SPINA BIFIDA CYSTICA AND OCCULTA

Some Aspects of Spinal Dysraphism

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In recent years the treatment of spina bifida cystica has altered in various ways (Forshall & Rickham, 1960; Sharrard *et al.*, 1963). Surgeons are now tending to operate at birth, and orthopaedic surgeons are being asked to examine the children prior to operation. A much more intelligent interest is being taken in the urological complications and other congenital anomalies present in these children, *e.g.* hydronephrosis and dislocation of the hip. It is high time that the whole problem was viewed from the standpoint of embryology and the knowledge obtained from operations on cases of spina bifida occulta of which we have a series of over 80.

At present anyone hearing mention of spina bifida will immediately think of meningocoeles and myelocoeles, but there is also spina bifida occulta. Both kinds of spina bifida are forms of spinal dysraphism—both have the same origin—failure of complete development of the median dorsal region of the embryo.

By definition, any form of spina bifida has incomplete development of the vertebral elements. It is only the degree of failure of development of embryonic ectoderm, mesoderm and endoderm which determines whether any particular foetus will have spina bifida cystica or spina bifida occulta.

In spina bifida cystica the most important abnormality is the primary dysplasia of the spinal cord, but there are also associated extrinsic abnormalities including the ones which call attention to the patient. As Warkany *et al.* (1958) has shown in the rat, the spinal cord tissue which is actually formed in a meningomyelocoele is to some extent resorbed and degenerates so that isolated areas of neural tissue are to be found in the sac wall. Where there has been degeneration there will be attempt at repair and consequently we see gliosis, particularly in the subpial marginal nervous tissue. This gliosis will lead to strangulation of functioning tissues to some extent and will increase the amount of dysfunction. Because there has been delay in the formation of the spinal cord there is also delay in the formation of meninges, bones, muscles, and skin. The embryonic tissues attempting to form these structures are frustrated and consequently are formed incompletely. Some of the embryonic tissues develop as aberrant structures which may be partly functional. Those which are not functional will become fibrous tissues. They interfere by pressure or by fixation so that the spinal cord cannot ascend within the vertebral canal in the course of normal growth.

In spina bifida occulta similar processes occur, but in the majority of cases the spinal cord itself is normal although in some there is an element of myelodysplasia: the spinal cord may be bifid in part of its length (diastematomyelia) or there may be degrees of failure of closure of the neural tube so that one finds what can be termed as intrathecal myelocoele. Being intrinsic abnormalities these are not amenable to surgery. The associated aberrations in the development of bone, ligaments, blood vessels, or posterior nerve roots leave extrinsic abnormalities which are much more obvious in spina bifida occulta than in spina bifida cystica and cause clinical abnormalities and neurological deficit later in life.

These same extrinsic elements seen in spina bifida occulta may also occur in

spina bifida cystica, but as their existence is not commonly known, they have not yet been positively identified, except that a septum in association with diastematomyelia may be found. In the surgical treatment of spina bifida cystica therefore, it is insufficient simply to close the sac. The spinal cord and nervous elements must be released and any non-functional aberrant tissues removed. If this is not done these tissues are liable to fix the spinal cord and nerve roots and later produce the types of neurological deficit which have been found in spina bifida occulta.

The borderline between the two forms of spina bifida is very difficult to define. We have operated on about 80 cases of what we considered to be spina bifida occulta but analysis and definition will probably reduce this number to 60. Our diagnosis has been based on the superficial appearance of the back. Cystic swellings and myelocoeles are clearly cases of spina bifida cystica; lipomata in the lumbosacral region, hypertrichosis, naevi, dermal sinuses and small flat umbilicated areas we have regarded as occult cases because the majority have no expansion of the meninges outside the vertebral canal. The transition from a mild form of cystica to the true occult is illustrated by reference to a series of cases which have been operated on. The two cases cited below show the most severe forms of spina bifida occulta.

Case W. R. had a lumbosacral lipoma the pedicle of which attached to the dura and ended by plugging an open intradural myelocoele (fig. 1). The plug was able to be removed and its surface of contact with the spinal cord was lined by ependyma. The spinal cord rested normally against the vertebral bodies and there was no expansion of the meninges outside the vertebral canal.

Case H. J. had a subcutaneous lipoma in the lumbosacral area which was found to contain a small meningocoele with the spinal cord kinked posteriorly and adherent to the dorsum of the sac. The spinal cord was separated from the sac wall and appeared complete, no neural tissue being found in the material taken for microscopy.

The next stage of transition is the meningocoele without neural content. The neck of the sac is narrow or obliterated. We have found these concealed in a few cases of lumbosacral lipoma, their presence being demonstrated on myelography if the neck of the sac is patent. Where the neck is obliterated, a blind cyst is occasionally found in the root of the lipoma pedicle usually outside the level of the dural attachment; the blind cyst is sometimes associated with an epidermal cyst which may be extrathecal or partly intrathecal. There is no expansion of the dura outside the vertebral canal in these cases.

Occasionally there is no trace of a previous meningocoele, but the relation of the spinal cord or cauda equina to the dura suggests that there was one previously or that one very nearly developed. In these cases there is a localised firm adherence of the spinal cord or cauda equina to the dura dorsally so that the neural elements are held away from the vertebral bodies. The most obvious examples of this occur near the termination of the dural sac where the cauda equina is closely involved with the dorsal dura and the filum terminale ends at this point of adherence instead of in the midsacral region. Since the cauda equina continues its course to the outlets in their normal position there is a close resemblance to the doubling-back of the nerve roots on the wall of a meningocoele sac. This finding is commonest in cases of lumbosacral lipomata, their pedicle being usually continuous with the conus medullaris or the filum terminale.

The types of cases which are undoubtedly occult have laminae and spinous

processes commonly consisting of a mixture of fibrous tissue, fat, muscle, cartilage and bone with no meningeal extrusion but this fatty fibrous mass frequently has one or more connections in the midline through the dura to the spinal cord. These connections are aberrant posterior nerve roots sometimes consisting of nerves and sometimes of fibrous tissue. They are frequently associated with diastematomyelia which has no septum and, we believe, produce a traction effect. Our belief is based on the line of their intradural course and the fact that dividing them surgically has produced improvement in some cases with a previous neurological deficit. These

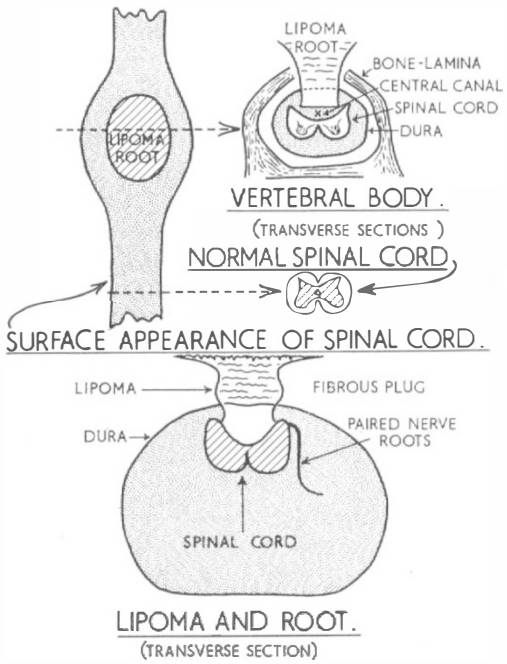

FIG. 1

Case W. R. A lumbosacral lipoma extending intrathecally and plugging an open intradural myelocoele.

aberrant nerve roots are the most likely abnormalities to be found in spina bifida cystica, they will be very difficult to differentiate from the nerve roots and tracts which adhere to the wall of the sac but their chance removal at operation may well account for some of the clinical improvement later which at present is ascribed to late myelination of tracts and fibres.

Because of these aberrant tissues, it is wiser in operating on spina bifida cystica to approach the neck of the sac through the lamina cranial to the junction of the root with the vertebral canal.

Our research on spina bifida occulta is supported by grants from the Medical Research Council and from the Scientific and Research Committee of the Newcastle Regional Hospital Board.

SUMMARY

In status dysraphicus there is dysplasia of the spinal cord and also defective development of the mesoderm and ectoderm in the midline of the dorsum of the embryo. Whilst there is no possibility of correcting the dysplasia of the spinal cord, the abnormalities of the other tissues, so far as they affect the spinal cord and nerve roots, are capable of surgical treatment. The methods of doing this in cases of spina bifida cystica are well known, but it is less well known that the aberrant non-neural tissues may cause delayed effects by interfering with nerve conduction as a result of growth during childhood.

The authors have operated upon more than 80 cases of spina bifida occulta and the types of abnormality found are described and classified. Their relationship to spina bifida cystica is discussed.

RÉSUMÉ

Dans le status dysraphicus il y a une dysplasie de la moëlle épinière ainsi qu'une défficience dans le developpement du mésodèrme et de l'éctodèrme correspondant à la ligne médiane du dos de l'embryon.

Bien qu'il ne soit pas possible de corriger la dysplasie de la moëlle épinière les abnormalités des autres tissus affectant la moëlle épinière et ses racines peuvent être soumis à un traitement chirurgical.

Le traitement chirurgical dans les cas de spina bifida cystica est bien connu, mais ce qui l'est moins est que les tissus aberrants peuvent causer, à retardement, des effets sur la conduction nerveuse resultant de la croissance au cours de l'enfance.

Les auteurs ont opèré sur plus de 80 cas de spina bifida occulta et ont decrit et classifié les differents types d'abnormalités qu'ils ont rencontrés et ont discuté de leurs relations avec la spina bifida cystica.

ZUSAMMENFASSUNG

Im status dysraphicus besteht eine Dysplasie des Rückenmarks sowrie eine Fehlentwicklung des Mesoderms und Ektoderms in der Mitte des Rückens beim Embryo. Obwohl eine Korrektur der Dysplasie des Rückenmarks unmöglich ist, sind die Anomalien der andern Gewebe, sofern sie das Rückenmark und die Nervenwurzeln afficieren, einer chirurgischen Behandlung zugänglich. Die Methoden, soweit sie Fälle von Spina bifida cystica betreffen, sind wohl bekannt, aber weniger bekannt ist die Tatsache, dass aberrante nicht-nervöse Gewebe infolge Wachstums im Kindesalter Spätfolgen auf die Nervenleitung haben können.

Die Autoren haben 80 Fälle von Spina bifida occulta operiert. Die beobachteten Anomalien werden beschrieben und klassificiert und ihr Verhältnis zur Spina bifida cystica diskutiert.

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