Congenital spinal arachnoid cysts

Report of two cases and review of the literature

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The purpose of the present communication is to describe two patients, one with a congenital extradural and one with a congenital intradural arachnoid cyst of the spine and to review the relevant literature. Congenital spinal arachnoid cyst is a very rare cause of spinal cord compression. It is one of the most favourable spinal lesions for surgical removal and recovery of neurological function. The term congenital is used here in distinction from the acquired variety. Acquired extradural arachnoid cysts may develop after operation when a small tear has been made in the spinal dura, after difficult lumbar puncture and after spinal injury. Swanson and Fincher in 1947 reported four cases of acquired extradural arachnoid cysts after 1,700 exploratory laminectomies—an incidence of 0.068%. Three cases were in the lumbar region and had followed a dural tear during disc removal. The fourth cyst was related to a non-penetrating injury to the lumbar spine. Acquired intradural arachnoid cysts are also associated with spinal adhesive arachnoiditis, although cyst formation is not constantly present in this condition.

The congenital intradural cysts are a separate entity and have been described in the literature under a variety of names—for example, spinal arachnoid diverticulae, leptomeningeal cysts, localized adhesive spinal arachnoiditis, and arachnitis adhaesa circumscripta. The appearances of adhesive arachnoiditis are absent on myelography in such cases and, although usually single, the cysts may be multiple.

CASE 1

INTRADURAL CYST H.R., a 22-year-old driller, was referred from another hospital by an orthopaedic surgeon on 7 December 1966 with the following history:

In June 1963 he suddenly developed pain in the lower back and slight weakness of his left leg. The weakness improved and the pain subsided after a few weeks of bed rest but it became worse again in November 1964. He

was diagnosed as suffering from a prolapsed lumbar intervertebral disc. In May 1965 he had a recurrence of low back pain and was dragging his left leg. He was treated with traction, heat, and a corset and improved. When reviewed in July 1965 he again complained of backache and left leg weakness. There was thought to be an element of 'functional overlay'. On 30 September 1966 he returned complaining of severe low back pain and weakness of the left leg and was transferred to the Regional Neurological Centre, where straight leg raising was found to be 70° on the left and 80° on the right. There was 1 in. of wasting of the left calf and both plantar responses were extensor but there were no sensory or other motor signs. The patient stated that when the pain in his back was severe the weakness of the leg was marked and as the pain subsided the weakness improved. The intermittency of symptoms and the relation of leg weakness to pain was striking. There was no relation of pain to coughing, sneezing, or stooping, although on direct questioning he stated that walking for some distance had brought on the pain and leg weakness on two occasions. There was no significant past history.

The spinal fluid dynamics were normal and the protein was 56 mg/100 ml.

Radiology Radiographs of the dorso-lumbar spine showed no significant abnormality. Lumbar route myelography was performed on 9 December. The Myodil flowed freely to the cervical region without any significant abnormality but on flowing back from the cervical region the contrast medium entered a well-defined oblong cavity at the level of D6 which was pear-shaped and lying posterior to the spinal cord. The Myodil was partially trapped within this cavity in both feet-down and head-down positions and a diagnosis of arachnoid cyst was made (Fig. 1).

Operation On the same day laminectomy of the 6th, 7th, and 8th thoracic vertebrae was performed. The dura was seen to be expanded and was opened widely, revealing a large intradural arachnoid cyst which was connected to the subarachnoid space by a small tubular structure to the left of the midline at the level of D6 (Fig. 2). This was divided between silver clips and the cyst (5 × 2.5 × 2 cm) was removed intact. Histological examination of the cyst showed it to have a fibrous tissue wall with few cells but with one area of compact cells with a whorled arrangement suggesting an arachnoidal origin. There was no evidence of cellular infiltration.

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FIG. 1. (H.R.) Myelogram showing the upper and lower limits of the cyst in head down and feet down position. The arrow points to the stalk of the cyst.

Post-operatively he had retention of urine requiring catheterization for a few days but otherwise his recovery was uneventful. On discharge his neurological state was normal with bilateral flexor plantar responses.

CASE 2

EXTRADURAL CYST J.M., a 55-year-old fire brigade instructor, was admitted on 26 June 1968 with the following history:

He was well until three months before admission when, after a golfing week-end, he complained of lower back pain for two weeks which was followed by a feeling of pressure and stiffness in the right leg, without any root pain. A few days later he developed a constricting feeling behind the right knee and to a lesser extent behind the left. There were occasional attacks of tingling and numbness of the toes. An orthopaedic surgeon treated him with traction for a presumed disc lesion but he progressively developed a spastic gait without urinary or bowel disturbance. On examination he had a spastic paraparesis, the right leg being affected more than the left. Both plantar responses were extensor. There was hypalgesia of the right L4 and 5 dermatomes but no convincing sensory level. He had no significant past history.

The spinal fluid dynamics were normal and the protein was reported as 28 and 40 mg/100 ml. in two different specimens.

Radiology Plain radiographs of the thoracic spine showed a well-marked widening of the interpedicular

distances from D6 to D8, most marked at the level of D7 (Fig. 3). Lumbar route myelography was performed on 27 June 1968. The Myodil flowed freely through the lumbar and thoracic region but, at the level of D8, the Myodil entered a cyst lying posteriorly within the spinal canal. The upward flow of Myodil was completely obstructed at the level of D7 and most of the Myodil entered the cyst and could not be made to re-enter the true theca. By tilting the patient the upper and lower limits of the rounded cyst were demonstrated to extend from the upper border of the body of D6 to the lower border of the body of D8 vertebra. The cyst was sausage shaped, lying posterior to the spinal cord and pressing forwards (Figs. 4 and 5). A diagnosis of arachnoid cyst was made.

Operation On 28 August 1968 laminectomy of 5th, 6th, 7th, and 8th thoracic vertebrae was carried out. There was extradural fat only at the upper and lower extremities of the exposure and pulsation of the dura only at the upper end. The dura was seen at the upper and lower limits of the exposure, the remaining dura being concealed by a posteriorly placed, thin-walled cystic swelling about 3 in. in length. This was easily elevated from the dura and at the junction of its middle and lower thirds it was connected by a 'stalk' passing through the dura to the left of the midline near the sleeve for the emerging nerve root. The connecting 'stalk' was divided between two clips and this large extradural arachnoid cyst was removed (Fig. 6). The dura, which was not opened, was considerably compressed from behind and

FIG. 2.

FIG. 3.

FIG. 4.

FIG. 2. (H.R.) Showing the two silver clips at the stalk of the cyst before operative removal.

FIG. 3. (J.M.) Plain radiographs showing widening of the interpedicular distances at D6, 7 and 8 levels.

FIG. 4. (J.M.) Myelogram in head down position showing the upper limit of the cyst. The arrow points to the stalk of the cyst.

FIG. 5. (J.M.) Lateral myelogram showing the upper and lower limits of the cyst which is compressing the spinal cord anteriorly. The arrow points to the neck of the cyst.

the volume of the spinal canal was increased as had been shown by the plain films.

Histological examination of the cyst showed a membrane consistent with arachnoid. There was no evidence of cellular infiltration. He made a satisfactory recovery apart from retention of urine for a few days for which he needed catheterization. His walking was much improved before discharge. When seen five months after the operation, he had returned to his original work, and was walking well but his right knee was still a little stiff. Tendon reflexes in the lower limb were still increased but the plantar responses were flexor.

REVIEW OF LITERATURE

EXTRADURAL CYSTS In 1934 Elsberg, Dyke, and Brewer reported four cases of spinal extradural cyst with compression of the spinal cord. One of the patients described by them, however, had previously been reported by Collins and Marks in 1915. In 1937, Cloward and Bucy reported nine cases of extradural cyst from the literature and described

one of their own. Since then many other reports have appeared (Good, Adson, and Abbott, 1944; Cuneo, 1955). Wise and Foster (1955) collected 33 cases and reported a personal case. Dastur (1963) described three cases of extradural arachnoid cyst with an account of the special radiological appearances on plain radiography of the thoracic spine.

Earlier reports claimed that these cysts were most common in adolescence. Seven out of 10 cases reported by Cloward and Bucy (1937) were under 15. However, Wise and Foster (1955) reviewed 33 cases of extradural cyst with an average age of 22.6 years, with twice as many male as female cases. In the majority of cases the cyst is situated in the thoracic region, sometimes thoraco-lumbar, rarely lumbar, and very rarely cervical. Meredith (1940) reported one case in which the cyst occurred in the cervical region.

Kyphoscoliosis is a very common associated abnormality in adolescent patients. In 1937 Cloward

FIG. 6. (J.M.) At operation the forceps are lifting the lower end of the cyst to show the two silver clips at its stalk before removal.

and Bucy described 10 cases and demonstrated that with one exception all reported cases of spinal extradural cysts which gave rise to symptoms during adolescence were associated with more or less marked radiological stigmata of kyphosis dorsalis juvenalis (Scheuermann's disease). If the symptoms of cord compression begin after the bony development of the vertebral column is complete, the above changes do not appear. These authors also advanced the hypothesis that extradural cysts, by interfering with the venous drainage of the vertebral bodies, were responsible for the kyphosis.

Aetiology These cysts are considered to be of congenital origin. Elsberg *et al.* (1934) proposed two theories of origin: (1) a congenital diverticulum of the dura mater, and (2) herniation of the arachnoid through a congenital defect in the dura. The fact that the attachment to the dura was always found at or near the dural opening for a nerve root favours these hypotheses. In some cases the communication with the subarachnoid space remains patent, in others it shuts off secondarily but the cyst continues to enlarge, especially if the cyst wall has an endo-

thelial lining. Hyndman and Gerber (1946) postulated that the cysts arise from cell rests. This theory seems unlikely, since in about 40 to 50% of the cases there is a direct communication between the cyst and the subarachnoid space. This was also the situation in our own patient with an extradural cyst.

INTRADURAL CYSTS Spiller, Musser, and Martin (1903) were the first to report the successful surgical treatment of a patient with an intradural cyst compressing the spinal cord. Skoog in 1915 reported two similar cases, stressing their rarity. Perret, Green, and Keller in 1962, described two cases of primary intradural arachnoid cysts which were unassociated with arachnoiditis. Teng and Papatheodorou in 1966, described 12 cases with intradural cyst; four of which had been reported by Teng and Rudner in 1960. In a review of the literature of the past 60 years they found only 21 individual cases, but stated that this condition is not rare and that such cysts are probably missed in prone myelography and remain undiagnosed. In a control study of 97 patients, in whom myelography was performed for prolapsed lumbar disc, small diverticulae were found in 44 cases. Although small cysts seldom produce symptoms, in two other instances the diverticulae were large and yet were asymptomatic. The average age of these patients is about 35 years and the most common site is in the mid-thoracic region. Cysts do occasionally occur in the cervical region (Hoffmann, 1960).

Aetiology The mode of origin of these cysts is obscure, but they are not caused by adhesive arachnoiditis. This has been proved by the absence of features of arachnoiditis on myelography, by the normal appearance of the cord at operation, and by the microscopic examination of the excised cyst, which shows no evidence of cellular infiltration or proliferation. Perret *et al.* (1962) maintain that such cysts are unassociated with arachnoiditis and arise from or within the septum posticum in the posterior subarachnoid space. A history of trauma to the spine is cited in many patients, which could possibly be either a causative or a contributory factor in the formation of these cysts.

DISCUSSION

Why these cysts almost always appear in the mid-thoracic region is not understood. Young children and adolescents who present with kyphosis that cannot be satisfactorily explained should be suspected of harbouring an extradural cyst. Pain is said to be slight or absent in extradural cysts (Elsberg *et al.*, 1934) but this was not the case in our patient.

In patients with extradural cysts the paraplegia is accompanied by an indistinct sensory level. This was well demonstrated by our case with an extradural cyst in whom, despite severe motor signs, there was no definite sensory level. Hence, in a patient with spastic paraparesis without a definite sensory level, the possibility of an arachnoid cyst should be considered. Remission of symptoms may occur spontaneously or after some orthopaedic measure. This is well shown by our case with an intradural cyst but such remissions are also well documented with extradural cysts (Wise and Foster, 1955). It is because of these remissions and absence of pain that such cases are sometimes first suspected of suffering from multiple sclerosis or other degenerative disease. This intermittency seems most probably to be related to variations in the volume of the cyst contents. A partial or complete block of the spinal subarachnoid space, as determined by Queckenstedt's test, is present in about 50% of the cases and the same applies to raised protein levels in the cerebrospinal fluid. However, the cerebrospinal fluid proteins are more consistently raised with extradural than with intradural cysts, in which the chemistry is usually normal.

Radiological investigations have included plain radiography of the spine and myelography. The majority of cases in which the cyst is within the dura mater cause no significant abnormality on the plain films. However, when the cyst is situated extradurally, it tends to erode the pedicles, which lose their normal inward convexity and appear flat or concave. There is nothing specific about these changes which may be seen with any other intraspinal space-occupying lesion. A definitive diagnosis is usually established by myelography. During this examination some of the Myodil collects in the cyst and, by altering the degree of table tilt, the cyst may be outlined. It is important to follow the flow of the Myodil column in both a cranial and a caudal direction, because the cyst may fill in one and not in the other procedure. In a few cases the cyst was outlined only when the examination was carried out in the supine position. Both intradural and extradural cysts give the same myelographic appearances. They are usually pear-shaped and lie on the dorsum of the cord, having a single orifice to the subarachnoid space. Both occur more frequently in the dorsal region and are usually single. Their patent communication with the subarachnoid space explains

the frequent relationship of symptoms to certain positions of the patient's body—that is, exacerbation in the erect position and relief when recumbent. This is commonly seen in intradural cysts.

SUMMARY

Two cases, one with extradural and one with intradural congenital arachnoid cyst of the spine are described. The literature is reviewed.

The inconsistency of the level of sensory disturbance with fairly advanced motor weakness of the legs and the spontaneous periods of remission are stressed in these lesions.

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