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Reference


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Two spine bifida defects in the same child

Sir: Congenital malformations of the central nervous system have been shown not to be uncommon in black people and reports from Nigeria show that hydrocephalus, anencephaly and myelomeningocele are the common anomalies seen. However, it is very rare for the same patient to have more than one defect of the spine; there are reports in the literature of such cases and the existence of lesions at three separate levels are extremely rare. The first case of a double spine bifida lesion in the same patient in Nigeria is presented in this report.

A 3 month old male child was admitted to hospital with a history that two swellings in the midline of the back had been noticed at birth. He was the third child of his mother. Pregnancy was uneventful throughout and was supervised at a private clinic in Bauchi State where she lived. Delivery was vaginal at term but was at home. The child cried spontaneously and passed meconium after birth.

Examination on 26 January 1988 showed a well nourished baby boy with a head circumference of 39 cm who weighed 5·1 kg. He was breathing spontaneously but had an intermittent stridor which had developed 18 days after birth. The anterior fontanelle was patent, flat and not tense. Two midline cystic

![Fig Thoracic myelomeningocele and lumbo-sacral lipomyelomeningocele.](http://jnnp.bmj.com/)

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swellings were noted in the thoracolumbar and lumbosacral regions. The upper swelling was covered by scar tissue and measured 5 cm x 3 cm. The lower swelling was covered by normal skin and measured 8 cm by 5 cm (fig). A sacral dermal sinus was seen below the lower swelling. Movements in both lower limbs were spontaneous but weak and the right lower limb moved better than the left. Anal sphincter tone was good. No other abnormality was noted. Radiographs of the chest and skull showed no abnormality. Thoracolumbar spine radiographs showed involvement of thoracic vertebrae T8-T11, and L5-S1, in the lumbosacral defects.

On 5 February 1988 at operation a ruptured thoracic myelomeningocele and a lumbosacral lipomyelomeningocele were found and repaired. The dermal sinus was not continuous with the spinal canal. 3.5 gm of fat was excised from the lipomyelomeningocele. The postoperative period was complicated by cerebrospinal fluid (CSF) leakage into the distal lesion which later broke down and required repeated aspiration and secondary wound closure. Movements in both lower limbs have not improved.

Very few cases of more than one lesion have been observed in the same patient. Tryfonas,2 reported three spinal bifida lesions in the same patient. The patient presented in this report had a thoracic myelomeningocele and lumbosacral lipomyelomeningocele (fig). A sacral dermal sinus was also present but it did not extend into the conus medularis. Johnson3 in 1857 was the first to describe a lipomyelomeningocele as a subcutaneous lipoma usually in the lumbosacral region. The lipoma usually passes through the defect in the muscles, vertebrae and dura into a low-lying sinal cord contiguous with the fat, which may be large or finger like. According to Schut,4 who reported an incidence of 25% lipomyelomeningocele to myelomeningocele, lipomyelomeningocele far outstrip other dysraphic anomalies like diastematomyelia, dermal sinus and thickened filum terminale.

Patients born with a lipomyelomeningocele are typically neurologically intact and present with a developing neurological deficit with increasing age. Many have a significant neurological deficit, by the time they attain the age of 4 years. Surgical repair not only aims at rectifying the cosmetic appearance but also at preventing neurological deterioration. The two spina bifida defects seen in this patient could each account for the decreased muscle power in both lower limbs. The good anal sphincteric tone without a patulous anus was noted. There was no talipes equinovarus deformity of the foot. The patient did not develop hydrocephalus post operatively and has not shown neurological deterioration during outpatient follow-up.

References
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Meningioma—an unrecognised complication of fibrous dysplasia of the skull?

Sir: Fibrous dysplasia is a congenital, non-familial, benign anomaly of bone development occurring in single or several bones, characterised by the replacement of normal bone by fibro-osseous tissue,1 with normal serum calcium, phosphate and alkaline phosphatase levels. The condition may be associated with skin pigmentation and endocrine abnormalities (Albright's syndrome).2 Approximately one-third of patients with fibrous dysplasia have involvement of the cranial or facial bones,3 complications of which include facial pain, headache and cranial nerve palsies due to bony compression, optic nerve damage being the most serious.

Although rare, malignant change occurring within dysplastic bone is well recognised. Various types of sarcoma have been reported including osteosarcoma, fibrosarcoma, chondrosarcoma and neurosarcoma.4 The present case report describes the development of a meningioma in association with fibrous dysplasia of the skull.

A 50 year old right handed Pakistani railway storeman was first seen in 1965 at the age of 28 years, with a long history of swelling and recent onset of double vision looked to the right. The diagnosis of fibrous dysplasia was confirmed on skull radiographs. In 1966 he had two operations in the first excess bone was removed from the region of the left orbit with some improvement in his visual symptoms, but when these recurred a large quantity of dysplastic bone was removed from the skull vault, leaving a full thickness bone defect which was later filled with an acrylic prosthesis. He did not receive radiotherapy and appears to have been lost to follow up.

The patient presented again in 1987 complaining that for the past 2 years there had been skull enlargement in the region of the left forehead and vertex, as well as episodes of visual blurring in the left eye, but no change in the longstanding diplopia. On examination there was obvious left frontal bossing. Although the visual acuities were normal, the visual fields revealed bilateral constriction and the fundi showed early papilloedema. The left eye movements were limited with restricted elevation and abduction. There were no other abnormal findings.

Routine biochemical indices were unremarkable, in particular serum calcium, phosphate and alkaline phosphatase levels were normal. A skull radiograph showed a large bone defect from his previous craniectomy. The frontal bones were thickened, particularly on the left, the left anterior clinoid was expanded and sclerotic with the appearances of fibrous dysplasia. CT of the head showed extensive fibrous dysplasia involving the frontal, parietal and sphenoidal regions.

Fig CT head scan showing contrast enhancing mass beneath dysplastic bone.