DIASEMATOMYELIA – THE “DOUBLE-BARRELLED” SPINE

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A perfectly preserved bony specimen of diastematomyelia approximately 1800 years old is presented. This abnormality is discussed, together with a brief review of the literature.

Diastematomyelia is an uncommon condition seen only rarely by general orthopaedic surgeons. For this reason it may be overlooked. We present a specimen of a “double-barrelled” spine (Fig. 1) which may serve to remind surgeons of its occurrence when considering a patient with back and lower extremity complaints.

Although the condition has been discussed extensively in the past (Matson et al. 1950; James and Lassman 1960; Winter et al. 1974; Hood et al. 1980), to the best of our knowledge no actual bony specimen has previously been presented in the orthopaedic or anatomical literature.

MATERIAL

The specimen is from the spine of an individual approximately 20 years old and was recovered during an excavation at the site of a burial tomb in the Negev desert of Israel. The tomb dates from the Roman period circa AD 100. Although the specimen is approximately 1800 years old, it had been immaculately preserved by the desert climate. The disarticulated spine consisted of 18 isolated vertebrae comprising most of the cervical, thoracic and lumbar segments; the sacrum and coccyx were missing.

When articulated the spine showed multiple malformations. Most remarkable was a sagittal bony septum at T12–L1 dividing the vertebral canal in two. The septum arose anteriorly from the back of a butterfly vertebral segment representing a failure of anterior body formation (Moe et al. 1978) at T12 (Fig. 2). It consisted of a plate of bone approximately 2 mm thick and 14 mm in its anterior–posterior dimension; it was trapezoidal,

2 cm long at its anterior base and narrowing to a summit of 1 cm where it attached to an undifferentiated mass of posterior elements (Fig. 3). It extended downward to include most of the subjacent L1 segment.

When compared with the spine above and below, the butterfly segment and the superior portion of the L1 vertebrae at the level of the bony septum were widened, producing a fusiform effect. The vertebral canal within was also clearly wider in the area of the septum than it was above or below, with a measured increase in interpedicular distance of 4 mm over the adjacent L2 level. This is the reverse of the normal pattern of increasing interpedicular distance as the lumbar spine progresses caudally. When viewed from above or below, the divided spinal canal had a “double-barrelled shotgun” appearance (Fig. 1).

Another failure of anterior vertebral formation in the butterfly pattern was seen at T9, although this defect was not associated with any septum. A severe congenital scoliosis derived from the two butterfly defects and extended into most of the thoracic spine and into the upper portion of the lumbar spine.

Additional malformations included multiple levels and degrees of spina bifida alternating with plate-like areas of fusion in the posterior elements (Fig. 3). There was marked disarray of the articular processes in the lower thoracic and upper lumbar spine with asymmetries, atrophy and, in some cases, complete lack of the articular surface.

DISCUSSION

The term “diastematomyelia” refers to the splitting of the spinal cord or cauda equina in the sagittal plane and not to the septum itself. However, it is almost always found in association with an osseous or fibrocartilaginous spur which divides the spinal canal. The septum is most commonly found at the lower thoracic or upper lumbar level, as in our case, but has been seen anywhere from the sacrum to T3 (Keim and Greene 1973; Winter et al. 1974). The division in the cord itself is usually longer than the septum, averaging from 5 cm (Keim and Greene 1973) to 10 cm (Herren and Edwards 1940) in

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Fig. 1

Fig. 2

Fig. 3

Figure 1 - The double-barrelled defect at L1 viewed from below with arrows indicating the bony septum: note the interpedicular widening at the level of the malformation. Figure 2 - The arrows indicate a butterfly defect in anterior body formation at T12. The bony septum arose posteriorly at this level and extended down to L1. Figure 3 - The view from behind shows marked disarray of the posterior elements. The arrow indicates the area of attachment of the bony septum. Note the spina bifida defects above and below this level.

length and with the spur most frequently found at the caudal portion of the split.

The embryogenesis and genetics of the disorder are subjects of conjecture (Bremer 1952; Gardner and Collis 1960) but it is agreed that the most common presenting feature is a congenital scoliosis. Diastematomyelia has been estimated to exist in 5% of all congenitally scoliotic patients (Moe et al. 1978). The lesion is associated with an overlying skin anomaly, such as a dimple or hairy patch, in 50% to 70% of patients (Eid, Hochberg and Saunders 1979). The classic association with a cavus foot appears to be less common than a club foot deformity (James and Lassman 1972). Foot disorders of some variety are present in 30% of all patients (Winter et al. 1974). The most common neurological signs are weakness of the legs, abnormal gait, or urinary incontinence (Banta 1981). Additional features vary from mild back pain to paraplegia (Keim and Greene 1973).

Radiographically, diastematomyelia is almost always associated with other congenital spinal defects; these usually are major failures of formation, as in our case, but may be as slight as a spina bifida occulta. The telltale bony spur is often difficult to see on plain radiographs, either because it is largely fibrocartilaginous, or because of overlying pathology, particularly posterior element abnormalities. Diastematomyelia may be suspected when increased interpedicular distance is seen in association with a localised fusiform widening of the spine (Hood et al. 1980). Myelographic studies should be instituted if the clinical situation warrants. The myelogram, a CT scan (James and Oliff 1977) or,

presumably, magnetic resonance imaging will usually demonstrate the division in the spinal cord and prove useful in diagnosing this uncommon abnormality.

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