'Meningoceles' and 'meningomyeloceles' (ectopic spinal cord)

Clinicopathological basis of a new classification

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While the terms 'meningocele' and 'meningomyelocele', as defined in a dictionary, appear clearly separate, these words have been qualified and assigned nuances by a variety of workers, rendering them somewhat confusing today. Thus for instance, a dictionary (Dorland, 1965) defines a meningocele as 'a hernial protrusion of a part of the meninges and substance of the spinal cord through a defect in the vertebral column'. This, however, appears to be an over-simplification, since in most cases the lesion of the spinal cord is more than a simple herniation into a sac. In fact, as shown by Cameron (1956) in his comprehensive investigation, the initial malformation in all cases of 'meningomyelocele' was an open myelocele which tended to be covered in time by an epithelialized scar. The classification of the congenital lesions of the spine and cord, associated with a spina bifida, proposed by Cameron and endorsed by Willis (1962) and others (for example, Greenfield, 1963), is as follows:

1. Spina bifida occulta, where the cord remains inside the canal.
2. Meningocele, where a cystic swelling is formed by dura and arachnoid and the cord remains in the canal.
3. Meningomyelocele, where the cord is closely applied to the fundus of a cystic swelling.
4. Myelocele, where part of the spinal cord is exposed to the surface. Willis also uses the term 'local myeloschisis' for the last category of Cameron, and further includes a fifth category of 'total myeloschisis' where almost the whole of the spinal canal is open. Both Cameron and Willis have thus made it clear that most if not all meningocele are epithelialized myeloceles. McMenemey (1967) uses the term 'rachischisis' in place of myeloschisis, while essentially adhering to this classification.

The present communication attempts a new classification of what have hitherto been described as 'meningoceles' and 'meningomyeloceles', on the basis of our clinicopathological observations of the last four years. It was apparent to us that the presence or absence of involvement of the cord and the roots constituted the basis of the anatomical lesion, the neurological deficit, and the nature of any surgical intervention. The two main lesions that impressed us were a tethering of the lower end of the spinal cord in some cases of meningoceles and 'meningomyeloceles', and the gross exposure of the spinal cord, best described as an ectopia, which is the hall-mark of the 'meningomyelocele'. It is also suggested that the latter term be dropped altogether in favour of the term meningocele with ectopic spinal cord. These two varieties of congenital anomaly of the spinal canal, meninges, and the cord, along with other similar anomalies, will be included in the proposed new classification of meningoceles.

MATERIAL AND METHODS

Fifty-one consecutive cases of spinal meningoceles of all types operated on by one of us (V.C.T.) constitute the present material. The neuropathologist (D.K.D.) assisted at many of these operations. Included in the material are two post-mortem specimens, which were available for complete dissection, one of a full-term baby with ectopic and tethered spinal cord, and the other of a 6 month foetus with total ectopia of the spinal cord (total myeloschisis), who also had iniencephaly and a completely exposed brain.

SURGICAL METHODS All babies with ectopic spinal cord were operated upon as soon after birth as possible; many babies with meningoceles were also operated on soon after birth. In the babies with ectopic spinal cord the operation consisted simply of providing a

skin cover to the exposed cord under local anaesthesia. No tissue was excised. The thin membrane attached to the spinal cord was buried along with it. No attempt was made to close the dura over the spinal cord. Skin and subcutaneous tissue were stitched in two layers, using the thick ridge of subcutaneous tissue which is usually present in these babies at the periphery of the defect. Whenever the cord was found tethered to the sacral vertebrae, it was released by dividing the tethering band as far down from the lower end of the cord as possible.

In the cases of meningocele without ectopic spinal cord, whenever the defect in the bony spines was small there was no opportunity to inspect the inner side of the spinal canal and the meningocele was excised at its neck flush with the posterior spines. However, when the defect in the spine was wide, it permitted inspection of the spinal canal and the dura was opened to find out the nature of the anomaly of the spinal cord and roots. Whenever the spinal cord or roots were found adherent to the stalk or the base of the mass, the stalk was divided a little distance above the level of the nervous tissue. The part of the stalk bearing this tissue was gently pushed back into the canal and the dura mater and the skin closed over it in layers.

Further details of the operative findings will be presented in the description of the different subgroups.

PATHOLOGICAL METHODS The biopsy specimens were fixed in formalin and blocked in paraffin. The sections were stained with haematoxylin and eosin for cellular details and with the Picro-Mallory method for connective tissues. Occasionally phosphotungstic-acid haematoxylin staining for glial elements and Kluver-Barrera's method for nerve cells and myelin were employed. When eosinophilic or amphophilic material was found accumulated in masses or in cells, periodic-acid-Schiff stain, Krajian's method for mucin and/or amidoblack for protein, were used. Similar procedures for fixation, embedding, and staining were carried out on necropsy material also. The entire fetuses were fixed in formalin in the two instances where they were available. The exposed or affected spinal cord was sectioned at all levels.

OBSERVATIONS

The fifty-one cases could be grouped into two main categories and into nine sub-categories according to the anatomical lesion. The two main categories were based on the absence or presence of an ectopic spinal cord. The second group was characterised by ectopia together with extensive and wide posterior spina bifida and corresponding deficiencies of the posterior spinal meninges and skin. Clinically, such cases manifested significant neurological deficit, generally paralysis of the legs and of the sphincters, soon after birth. In contrast, the first group was characterized by the spinal cord remaining within the dural sac and

spinal canal, with or without a tethering of the lower end of cord or roots. The clinical presentation here varied from complete neurological normality to extensive neurological deficit.

We shall now consider the various subgroups of the two main groups in the following order:

- IA. Simple meningocele
- IB. Meningocele with aberrant neural tissue
- IC. Meningocele with external fistula
- ID. Meningocele with haemangioma
- IE. Meningocele with tethered roots
- IF. Meningocele with tethered cord
- IIA. Meningocele with ectopic spinal cord
- IIB. Meningocele with ectopic spinal cord and tethering
- IIC. Meningocele with total ectopia of spinal cord

SUBGROUP IA SIMPLE MENINGOCELES (Fig. 1a) The four cases in this subgroup presented with a cystic swelling which was translucent, and which at operation was found to have a narrow stalk emerging out of a single spine. The defect in the spine was so narrow that clinically it could not be palpated as a spina bifida and, even after excising the mass and pushing in the stalk, it would not admit the tip of a finger. There was no neurological deficit pre-operatively or post-operatively in any of these. Histological examination carried out in three of these cases revealed the structure of a simple meningocele, without any neural components. These lesions varied in size and in site, being encountered in dorsal, lumbar, or sacral spine. The amount of skin cover also varied, being incomplete in two, but this had no relationship to the histological features.

SUBGROUP IB MENINGOCELE WITH ABERRANT NEURAL TISSUE (Fig. 1a) The four cases in this subgroup also presented with a cystic swelling which was translucent, and which at operation was found to have a narrow stalk emerging out of a spina bifida in a single spine which was as narrow as in subgroup IA. Again there was no neurological deficit either before or after the operation. This group also showed the variation in site and size of lesion and in skin cover, as in subgroup IA.

The feature which distinguished this subgroup from subgroup IA was the presence, on histopathological examination, of aberrant nervous tissue, in one form or another. In one instance there were ependymal canals and large neurones (Fig. 2), suggesting the presence of cord tissue. However, as there was no neurological deficit before or after operation and as the cord was not even visualized at operation (in the D4 region), this was obviously aberrant central nervous tissue. In two other instances, besides abnormal neural or neuroglial tissue, derivatives of the mesodermal layer, such as abnormal smooth muscle masses or hamartomatous vessels (Figs. 3a, b) were observed. The fourth case with a fundal ulcer, showed, besides evidence of acute inflammation, one spinal ganglion with rootlets (Fig. 4).

a SIMPLE MENINGOCELE: Subgroups IA,B

b MENINGOCELE WITH EXTERNAL FISTULAS: Subgroup IC

c MENINGOCELE WITH TETHERED ROOTS: Subgroup IE

d MENINGOCELE WITH TETHERED CORD: Subgroup IF

e ECTOPIC SPINAL CORD: Subgroup IIA

FIG. 1. Main operative findings, in diagrammatic sagittal sections, in the various subgroups of meningoceles. U = upper end. L = lower end. The curved lines within the mass in (a) and (b) indicate the fibrous septa frequently found.

FIG. 2. (NP-D-830). Aberrant cord tissue and ependyma lined canals in otherwise simple meningocele (H and E, $\times 66$).

FIG. 4. (NP-E-415). Part of an incipient spinal ganglion and some root tissue in a cystic space in the deep subcutis of a meningocele (H and E, $\times 65$).

SUBGROUP IC MENINGOCELE WITH EXTERNAL FISTULA (FIG. 1B) Both the cases in this subgroup presented with a mass in the lumbar region, continually discharging cerebrospinal fluid (CSF) through an opening on the surface. Around the opening one case showed a raw irregular surface and the other had good skin cover. At operation the discharging fistula was seen to be the outer end of a track leading down to the neck of the lesion in the dural canal. There was no neurological deficit either before or after operation, although the one specimen which was examined histopathologically revealed some abnormal degenerating root tissue. Both these children died of infection.

SUBGROUP ID MENINGOCELE WITH HAEMANGIOMA The single case in this group presented with a cylindrical swelling sticking out like an umbilical cord above the skin surface, through a single (L4) spina bifida. The swelling consisted of a bluish vascular mass covered by a thin membrane. At operation, the mass was found attached to the dura by a firm stalk which filled the

defect in the spine. Histologically, it was a cavernous haemangioma.

SUBGROUP IE MENINGOCELE WITH TETHERED ROOTS (Fig. 1c) Of the five cases in this subgroup information of the anomaly at the first operation is available only in three cases, the other two being included on the basis of a retrospective analysis. In the former three cases, the swelling was partly cystic, with a central firm tender mass, and it was only partially translucent. While clinically the spina bifida did not appear to be large, at operation the defect was clearly wider than in the preceding subgroups of cases, though confined to one spine. The defect was wide enough to admit the thumb. The characteristic feature on opening the dura was the presence of recognizable nerve roots closely adherent to and forming a part of the deeper portion of the stalk. These roots formed a part of the fibrovascular matrix of the stalk. Histopathological examination of the carefully dissected supra-radicular portion of the stalk and of the meningocele revealed

(a)

(b)

FIG. 3. (NP-D-973). (a) Nests of smooth muscle fibres and (b) hamartomatous vessels in fibrous matrix in the deep subcutaneous part of meningocele (H and E, $\times 66$).

only fibrous tissue. Specimens from two other cases showed in one instance an epidermoid cyst which had formed by a process of invagination (Fig. 5) and in the other some abnormal collections of striated and smooth muscle fibres.

FIG. 5. (NP-E-760). Branching epidermoid cyst entering subcutaneous tissue of meningocele (H and E, $\times 66$).

The meningocele in two of the cases had complete skin cover, in one there was a small raw area with surrounding thin membrane, another had a large area covered by thin dry membrane and in the fifth no information was available. When the membrane was present, it was encircled by a thick ridge of subcutaneous tissue, much like the ridge seen in ectopic spinal cord (see later).

The two cases which have been included on retrospective analysis had neurological deficit which was believed to have resulted from the first operation. The deficit was in the form of sphincter disturbance and perianal anaesthesia only, suggesting the involvement of a few sacral roots. In one of these cases the spina bifida was noted at the second operation to be wide, but restricted to one spine.

SUBGROUP IF MENINGOCELE WITH TETHERED CORD (Fig. 1d) The meningocele in all five cases presented as a large tender swelling in the lumbar or lumbosacral spine. All had a wide spina bifida, clearly palpable as a defect of at least three spines. Four of the five patients who were between the ages of $1\frac{1}{2}$ years and 9 years at the time of examination manifested neurological signs in the form of paraparesis, wasting, sphincter disturbance, and anaesthesia. Maximal wasting with contractures and trophic ulceration were seen in the oldest child. By contrast, the fifth case examined at the age of 13 days showed no neurological signs.

At operation in all the five cases the mass was found adherent to the dural sac and through it to the spinal cord. This meant that the spinal cord was down at the level of the bony defect which was in the lower lumbar or sacral spines. This constituted a form of tethering down of the spinal cord. While in four of the cases it was the lower end of the cord which was seen to expand out to form the deeper part of the stalk of the mass, in one instance the base of the mass was adherent to a slightly higher level of the cord which had been pulled out into the mass (Fig. 6).

FIG. 6. View at operation of the meningocele (L4 to S1) as seen from the right side of the patient. The tethered spinal cord has humped out of the spinal canal and is adherent to the under surface of the mass (which is gently hooked to the left); the roots are also stretched and tethered. The cord assumes an inverted 'U' shape as it extends caudally (towards the right) beyond the tether.

FIG. 7. (a) Anteroposterior and (b) lateral views of myelogram, showing the wide dural sac. (b) shows the spinal roots coursing horizontally from the spinal cord, which was found adherent to the posterior wall of the dural sac. (a) shows the lower end of the cord as a filling defect on one side of the midline.

Clinically, two cases showed peripheral cystic components to the swellings which at operation were confirmed to be meningeal sacs filled with CSF. The central firm and tender part of the swelling was directly adherent to the dural sac underneath and was capped by a lipomatous mass. In two other cases the swelling was soft and non-cystic throughout. At operation, this swelling consisted mainly of a lipomatous mass with a central stalk. The fifth case, who had an earlier operation elsewhere for excision of a 'lipoma', probably belonged to the second category. The latter three cases had a bulky dural sac, although it was accommodated in the widened spinal canal. This feature was obvious on the plain radiographs of the spine, in the form of widened pedicles.

Myelography done in one of the cases demonstrated the enlarged dural sac (Fig. 7a, b).

In two of the three cases where an operated specimen was available for histological examination, small irregular islands of aberrant neurological tissue were encountered amidst the fibrous tissue of the stalk and of the deeper part of the fatty cap respectively (Fig. 8). In the third case, clear teratomatous tissues were detected with abnormal entodermal (intestine-like), mesodermal (cartilaginous), and ectodermal (abnormal CNS tissue) elements (Fig. 9a, b, c). In none of the cases was there a true lipoma. As four of the five cases showed a clear improvement in the neurological status post-operatively, and the child without any pre-operative deficit showed

FIG. 8. (NP-D-860). Irregular islands of neuroglial tissue amid masses of collagen in the deeper part of the fatty cap of the meningocele with tethered cord (Kluver-Barrera, $\times 66$).

FIG. 9b

FIG. 9c

FIG. 9a

FIG. 9. (NP-E-573). (a) Mucin-laden cells of abnormal epithelium; (b) irregular masses of cartilage; and (c) nervous tissue with attempted lamination of some of the cells: in meningocele with tethered cord. ((a) Krajan's mucicarmine stain, $\times 105$; (b) same stain, $\times 40$; (c) H and E, $\times 66$).

no worsening, it became obvious that the neural tissue removed was aberrant.

SUBGROUP IIA MENINGOCELE WITH ECTOPIC SPINAL CORD (MENINGOMYELOCELE) (Fig. 1e) As expected, this formed the largest subgroup and consisted of 22 cases. As stated at the outset, the essential morbid anatomical feature of these cases was an exposure or ectopia of spinal cord of greater or lesser extent. When examined soon after birth, the spinal cord was found to be without any covering in the midline. A thin membrane extended from the border of the exposed cord to the skin edge. In several cases, the upper skin edge was directly adherent to the underlying spinal cord. When examined some time after birth, the thin membrane as well as the spinal cord, were seen to be covered by granulation tissue or even frank slough. The only case operated on at the age of 4 months showed an epithelialized membrane and spinal cord.

The spinal cord generally ended at a level lower than the usual, even though it was not tethered down by the filum terminale. In one case the cord ended at the level of S4 vertebra—that is, below the level of the lower end of dural sac at S2, and it was adherent to the posterior surface of the bodies of S3 and S4 vertebrae. The margin of the lesion was well defined, being made up of a thickened ridge of subcutaneous tissue, which appeared to be at the junction of the dura and the deep fascia. The spina bifida, as assessed both clinically and at operation, was very wide in all the cases and extended over more than three spines.

As expected, all cases presented with severe neurological deficit, generally paraplegia, sphincter disturbance, and anaesthesia. That this was noticeable even in children examined six hours after birth pointed to the vulnerability of the exposed cord.

As the surgical procedure was entirely reconstructive, no material was biopsied for pathological examination, except for a small bit of dura in one case.

SUBGROUP IIB MENINGOCELE WITH ECTOPIC SPINAL CORD AND TETHERING There were seven cases in this subgroup, one of which was available for dissection after death. They all presented with an exposed spinal cord in a manner identical with that described in the previous subgroup. The interesting feature in all these cases was the presence of a prominent filum terminale of varying thickness. In the necropsied case it was as thick as the lower end of spinal cord (Fig. 10). At operation, the filum terminale was attached to the sacrum in a variety of ways. The exact nature of this attachment was not clear in two cases, as the lesion was higher up and there was no opportunity to examine the posterior surface of the sacrum. In three, the attachment was through a thick fibrous band, while in the other two it was through a hard nubbin filling the lower end of the gap in the bifid sacral spine (Fig. 11).

In four of the cases a biopsy specimen of the tethering tissue was available for histological examination. In two of these specimens cord tissue could be recognized, one containing ependymal cell groups within possibly ill-formed homogeneous cord tissue (Fig. 12), and the

FIG. 10. (NP-E-338). Dissected post-mortem specimen of lumbosacral region of 7-day-old baby, showing thin atrophied spinal cord (in the middle of the upper edge of the picture) tapering down to a thick filum terminale, which then ends in the oval fibrous mass attached to the sacrum. (On the right of this is another irregular elongated white mass, which is the iliac bone and which separates the cauda equina region from the dissected right gluteal region.)

other including mature spinal cord with anterior horn cells and both spinal roots (Fig. 13). The cord tissue in the former seemed aberrant both from its appearance and from the observation that the child did not show obvious clinical worsening after operation. In the third case, the cartilaginous nubbin removed at operation was confirmed to be cellular cartilage. In the fourth case, the nubbin was found to consist of posterior root and ganglionic tissue embedded in dense fibrous matrix.

All the cases in this subgroup were examined and operated on in the first three days of life. They presented with wasting of one or both legs and a talipes deformity. This was not noticed in the cases belonging to the previous subgroup. As all these cases also showed the tethering of the cord, this was probably responsible for the wasting. Five of the seven cases showed a kyphosis of the vertebral column at the site of the lesion.

SUBGROUP IIC MENINGOCELE WITH TOTAL ECTOPIA OF SPINAL CORD The only case in this group was a stillborn foetus, who presented with a frog-like face and a grossly ill-developed and exposed brain, looking like a combination of iniencephaly and anencephaly.

DISCUSSION

As stated at the outset, this study was prompted by the lack of clarity on the true nature of so-called 'meningoceles' and 'meningomyeloceles'. We have attempted to show how the term 'meningocele' can legitimately include several varieties of congenital pathological lesions involving the spinal cord, roots, meninges, and vertebrae in dorsal, lumbar, and sacral regions. These lesions include

FIG. 11. The attachment of filum terminale in ectopic spinal cord with tethering.

FIG. 12. (NP-D-767). Ectopic spinal cord with tethering; biopsy specimen from S2-3 level. Core of ependymal cells in what looks like a widened central canal in a ring of (?) ill-formed cord tissue; the cells in this could be fibroblasts or altered astrocytes (H and E, $\times 25$).

all congenital swellings that present through posterior spina bifida. Lesions that remain strictly within the spinal canal, such as diastematomyelia, are not included here. Where the exposure of the spinal cord was the main feature of the lesion, with only a thin leptomeningeal membrane covering it, we have described the condition as a meningocele with ectopic spinal cord (our group II) to replace the older inadequate terms 'meningomyelocele' and 'myelocele'. For all practical purposes this group may be referred to as ectopic spinal cord (ESC) only. The rest of the cases, without the exposed spinal cord, comprise our group I and this may be referred to as meningocele.

The essential morphological features of the subgroups of group I are summarized in the drawings (Fig. 1). As detailed earlier, the common feature of subgroups IA, IB, IC, and ID was a narrow defect in a single spine. The important variable features were the occasional inclusion of aberrant CNS tissue within the mass and the extent of skin cover. When there was incomplete skin cover and some granulation covered the raw area, the appearance simulated that of ectopic spinal cord. However, the correct diagnosis could be readily established by palpation of the size of the bony defect. One of the varieties of 'open spinal meningocele' described by Chapman (1967), is very similar to that described by us as meningocele with external fistula (subgroup IC). He too has emphasized the proneness to infection of these cases.

Considering these four subgroups together, six out of 11 cases showed abnormal inclusions of tissues of ectodermal, mesodermal, or endodermal origin, five of these being clearly tissues pertaining to spinal cord or roots (subgroups IB and IC); the sixth was a haemangioma (subgroup ID). Such an inclusion of neural tissue, though aberrant and producing no clinical deficit on excision, would seem to be inconsistent with the hitherto accepted definition of a meningocele as simply 'a hernial protrusion of the meninges' (Dorland, 1965).

Subgroups IE and IF had the common feature of a wider spina bifida (Fig. 14). It seems noteworthy that, at least in our experience, a wide defect in a single spine (subgroup IE) has always been in the lower lumbar or sacral regions, with tethering of

FIG. 13. (NP-E-366b). Ectopic spinal cord with tethering; biopsy specimen from S2 level. Mature cord tissue with central canal in the centre and groups of anterior horn cells. The haemorrhagic areas are a result of the operative procedure (H and E, $\times 25$).

roots only; while a still wider defect in several spines (subgroup IF) was always associated with a tethering of the cord itself. Another important clinical feature was tenderness on palpation of the mass, which could be elicited even in the new born, and which disappeared after operation. At operation, especially when the mass was tender or when the spinal defect was wide, the dural sac was opened and the stalk of the mass was carefully examined for the presence of adherent roots or cord.

The tethering of the cord or roots to the under-surface of the meningocele was probably responsible for the neurological deficit in the cases of subgroups IE and IF. The two cases of subgroup IE, where parents reported neurological deficit, had probably suffered inadvertent damage to the roots during the first operation. In the subgroup IF, as the neurological deficit was most pronounced in the oldest child and was non-existent in the new born, it seems reasonable to presume that greater damage to the tethered cord resulted from the increase with growth in the length of the spinal column. Walker (1944) suggested the same mechanism for the progressive neurological deficit in a 23 year old patient with intraspinal lipoma adherent to the lower end of cord and roots. Streeter (1919) showed that when the embryo reaches 30 mm in length, there begins a disproportion in the rate of growth between the vertebral column and the spinal cord, the former elongating rapidly.

It is interesting to note that in two of the three cases of subgroups IE and in all three cases of subgroup IF, where biopsy specimen was available from the first operation, some abnormal or hamartomatous component of one or more of the three germinal layers was detected. All three cases of

subgroups IF showed aberrant neuroglial tissue, with, in one, evidence of teratomatous tissue as well. Thus, in a general way, meningoceles of subgroups IE and IF compared with those of subgroups IA to ID in respect of their content of aberrant tissues. These histopathological findings indicate that the defect in development of the spinal neural tube which results in the formation of the meningoceles, arises early in foetal life.

Our observations also provide evidence that the fibro-fatty mass which forms the body of these meningoceles (two cases of subgroup IE and all five of subgroup IF), is not a true 'lipoma'. In virtue of its content of a variety of hamartomatous tissues, it is at best a hamartoma. This has also been suggested by Bouton, Martin, and Rickham (1966). The term 'lipoma of cauda equina', as used among others by Dubowitz, Lorber, and Zachary (1965), is best dropped. They have reviewed the literature well and shown how a fibro-fatty mass adherent to the lower end of the cord has been known for over 100 years. Their cases, as also many of those described by James and Lassman (1960) under the term 'spinal dysraphism', are very similar to those of our subgroup IF. They could be described better under the term 'meningocele with tethered cord', as we have used it. The entity 'meningocele with tethered roots' does not seem to have been described before.

A comparison of the drawings of the morphological defect in subgroup IF with that in subgroup IIA shows the extent of similarity between them. The common features are a wide defect in the spine extending over at least three vertebrae (Fig. 14), the presence of the spinal cord in the lesion even when that is in the lower lumbar or sacral region, and a consequent horizontal disposition

FIG. 14. Diagrammatic view as seen from behind at operation (and also radiologically) of the range of defects in posterior spines. The wide defect in subgroups IF, IIA, and IIB often extended over more than three spines.

of some of the roots of the cauda equina in a wide intraspinal meningeal sac (Fig. 1). The most significant feature differentiating group II from subgroup IF was the absence of the skin cover and the underlying mass, with resultant ectopia of the spinal cord. As indicated in the introduction, the term 'meningomyelocele' appears to us highly unsatisfactory. While Cameron (1956) in his careful study of necropsy specimens of the spine and the cord in 'spina bifida cystica' demonstrated clearly that the basic lesion in all cases of so-called 'meningomyelocele' was in fact an open 'myelocele', he unfortunately retained the former term in his final classification. Most subsequent medical literature on this subject has tended to perpetuate this terminology. Gardner (1968) in a recent evaluation, rightly drops the term 'meningomyelocele' in favour of 'myelocele', which he prefers 'because neural tissue constitutes the primary component of the sac, the meningeal portion being entirely secondary'.

The neurological deficit in cases of this subgroup was noticeable even in infants examined six hours after birth. This points to the vulnerability of the exposed cord before birth, during labour, and after birth. Sharrard, Zachary, Lorber, and Bruce (1963) clearly demonstrated that children operated on during the first two days of life showed much less residual paralysis than those operated on at the age of 6 months. Considerations of the anatomy of this group of ectopic spinal cord and the practical implications of it have been published elsewhere (Talwalker, 1967).

The seven cases of subgroup IIB presented a clinical picture similar to that in cases of subgroup IIA, with the notable exception of the presence of wasting of the leg muscles and of a talipes deformity. Many also showed kyphoscoliosis. While the spinal lesion here was again similar to that in subgroup IIA, at operation some form of tethering of the lower end of the spinal cord was witnessed in all except one. The tethering mechanisms, summarized in Fig. 11 were different from those encountered in subgroup IF, in that the tethering bands and nubbins were now found attached to the posterior surface of the body of the sacrum. This was reminiscent of the attachment of the normal filum terminale which, in fact, appeared to be thickened. Streeter (1919) has shown that the 'filum terminale represents that portion of the spinal cord, caudal to the second coccygeal segment, which has undergone dedifferentiation and has finally become converted to a fibrous strand'. It is conceivable that there was an arrest or inhibition of this dedifferentiation in the cases we are describing here, with the result that the filum terminale did

not become completely thinned out and fibrosed, but retained some neural tissue. The latter was evident in the histological examination of the tethering tissues, as described under 'Observations'. The relation between this anomaly and the wasting of leg muscles is not clear. The wasting may be due to the tethering and consequent stretching effect on the cord during intrauterine development. It must be pointed out that in one case of subgroup IIB, where tethering was not noticed, it might have been masked by granulation tissue or been actually absent.

As the number of cases on which our classification is based is rather small, it is possible that there may be a few other subgroups which we may not have either come across or recognized. To that extent our classification remains open to elaboration.

SUMMARY

A clinicopathological study is presented on 51 cases of various types of 'meningocele' and 'meningomyelocele'. The 21 cases of 'meningocele' (group I) fell into the categories of (A) simple meningocele (four cases), (B) meningocele with aberrant neural tissue (four cases), (C) meningocele with external fistula (two cases), (D) meningocele with haemangioma (one case), (E) meningocele with tethered roots (five cases), and (F) meningocele with tethered cord (five cases).

The term ectopic spinal cord is suggested in place of 'meningomyelocele' (group II), as the former term describes the main feature of the lesion. Of the 30 cases of ectopic spinal cord, 22 were of the usual variety (IIA), seven showed ectopic spinal cord with tethering (IIB), and one case had total ectopia of the cord (IIC).

The frequent involvement of spinal cord and root tissue, often of an aberrant or hamartomatous nature, in group I, besides their invariable involvement in group II, point to a similarity between the two groups, which have therefore been included under the common designation of meningoceles.

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The February 1970 Issue

THE FEBRUARY 1970 ISSUE CONTAINS THE FOLLOWING PAPERS

- The cerebrospinal fluid homovanillic acid concentration in patients with Parkinsonism treated with L-dopa
G. CURZON, R. B. GODWIN-AUSTEN, E. B. TOMLINSON, and B. D. KANTAMANENI
- Relief of intention tremor by thalamic surgery
KHAIRY SAMRA, JOSEPH M. WALTZ, MANUEL RIKLAN, MAXIM KOSLOW, and IRVING S. COOPER
- Simple and complex finger movement performance of patients with Parkinsonism before and after a unilateral stereotaxic thalamotomy
E. PERRET, E. EGGENBERGER, and J. SIEGFRIED
- Colour imperception in unilateral hemisphere-damaged patients
GIUSEPPE SCOTTI and HANS SPINNLER
- The familial incidence of intracranial aneurysms
VIJAY K. KAK, COLIN A. GLEADHILL, and IAN C. BAILEY
- A study of a patient who had alexia without agraphia
W. J. K. CUMMING, L. J. HURWITZ, and N. T. PERL
- Changes in motor innervation and cholinesterase localization induced by botulinum toxin in skeletal muscle of the mouse: differences between fast and slow muscles
L. W. DUCHEN
- The incidence of abnormality in control human peripheral nerves studied by single axon dissection
N. ARNOLD and D. G. F. HARRIMAN
- Remarkable recovery of a steroid-responsive recurrent polyneuropathy
DARRYL C. DEVIVO and W. KING ENGEL
- Changes in the forearm associated with median nerve compression at the wrist in the guinea-pig
MARGARET H. ANDERSON, PAMELA M. FULLERTON, R. W. GILLIATT, and J. E. C. HERN
- The prediction of recurrence in meningiomas
M. RUFUS CROMPTON and PETER C. GAUTIER-SMITH
- Uric acid and serum lipids in cerebrovascular disease
Part 2: Uric acid-plasma lipid correlations
JOHN PEARCE and HASAN AZIZ
- Lymphocyte transformation induced *in vitro* by PHA and PPD in multiple sclerosis
W. CENDROWSKI
- Subdural haematoma after the treatment of chronic hydrocephalus by ventriculocaval shunts
R. D. ILLINGWORTH
- A comparison of methohexitone and thiopentone in electrocorticography
R. PAUL and RUTH HARRIS
- Congenital spinal arachnoid cysts. Report of two cases and review of the literature
IFTIKHAR A. RAJA and JOHN HANKINSON
- Steroid myopathy complicating McArdle's disease
F. L. MASTAGLIA, J. P. K. MCCOLLUM, P. F. LARSON, and P. HUDGSON
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