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Double Meningocele

It is very unusual to find a meningocele in more than one area: we are, therefore, reporting a case in which there were two distinct meningoceles with a normal intervening spinal canal.

Case Report

The girl was born in March 1970, the 8th child of healthy parents. Her birthweight was 3203 g. At the age of 6 weeks she was referred with a diagnosis of double meningocele.

Over the upper thoracic spine there was a lobulated swelling 5 cm in diameter partly covered by skin and partly by an opaque membrane (Fig. 1). A small daughter cyst 0·7 cm in diameter was present just below and to the right of the main swelling. The tumour was translucent and increased in tension when the child cried and was thought to be a meningocele.

Another swelling 8 × 6 cm was present over the lumbar and sacral spines. This was covered by normal skin and was initially diagnosed as a lipoma as it did not pulsate or increase in tension with crying.

The lower limbs were normal and, as far as could be judged, sphincter function was unimpaired. There was no evidence of hydocephalus and general examination did not reveal any other abnormalities.

First operation. At age 8 weeks the thoracic swelling was explored through a transverse elliptical incision. The superficial part consisted of fat; under this was a bilocular dural sac which contained clear CSF but no nervous tissue (Fig. 2 A–B). The base of the sac was doubly ligated at the level of the laminae and the whole swelling excised. The laminar defect was identified and its edges sutured together with silk, followed by repair of the superficial layers.

On the day after operation, the right foot was noticed to be flaccid though the left was normal. Five days later the left foot became flail, but no further progress of the paraparesis occurred. The wound was slow to heal as it developed some superficial infection from which *Staph. aureus* was cultured.

Second operation. At age 14 weeks, through a longitudinal elliptical incision, the lumbar swelling was explored, and was found to consist of a superficial mass of fat covering a small meningomyelocele (Fig. 2 C–D). When the dura was opened, clear colourless CSF and several nerve roots emerging to fuse with the dome of the sac were seen. The fatty mass was dissected away, and after allowing the CSF to escape, the neural elements

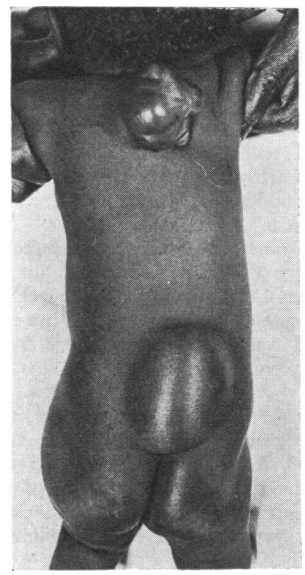

FIG. 1—*Two distinct meningoceles, one dorsal and the other in the lumbosacral region.*

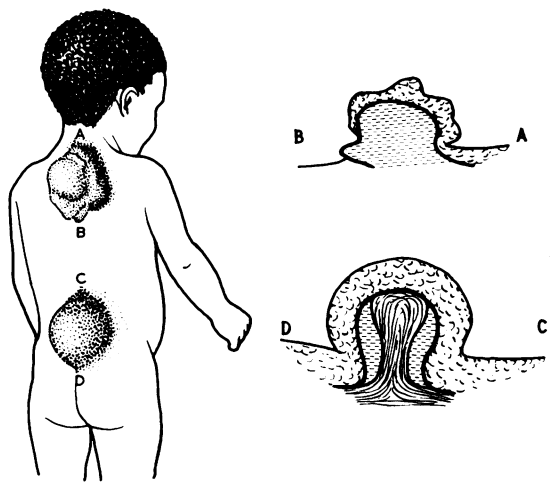

FIG. 2.—*Diagram to illustrate the contents of the meningoceles. The thoracic sac A–B was bilocular containing CSF only. The lumbosacral swelling C–D contained nerve roots attached to the fundus.*

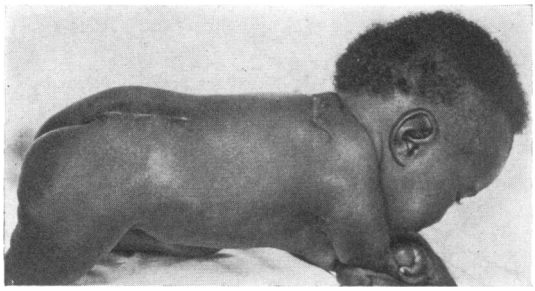

FIG. 3.—*Patient after surgical repair.*

were replaced within the spinal canal. The meninges were repaired and the bony defect covered by two layers of lumbar fascia.

There was some superficial wound infection, as there had been after the first operation, but healing was complete after 17 days (Fig. 3). No change in the neurological state of the legs occurred but it was noticed that the child was passing urine very frequently and was constantly wet.

Follow-up. At the age of 9 months, the child was alert and progressing normally. Her head circumference was 43 cm and there was no evidence of hydrocephalus. The neurological function of the legs had returned to normal, and the mother declared that she was no longer wet and had normal sphincter function.

Discussion

The presence of two distinct spinal meningoceles is very unusual. Potter (1962) says that she has observed 2 cases in the dorsal and lumbar regions, in one of which there was an associated hydrocephalus and Arnold-Chiari malformation. Bertan and Wilson (1968) described a case of combined thoracic and lumbosacral myelomeningocele. This child also developed hydrocephalus after successful excision of both sacs and succumbed to septicaemia after the insertion of a ventriculoatrial shunt.

Occipital meningoceles are often associated with spina bifida of the upper cervical segments, and in severe cases may constitute an encephalomyelocele. In such cases, however, there is no segment of normal spine in between the occipital and the spinal defects.

Fahrenkrug and Højgaard (1963) described the radiological findings in multiple paravertebral lumbar meningoceles, but their patient was not operated upon.

Meningoceles have been described with other congenital anomalies of the vertebral column, while hydrocephalus and the Arnold-Chiari malformation are common associations. Von Recklinghausen's disease is frequently found with intrathoracic meningoceles, but its rare coexistence with lumbosacral meningoceles and meningomyeloceles is regarded as no more than coincidental (Levene, 1959).

Summary

A child with both a thoracic meningocele and a lumbar meningomyelocele is described. Both lesions were successfully operated upon; hydrocephalus did not develop and there was no obvious neurolgical defect.

I acknowledge, with thanks, the permission of the Chief Medical Officer, Ministry of Health, Uganda, to publish this paper.

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IAN C. BAILEY

The Neurosurgical Unit, Mulago Hospital, Kampala, Uganda, East Africa.

Classical and Mild Phenylketonuria in a Family

A few patients with untreated phenylketonuria (PKU) escape the severe mental retardation usual in the untreated disease. Some of these patients have classical PKU but most have a related condition which has been called mild PKU or hyperphenylalaninaemia. Hsia, O'Flynn, and Berman (1968) reviewed 43 untreated phenylketonurics who were 'atypical' in that the IQ was 70 or higher. Of these patients, 18 had plasma phenylalanine levels of 21 mg/100 ml or more and were regarded as having classical PKU, while 19 had plasma phenylalanine levels of 20 mg/100 ml or less and were regarded as having hyperphenylalaninaemia; in 6 cases the plasma phenylalanine level was not known.

We describe here a family of 5 children, 4 of whom have PKU. The youngest child has classical PKU diagnosed in the neonatal period and is on a low phenylalanine diet. 3 of her 4 older sibs have untreated mild PKU each with a blood