised, no tumour was visible in the vertebral canal. On opening the dura a very capacious subarachnoid space was revealed, but again there was no tumour. The three intervertebral foramina on the left, at the apex of the scoliosis, appeared dilated, especially that between the 2nd and 3rd thoracic vertebrae, which had a cross-sectional diameter of about 5 cm, and through this the meninges passed out into the chest as a cystic diverticulum. The wall of the cyst was so thin that the carbon-mottled lung surface and its movement on respiration could be seen through it. The spinal cord was tightly stretched across the apex of the scoliosis. There seemed no indication to interfere with the meningocele. Lipiodol was placed in the cavity, and its position, in the upper chest opacity, was confirmed radiographically.

Thirty-six hours post-operatively, the patient developed

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A selected set of references is appended, but a full list of 69 references may be obtained by application to: The Department of Neurological Surgery, Radcliffe Infirmary, Oxford.

FIG. 1. Case 1. Upper thoracic scoliosis with meningocele filling the left thoracic apex.

a left-sided pneumothorax which proved fatal. Necropsy provided no explanation for the pneumothorax and merely confirmed the surgical findings (Fig. 2). During the operation it had been observed that there was abnormal mobility of the vertebral column at the site of the meningocele, and this also was confirmed at necropsy. It was suggested that this abnormal mobility at the site where the cord was tethered over the kyphus may have contributed to the paraparesis.

CASE 2 E.S. (R.I. No. 147752), a 48-year-old housewife, was admitted on 25 September 1951 after a rounded opacity in the right lower chest had been discovered during a barium meal examination for dyspepsia (Fig. 3). She also complained of low backache and slight weakness and nocturnal paraesthesiae of the left leg.

On examination she was found to have 'café-au-lait' patches of neurofibromatosis. There was slight weakness of the left leg without reflex or sensory changes. Chest radiographs showed the opacity to be centred over the right sides of the 11th and 12th thoracic vertebrae, with considerable scalloping involving the posterior surfaces and the right sides of the bodies of the 10th thoracic to the 1st lumbar vertebrae. The vertebral canal was widened with thinning of the pedicles over the same region, and the head of the right 12th rib was eroded. There was slight kyphotic deformity of the spine at this level, but no significant scoliosis. Lumbar puncture revealed normal cerebrospinal fluid with normal manometry. The diagnosis of antero-lateral meningocele was

FIG. 2. Case 1. Necropsy specimen showing the meningocele seen from the thoracic cavity, with the vertebral bodies removed to show the continuity of the spinal meninges with the wall of the meningocele.

confirmed by air myelography: air injected into the lumbar subarachnoid space passed freely into the meningocele (Fig. 4).

Because of the symptoms and the slight left leg weakness, thoraco-lumbar laminectomy was performed on 22 October 1951. As in the first case, there were thinned laminae, and expansion of the vertebral canal and subarachnoid space extending over at least three segments with no evidence of either tumour or compression or distortion of cord or roots. The meningocele passed out through an intervertebral foramen dilated to 2.5 by 3 cm. The intercostal nerve and accompanying artery were seen passing through the cavity. The neck of the meningocele was dissected free from the bony margins of the intervertebral foramen and was ligated but not divided. There were no post-operative complications and the pain and paraesthesiae disappeared immediately.

Fifteen years later (1966), there has been no recurrence of symptoms. On examination there is now definite

FIG. 3. Case 2. Lateral radiograph of the chest with the meningocele outlined by arrows, and showing the posterior vertebral body scalloping.

kyphosis with little scoliosis, and, while the patient regards the power in her legs as normal, the left thigh is a little wasted and the left knee and ankle reflexes are exaggerated. Sensation is normal. Radiographs show the opacity to be essentially unchanged in size with comparable posterior scalloping but increased lateral scalloping. The rib erosion is unchanged.

CASE 3 A.G., a 24-year-old student with neurofibromatosis, was found to have a rounded opacity in the left lower thorax at routine chest radiography in 1955. The posterior surfaces of the bodies of the 8th to the 12th thoracic vertebrae were considerably scalloped and the bodies themselves were irregular in outline. There was a slight scoliosis concave towards the opacity. The left pedicles of the 10th to the 12th thoracic vertebrae were thinned and that of the 9th was absent. The interpedicular distance was increased at these levels. The heads and necks of the left 9th and 10th ribs were eroded (Fig. 5, a + b). There was a prominent posterior 'café-au-lait' patch in the left paravertebral position over the lower

FIG. 4. Case 2. Air myelography has resulted in the replacement of some of the cerebrospinal fluid in the meningocele by air. With the patient in the vertical position a fluid level results as seen. The arrows indicate the thin meningocele wall outlined by air.

six ribs. The character of the opacity and more particularly the associated bony abnormalities suggested the diagnosis of antero-lateral meningocele, but, as there were neither symptoms nor signs referable to the chest or the spinal cord, no treatment was advised. Over the succeeding 12 years he has continued to be symptom-free, but, radiologically, there has been a definite increase in the size of the meningocele: its volume was estimated as 55 ml. in 1955, and as 241 ml. in 1967.

CASE 4 T.M. (R.I. No. 2563), a 24-year-old chef, a binocular twin, had neurofibromatosis though there was no family history of such. At routine chest radiography on 13 April 1966 an opacity was seen over the heads of the left 10th and 11th ribs, which were grossly eroded (Fig. 6). There was slight kyphoscoliosis and gross posterior scalloping of the bodies of the 8th to the 11th thoracic vertebrae, with posterior collapse (reversed wedging) of the 10th thoracic vertebral body. There was expansion of this segment of the vertebral canal. He had complained for three years of slight mid-thoracic and lumbar back-ache with occasional cramp in the left leg, and had obtained some relief from a plaster of Paris support.

On examination the power and the tone in the legs were normal, although the left knee reflex was diminished and the ankle reflex was absent. There was no sensory

FIG. 5a and b. CASE 3. Penetrated radiographs (1955) showing (a) the meningocele lying over the eroded heads of the left 9th and 10th ribs; (b) the meningocele and the degree of posterior scalloping.

deficit. Again it was felt that at present no treatment was indicated and that no further investigations were necessary. However we had previous records of this patient. He was first seen in 1943 as a 10-month-old baby, when he presented with right upper lobe pneumonia, and review of the chest radiographs taken at that time now reveal thinning of the left 10th and 11th ribs and the suggestion of a lower thoracic scoliosis. Subsequent radiographs taken because of recurrent chest infection confirmed these abnormalities. At the age of 5 he presented again with a left occipital skull defect, and at this examination 'café-au-lait' patches were found on his trunk and these had appeared only over the preceding few months. Because of the known association of bony dysplasia with neurofibromatosis, further radiographs were taken and those of the vertebral column (Fig. 7) showed posterior scalloping of the 8th to 11th vertebrae and, to a lesser extent, the 2nd to 4th lumbar vertebrae inclusively, with widening of the canal and thinning of the pedicles. The rib changes were confirmed, and thinning of the 10th and 11th thoracic transverse processes with, in fact, costo-transverse disarticulation was now

also seen. As there was no neurological deficit and lumbar puncture revealed normal cerebrospinal fluid with normal manometry, no surgery was undertaken. Frequent radiographs were taken up to the age of 13 (1953) and these showed only slight increase in the scalloping. The child remained well and active.

It was not until 1966, when he was 24, that the meningocele was discovered on a routine chest radiograph taken in connection with his work as a chef. Retrospective examinations of previous chest radiographs show that it was not present in 1957, although by 1961 it was just visible; at that time it was smaller and almost obscured by the cardiac shadow. The volumetric increase, estimated radiographically, was from a few ml. in 1961 to 65 ml. in 1966. There has been a further increase in the last year to 85 ml.

REVIEW OF THE LITERATURE

Of the 70 cases now reported in the literature, 38 were women and 31 were men. We have chosen to

FIG. 6. Case 4 in 1967. Penetrated radiographs showing (a) the meningocele in the lateral projection and also showing the severe posterior vertebral scalloping with posterior collapse of the 10th thoracic vertebral body; (b) underlying left 10th and 11th ribs are shown to have eroded heads and thinned necks, and there is upward dislocation of the 10th rib.

exclude from this series three cases that have been included in some previous series. In each we feel that the evidence for the protrusion being an antero-lateral meningocele is inconclusive.

AGE OF PRESENTATION The meningocele is usually detected in middle life, the average age in the recorded cases being 43.3 years; 74% presented between the age of 30 and 60 years.

SITE In 37 cases (52%) the meningocele was on the right; in 27 cases (40%) it was on the left. In eight cases there was more than one meningocele and in five of these there was a meningocele on each side. In one case the side was not specified. The apparent right-sided preference may be artificial, as small left-sided meningoceles may be obscured in the radiograph by aortic and heart shadows. As an example of this, LaVielle and Campbell (1958) thought that they were dealing with a single meningocele until the post-operative film, as well as

showing satisfactory removal of the right-sided 10th thoracic meningocele, also showed a previously unrecognized meningocele at the same level on the left side. In our fourth case, similarly, the meningocele was recognized only when it had expanded beyond the cardiac shadow. Wilhelm (1954) suggested that there might be particular paucity of lower left meningoceles, and that this could be due to the buttressing effect of the descending aorta. We found a random distribution along the thoracic spine, with the exception that, for no obvious reason, as many as 13 (20%) occurred between the 10th and 11th thoracic vertebrae.

MODE OF PRESENTATION (Table 1) *Chance findings* Thirty-two meningoceles were discovered on routine radiological examination of the chest, nine on radiography for completely unrelated symptoms, and there was one chance necropsy finding. A total, therefore, of 42 (60%) could be considered chance findings. Many of the others presented with symp-

FIG. 7. Case 4 in 1948. On the left showing the thinning of the proximal parts of the 10th and 11th ribs. In the lateral projection posterior vertebral body scalloping is seen in the lowest thoracic vertebra and again in the mid-lumbar vertebrae.

toms only doubtfully related either to the meningocele or to the part of the spine bearing it.

Pain Though not a characteristic feature, pain was present in 16 cases (23%). All but one of those who had pain also had kyphosis, and in the exception the presence or otherwise of kyphoscoliosis was not specified. In nine the pain was referred to some—though not necessarily the appropriate—part of the vertebral column, and in six it was of a radiating, intercostal type, conceivably referable to the meningocele. In no case was there mention of pleuritic pain.

Neurological abnormalities These were on the whole slight and uncommon. Only six patients (9%) had significant paraparesis, but another seven had slight abnormalities such as hyper-reflexia, slight weakness, vague sensory impairment, and the like. Kyphoscoliosis was specified as being present in all but one of those who had any kind of neurological deficit in the legs.

TABLE I
MODE OF PRESENTATION

Presentation	(no.)	(%)
Chance finding	42	60
Pain	16	23
Dyspnoea	8	11
Cough	7	10
Paraparesis	6	9
Direct presentation:	2	3
1 rupture		
1 bleed		
Dyspepsia	1	

Pulmonary symptoms Cough was present in seven (10%) and dyspnoea in eight (11%). Only in those cases in which the meningocele assumed massive proportions was the respiratory embarrassment severe, as in the case reported by Waterfall (1966).

Presentation directly related to the meningocele This occurred twice. Portigliatti-Barbos (1953) had a case in which the meningocele ruptured during a bout of coughing, when it was penetrated by a previously fractured rib. The patient died from medullary coning due to leakage of cerebrospinal fluid into the pleural cavity. Another patient presented with spontaneous haemothorax originating from the very vascular wall of the meningocele, and survived its excision. This vascularity proved fatal at one operation and was a serious problem at another. Dyspepsia and dysphagia were not previously recorded but may have been a feature of our second case.

RADIOLOGICAL FINDINGS (Table II) **Kyphoscoliosis** Some degree of kyphoscoliosis varying from slight to very gross was present in 47 cases (67%). In 15 cases (21%) there was said to be none. The kyphoscoliosis usually had the meningocele at its apex on the convex side, but in our third case there was a slight scoliosis concave toward the meningocele. When there were bilateral meningoceles, there was no obvious reason why the scoliosis should have been to one side rather than to the other.

The incidence of kyphoscoliosis in people with

neurofibromatosis has been variously estimated as being between 9% according to Hunt and Pugh (1961) and 38% by Laws and Pallis (1963). This association has been further emphasized in this series in that, if a patient had a meningocele, kyphoscoliosis appeared to be present when there were other recognizable stigmata of neurofibromatosis. Thus of 46 patients with meningocele and obvious neurofibromatosis, 39 (85%) had kyphoscoliosis, while of the 15 without any evidence of neurofibromatosis only five (33%) had kyphoscoliosis.

Scalloping This was present in 48 cases (69%) and again was usually associated with kyphoscoliosis. The number of vertebral bodies affected varied from two to six, and in our fourth patient the spine seemed to be involved at two separate levels, lower thoracic and lower lumbar. Reversed wedging—that is, collapse of the posterior part of the vertebral body—has been noted by Laws and Pallis (1963) and in our fourth patient was shown tomographically to be related to the degree of posterior scalloping.

Enlargement of the intervertebral foramen This was a common finding but was specifically said to be absent in five cases. It was also common for several adjacent foramina to be enlarged, even though the meningocele emerged through a single foramen.

Rib changes These were recorded in 36 cases (51%) and were probably not present in nine cases. They varied from splaying with increase in the intercostal space, to gross thinning of the adjacent surfaces of the heads and necks at the site of the meningocele. Costo-transverse dislocation occurred with the most marked thinning, and Waterfall (1966) described progression of rib changes accompanying the expansion of the meningocele. The possible significance of rib changes before the development of the meningocele will be discussed later. These proximal rib changes are different from those commonly associated with neurofibromatosis—that is, the ‘twisted ribbon’ type of deformity which usually occurs in the more distal parts of the shaft of the rib.

Other bony abnormalities These are rare, although synostosis of adjacent vertebral bodies was

seen in the vicinity of the first meningocele that was described by Pohl (1933).

Radiological evidence of enlargement In 12 cases increase in the size of the meningocele was known to have occurred over periods of from two to 10 years. In two cases, the meningocele ultimately attained a massive size, occupying two-thirds of the hemithorax. No enlargement of the meningocele occurred in six instances followed for periods of from one to nine years.

Abnormalities in adjacent organs Pulmonary cysts have been seen at the same level as the meningocele and at laminectomy an arachnoid cyst was found just above another meningocele.

THE INCIDENCE OF NEUROFIBROMATOSIS In 46 cases (64%), there was definite evidence of neurofibromatosis; six were said to have none, but one of these had a positive family history. In an attempt to explain the absence of signs of neurofibromatosis in some cases, Wilhelm (1954) suggested that their appearance might be delayed until after the meningocele was discovered. Voisin, Macquet, Wattel, Mahieu, Leduc, and Jacob (1964) likewise emphasize the evolutionary progression of overt evidence of neurofibromatosis. Our fourth patient had bony abnormalities before cutaneous manifestations appeared, and Teng and Eastman’s (1958) patient had but one small subcutaneous occipital neurofibroma to suggest the presence of this disease. Cross, Reavis, and Saunders (1949) considered that the meningocele might be a *forme fruste* of neurofibromatosis, and the only apparent manifestation. The siting of the skin stigmata did occasionally seem to have some relevance, as with the large overlying café-au-lait patch in our fourth patient, and adjacent intercostal neurofibromata have also been described.

Other conditions known to be associated with neurofibromatosis also occurred in this series and included two phaeochromocytomata, two pontine gliomata, and a presumed optic nerve glioma. In one patient sarcomatous change occurred in a peripheral neurofibroma.

The Differential Diagnosis This always included

TABLE II
RADIOLOGICAL DATA

	Kyphoscoliosis		Vertebral scalloping		Foraminal enlargement		Rib changes		Evidence of enlargement	
	(no.)	(%)	(no.)	(%)	(no.)	(%)	(no.)	(%)	(no.)	(%)
Present	47	67	48	69	50	71	36	51	12	17
Definitely absent	15	21	7	10	5	7	9	13	6	9

neurogenic tumour—for example, neurofibroma, neuroblastoma, ganglioneuroma, etc—and in one case ruptured aortic aneurysm. Thirty-six of the meningoceles were operated upon with a presumptive diagnosis of intrathoracic neurogenic tumour—usually because of the risk of malignancy, which is quoted by Kent, Blades, Valle, and Graham (1944) as being as high as 40% in this type of tumour. However some authors, including Hilton and McCarthy (1959), have suggested that in the presence of neurofibromatosis, a paravertebral shadow is more likely to be a meningocele than a neurogenic tumour. Supporting this relative dissociation between generalized neurofibromatosis and intrathoracic neurogenic tumour, Bikfalvi (1964) found evidence of the general disease in only one of the 28 cases of intrathoracic neurogenic tumour upon whom he operated. However Wilhelm (1954) questions the practical value of such deductions when intrathoracic neurofibroma is so much more common than intrathoracic meningocele.

Laws and Pallis (1963) support the view that the associated adjacent bony abnormalities—that is, in the vertebrae and ribs—are themselves sufficiently characteristic for the correct diagnosis to be made, while others argue the need for special investigations. With Laws and Pallis we think that by virtue of these characteristic vertebral abnormalities, and particularly their extending over several segments, it should be possible to make the correct diagnosis without resorting to myelography, as long as there is no evidence of paraparesis. In all cases with paraparesis myelography is necessary to assess possible cord compression due to a dumb-bell tumour or kyphoscoliosis. If the chest opacity is felt to be an intrathoracic neurofibroma, and in spite of there being no evidence of cord compression, there is still concern at the possibility of an intravertebral extension, the relatively simple procedure of lumbar puncture might be of some help: a normal Queckenstedt response and a normal amount of protein in the cerebrospinal fluid might provide considerable reassurance. However if there were evidence of a partial or complete block, myelography is needed to differentiate between the possible causes.

Regarding the cerebrospinal fluid itself, we have not been able to explain the findings in our first patient. There was evidence of obstruction by the impaired response to jugular venous compression, but, whereas the protein content of the fluid was only 100 mg/100 ml., the fluid was yellow, which one usually expects only in association with much higher concentrations of protein. We found no similar reports in the literature.

Diagnosis The correct diagnosis was made pre-

operatively in 29 cases: in 18 by Myodil or Lipiodol myelography as first used by Schüller and Uiberall (1938). Myelography failed to reveal the meningocele on two occasions, merely indicating some degree of block at the appropriate level, probably due to kyphoscoliosis. Diagnosis was achieved by direct puncture and aspiration twice, and confirmed by direct instillation of Lipiodol on two other occasions. Indigo carmine was instilled directly into another meningocele and then stained cerebrospinal fluid was withdrawn at lumbar puncture. The diagnosis was made in one case by comparing the cerebrospinal fluid pressures of simultaneous direct puncture and lumbar puncture. The safest diagnostic procedure would seem to be air-myelography, which was first performed in this type of abnormality by Cross in 1949 and has been used successfully eight times since.

TREATMENT *Corrective surgery* This was performed on 42 meningoceles, with excision via thoracotomy on 33, and by laminectomy on two. A simple ligation after laminectomy was made on our second case. Exploration without any attempt at surgical correction was made twice and was combined with aspiration of the exposed meningocele once. Neurotomy for intractable pain was necessary in one case.

Observation by follow-up This procedure, without operation, was decided upon for 20 cases.

RESULTS OF OPERATION *Improvement* It has been difficult to evaluate improvement after operation because the symptoms have often been vague and sometimes difficult to relate purely to the meningocele. When pain was a feature, relief was obtained in four cases, though it was unaffected in three others. Dyspnoea and headache were each improved in two cases. Our second patient's dyspepsia was cured and weakness of her left leg subjectively and objectively improved, though when seen 15 years after operation there were still abnormal neurological signs to be found in the limb. There has been no report of improvement of paraparesis.

Operative mortality There were eight (19%) deaths directly related to the operation, though this figure is somewhat artificially weighted by the three early failures which occurred before the use of antibiotics. Three died from leakage of cerebrospinal fluid and subsequent infection, two from respiratory failure, one due to pneumonitis and the other, our first case, due to unexplained pneumothorax. Wilhelms' (1954) patient died from circulatory failure due to a coincidental pheochromocytoma, and Byron, Alling, and Samson's (1949) because of operative haemorrhage. The reason

for one death is not stated, and there was another death arising from the consequences of a paraplegia that had appeared only after two operations on the meningocele.

Operative morbidity Non-fatal leakage of cerebrospinal fluid occurred after four operations, and in two there was evidence of meningitis. Operation has been followed by permanent paraparesis, temporary aggravation of existing paraparesis, and Horner's syndrome.

Recurrence Radiological evidence of recurrence was seen only once by Bogedain, Carpathios, and Lawland (1963) and that was four years after operation. In this case the closure of the defect had been difficult, and an immediate cerebrospinal fluid fistula had necessitated re-operation and fascial repair. At necropsy, five years after the original operation, the graft was found to be thinned but intact.

HISTOLOGY In 16 cases histology of the meningocele wall showed it to be thin (measuring only 1 to 2 mm), smooth lined, and looking very similar to the spinal meninges with which it became continuous at the intervertebral foramen. Microscopically dura mater was seen in 13 cases, while both dural and arachnoidal meninges were recognizable in three. Hutchin and Mark (1964) found well-defined lining layers of cells in each of his three cases. In the first the lining cells were cuboidal, while in the second and third cases the cells were flattened and squamous. Bikfalvi (1964) also found a well-defined lining layer of cells that were either cuboidal or cylindrical. Nerve elements have been found in the wall, being isolated nerve fibres in six cases with additional ganglion cells in two instances. Portigliatti-Barbos (1953) found some smooth muscle fibres incorporated in the wall of the meningocele that he examined. The meningocele removed by Nanson (1957) appears to have been a little different from the others examined in that, although the upper part was only 2 mm thick and smooth lined, the lower wall was 2.6 cm thick and lined by gelatinous tissue. Microscopically this lower part seemed to be the picture of a degenerating neurofibroma. The histology of another case is described only as being that of a 'cystic lesion'.

DISCUSSION

One interesting aspect of this unusual condition is its uncertain aetiology. In discussing the aetiological possibilities it would seem necessary first to consider the frequent association with neurofibromatosis.

Neurofibromatosis (von Recklinghausen's disease) is a hereditary condition transmitted by the dominant gene, and the well-known visible

stigmata, if not already present at birth, develop or evolve in a progressive manner throughout life. With the obvious defects of skin and nerves it was originally considered to be an example of neuroectodermal dysplasia, but since it has become clear that all organs can be affected, it is now thought to be an abnormality sited at the meso-ectodermal junction. For our purpose we can confine our interest to the skeletal abnormalities that Hunt and Pugh (1961) found to occur in 50% of those with neurofibromatosis. More specifically, considering vertebral abnormalities alone, the most commonly recognized is scoliosis, which is variously quoted as occurring in from 9 to 38%. Laws and Pallis (1963), in an intensive radiological survey of the vertebral abnormalities in 18 patients with neurofibromatosis, found that 38% had scoliosis, 28% had posterior scalloping, 22% had enlargement of the intervertebral foramen, and 18% erosion of pedicles. In the region of the spine bearing such abnormalities of the vertebrae, the subarachnoid space was found to be expanded in each of the three cases that had myelography. The expansion occurred not only into the posterior excavations of the vertebral bodies, but also as saccular pouches passing out into the intervertebral foramina. They also described one intrathoracic meningocele with

FIG. 8. Myodil outlining the saccular antero-lateral expansions of the cervical subarachnoid space in a woman with neurofibromatosis.

proximal rib erosion. Such abnormalities, extending over several segments, have been found at all levels along the vertebral column and have been seen in association with antero-lateral meningocele in the cervical (Fig. 8) and in the lumbar region.

Even such a brief account of the vertebral abnormalities already recognized as occurring in neurofibromatosis must emphasize the close relationship between these and the abnormalities which review of the literature shows so often to be associated with intrathoracic meningocele. The appreciation of these similarities should make consideration of the various theories of aetiology a little clearer.

AETIOLOGY *Nerve root sleeve prolongation* Sengpiel, Ruzicka, and Lodmell (1948) suggested that the sleeve of meninges that normally accompanies the nerve root and fuses with the intercostal nerve perineurium in the intervertebral foramen might be abnormally prolonged, ending outside the foramen and thereby giving rise to a form of congenital herniation of the subarachnoid space. The pressure difference between the cerebrospinal fluid inside the 'hernia' and the intrapleural pressure outside, especially during manoeuvres such as coughing and sneezing, would then tend to cause progressive expansion of this meningocele. There is some support for this hypothesis, especially in the fact that nerve elements have frequently been found in the wall of the meningocele. The finding, at operation on our second case, of the intercostal nerve and artery passing through the meningocele must also give some support to this argument.

Cystic degeneration in a neurofibroma As previously mentioned, Nanson (1957) found histological evidence of a degenerating neurofibroma in the lower pole of one excised meningocele and suggested that such cystic degeneration occurring in an intrathoracic neurofibroma might be the primary aetiological process and that communication with the subarachnoid space occurred only as a secondary feature. Cystic change can occur in intrathoracic neurogenic tumours. Neither of these theories, however, explains the associated bony changes in adjacent vertebrae.

Trauma Cross *et al.* (1949) described an injury to the back immediately before the development of the presenting symptom of pain in their patient, while Stüber (1949) in his report of a meningocele found at necropsy noted a history of back trauma that had occurred seven years previously. Some authors consider that trauma could be important, especially when there was no evidence of generalized neurofibromatosis. In general, however, there is little to implicate trauma in the development of antero-lateral meningoceles.

Dural dysplasia Several authors have suggested that the meningocele develops at a site of dural dysplasia. If the dura were to be poorly developed, the pulsatile pressure of the cerebrospinal fluid and its erosive effect would be directly transmitted to the adjacent skeleton. Beginning with the vertebral bodies, the pedicles, and the laminae, and eventually also affecting the ribs, the picture of multiple bony erosions would ultimately be arrived at. This is a logical explanation, but at present without factual support. It is perhaps worth noting that pulsation of the meningocele at operation has never been described.

Bone dysplasia If the vertebrae themselves were dysplastic, either congenitally thin or in some other way more vulnerable, then herniation of the unsupported, although normal, meninges would likewise occur, again as a result of the differential pressure across the meninges. Against this hypothesis is the evidence that there has been only one recurrence after excision, in spite of the fact that at no time has any attempt been made to repair the bony defect. Again, the suggestion that the abnormality of bone alone is responsible for the development of the meningocele must be purely hypothetical.

Regional dysplasia or dystrophy Being less specific as to which are the primary and which the secondary effects, some authors have suggested regional dysplasia or dystrophy as being responsible for the development of these meningoceles. Affecting many different tissues and in this way relating the widespread changes in meninges, vertebrae, and ribs that are found in neurofibromatosis and also in the vicinity of an antero-lateral meningocele, it must be an attractive hypothesis.

Moore (1941) considered the regional changes in neurofibromatosis to be dystrophic, and due to faulty autonomic innervation. Hagelstam (1946) found histological evidence of an endarteritic phenomenon in the affected tissues that he studied, and considered that it supported the theory of 'neurotic atrophy'.

Congenital derangement Intrathoracic meningocele is probably rarely truly congenital—that is, present at birth. In support of this statement many of the cases, including our fourth, have had the evidence of previously normal chest radiographs. However the extent of the adjacent congenital derangement of vertebrae and ribs is such that the secondary development of a meningeal herniation should not be surprising. The manner in which such abnormalities of vertebral body, arch, and ribs, are linked can be explained embryologically. The mesenchymal sclerotome migrates dorsally from its condensation position around the notochord to

form the neural arch and laterally to form the transverse processes and ribs. Initially in continuity—and presumably therefore similarly vulnerable to dysplastic influences—they only later separate and by means of specific ossification centres develop independently. Our fourth case provides a record of congenital derangement preceding, and possibly contributing to, the development of the meningocele. It does not, however, exclude the possibility of there being both a congenital derangement and a regional dysplasia or dystrophy. The dysplasia or dystrophy could be considered secondary in that its evolution might be due to some autonomic or neurotrophic derangement. The factor or factors responsible for the greater frequency and size of antero-lateral meningoceles in the thoracic region is also unexplained. It is probably necessary to invoke as an explanation more than just the facility for chest radiography in modern medicine. The simplest explanation lies in the difference in pressure between the spinal subarachnoid space and the pleural cavity, which at rest would amount to around 10 cm water and which at times of inspiration or cough might be much greater. Perhaps supporting this, there was no further expansion in the size of the meningocele in our second case after ligation of its neck. Against this hypothesis is the fact that after excision there has been only one recurrent meningocele, whereas the pressure gradient should have remained unchanged by the operation.

CONCLUSION

Intrathoracic meningoceles, *per se*, rarely cause any symptoms. The one symptom that seems unequivocally related to the meningocele, but then only when it is a very large one, is dyspnoea. Pain is almost certainly related to the associated kyphoscoliosis, in particular to the degree of angulation, and LaVielle and Campbell (1958) were able to relieve it by fusion after it had been uninfluenced by the prior removal of the meningocele. It is difficult to imagine the meningocele influencing the spinal cord and causing paraparesis, and more probably again it is the kyphoscoliosis that is to blame. However Bueno and Osácar (1961) have described displacement of the cord towards the meningocele until incision of the arachnoid, after which it returned to a more normal position; but this patient had no paraparesis. Chandler and Herzberger (1963) at laminectomy observed a previously non-pulsatile cord to pulsate after excision of a right-sided meningocele at the level of the fifth thoracic vertebra. They felt that as a result of this procedure there had been some improvement in the power of the right arm, which together with both legs had been weak before the

operation. The paraparesis remained unchanged. Our first case had a paraparesis that was attributed to his kyphoscoliosis, possibly due to excessive mobility of the spine at this point.

There is little information as to the influence of the meningocele or of its removal on the progression of the adjacent bony changes. Our fourth case has shown some added rib thinning since the development of his meningocele, while Waterfall's (1966) case showed progressive rib splaying with increase in the size of the meningocele. Our second case has some added kyphosis and increased lateral vertebral body scalloping in spite of ligation of the meningocele neck.

TREATMENT As intrathoracic meningoceles are often symptomless, and as they do not invariably increase in size, and as the risks of operation are by no means negligible, the question of treatment is difficult. If the meningocele is a chance radiological finding and is causing no symptoms, the right course would seem to be continued observation to determine whether it is enlarging, and the temptation to remove it simply because it is there may have to be resisted. If there is evidence of progressive enlargement—and this has occurred in two-thirds of the 18 cases in which there have been serial observations—an initial attempt should be made to ligate the neck of the sac via laminectomy. This may not always be possible, but, depending on the anatomical arrangements, it may be feasible to occlude the neck of the sac in other ways—for example, by plicating sutures.

If it seems impossible to deal with the lesion via laminectomy, and more specifically if the meningocele is massive and producing respiratory symptoms, it should be approached by thoracotomy. If it can be excised, this should be done, but it is essential that a watertight closure of the neck be obtained. If the thickness of the wall of the sac, or the presence of adhesions to surrounding structures appears to prohibit a satisfactory closure, the attempt should be abandoned. The same principles apply if the meningocele is discovered accidentally at thoracotomy—for example, in an exploration for a supposed neoplasm.

SUMMARY

The four cases here presented make a total of 70 recorded cases of intrathoracic meningocele, and these have been reviewed. Discovery has usually been as a chance finding, but dyspnoea, due to a large meningocele, has been reported. There is evidence both for and against progressive increase in size.

The possible aetiology has been discussed, and

we think there is evidence for a congenitally based, regional bony derangement, affecting vertebrae and ribs, being present before and possibly contributing to the later development of a meningocele at this site. Other factors, such as abnormal peripheral prolongation of meningeal sheaths along the intercostal nerves, and the pressure difference between the spinal subarachnoid space and the pleural cavity, may also play a part.

The frequent (64%) association with neurofibromatosis (von Recklinghausen's disease) may be related to the frequency of skeletal, and especially vertebral, abnormalities in this condition. However antero-lateral meningocele may occur without external evidence of neurofibromatosis, and the vertebral changes described may therefore be the only evidence of the disease.

As long as there is no clinical evidence of spinal cord compression, it should be possible to reach the correct diagnosis without resorting to myelography. On diagnosis, follow-up observation should be undertaken, and operation carried out only for respiratory distress due to a large meningocele, or if there is evidence of rapid progression in size.

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ADDENDUM

Since compiling the material for this publication, one of us (J.B.M.) has been fortunate in being able to observe a further case of intrathoracic meningocele which illustrates the risks attendant upon operation for this condition.

A 49-year-old man with neurofibromatosis had his right apical opacity explored by a thoracic surgeon. It proved to be a meningocele and he developed a post-operative cerebrospinal fluid fistula and was

transferred for neurosurgical opinion. Happily it resolved without the need for further surgery.

REFERENCES

- Bikfalvi, A. (1964). Zur Frage der intrathorakalen lateralen Meningozele. *Brunn's Beitr. klin. Chir.*, **208**, 292-304.
- Bogedain, W., Carpathios, S. T., and Lawland, F. (1963). Intrathoracic meningocele. *Amer. Rev. resp. Dis.*, **87**, 757-761.
- Buono, M. S. del, and Osácar, E. M. (1961). Intrathoracic meningocele association with cutaneous neurofibromatosis. *Acta neurochir. (Wien)*, **9**, 561-581.
- Byron, F. X., Alling, E. E., and Samson, P. C. (1949). Intrathoracic meningocele. *J. thorac. Surg.*, **18**, 294-303.
- Chandler, A., and Herzberger, E. E. (1963). Lateral intrathoracic meningocele. Case report with preoperative diagnosis. *Amer. J. Roentgenol.*, **90**, 1216-1219.
- Cross, G. O., Reavis, J. R., and Saunders, W. W. (1949). Lateral intrathoracic meningocele. *J. Neurosurg.*, **6**, 423-432.
- Hagelstam, L. (1946). On the deformities of the spine in multiple neurofibromatosis (von Recklinghausen). *Acta chir. scand.*, **93**, 169-193.
- Hilton, H. D., and McCarthy, H. H. (1959). Intrathoracic meningocele. *J. thorac. Surg.*, **37**, 261-268.
- Hunt, J. C., and Pugh, D. G. (1961). Skeletal changes in neurofibromatosis. *Radiology*, **76**, 1-20.
- Hutchin, P., and Mark, J. B. D. (1964). Intrathoracic meningocele not associated with neurofibromatosis. *J. thorac. cardiovasc. Surg.*, **48**, 29-39.
- Kent, E. M., Blades, B., Valle, A. R., and Graham, E. A. (1944). Intrathoracic neurogenic tumors. *J. thorac. Surg.*, **13**, 116-161.
- LaVielle, C. J., and Campbell, D. A. (1958). Neurofibromatosis and intrathoracic meningocele. *Radiology*, **70**, 62-65.
- Laws, J. W., and Pallis, C. (1963). Spinal deformities in neurofibromatosis. *J. Bone Jt Surg.*, **45B**, 674-682.
- Moore, B. H. (1941). Some orthopaedic relationships of neurofibromatosis. *J. Bone Jt Surg.*, **23**, 109-140.
- Nanson, E. M. (1957). Thoracic meningocele associated with neurofibromatosis. *J. thorac. Surg.*, **33**, 650-652.
- Pohl, R. (1933). Meningokele im Brustraum unter dem Bilde eines intrathorakalen Rund schattens. *Röntgenpraxis*, **5**, 747-749.
- Portigliatti-Barbos, M. (1953). Neurofibromatosis e meningocele toracico. *Friuli méd.*, **8**, 477-484.
- Schüller, A., and Uiberall, H. (1938). A case of neurofibromatosis. Recklinghausen combined with lateral spinal meningocele. *Confin. neurol. (Basel)*, **1**, 312-317.
- Sengpiel, G. W., Ruzicka, F., and Lodmell, E. A. (1948). Lateral intrathoracic meningocele. *Radiology*, **50**, 515-520.
- Stüber (1949). Doppelseitige seitliche Meningozele thoracalis. *Zbl. allg. Path. path. Anat.*, **85**, 275.
- Teng, P., and Eastman, P. (1958). Intrathoracic meningocele. *Neurology (Mineapp.)*, **8**, 153-156.
- Voisin, C., Macquet, V., Wattel, F., Mahieu, P., Leduc, M., and Jacob, M. (1964). Les manifestations thoraciques de la neurofibromatose de Recklinghausen. *Lille méd.*, **9**, Suppl. 129-39.
- Waterfall, M. C. (1966). Intrathoracic meningocele and neurofibromatosis. *Proc. roy. Soc. Med.*, **59**, 564-565.
- Wilhelm, E. (1954). Meningozele des Brusttraumes. *Thoraxchirurgie*, **2**, 147-155.