Intramedullary dermoid

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Dermoids belong to the group of congenital cystic tumours developing from an embryonic rest. Such tumours vary from those which contain only epidermal cells to those containing various dermal derivatives, such as glandular cells (sebaceous or suderiferous), and even hair and teeth. So they vary from pure epidermoids to dermoids and then through the teratoid tumours to the teratomas.

These tumours can arise wherever two ectodermal surfaces fuse together in the developing embryo when an extodermal implant is retained deep in the surface. This may be the case at about the fourth week of intrauterine life when the neural ectoderm separates from the cutaneous ectoderm along the mid-dorsal aspect of the embryo (Ross Fleming and Botterell, 1959). As a result of such a midline inclusion an epidermoid or dermoid may develop between the skin and the brain ventricles or the central canal of the spinal cord. A congenital dermal sinus of varying length and completeness may persist in association with these tumours.

Dermoids and epidermoids are rare lesions. Within the skull they represent less than 1% of all brain tumours (Ross Fleming and Botterell, 1959). In the spinal canal they are far less frequently encountered. Sachs and Horrax, for example, up to 1949 could find only 61 cases in the literature representing 35 dermoids and 26 epidermoids, and Gros, Vlahovitch, and Mohasseb (1960) added two cases of intraspinal dermoids. Manno, Uihlein, and Kernohan (1962) published a survey of the literature of intraspinal epidermoids only. According to Sachs and Horrax (1949), dermoids are more often situated in the thoracic region of the spinal canal, whereas epidermoids are roughly equally distributed in the thoracic and lumbar region. In all four cases of intraspinal dermoid reported by Slooff, Kernohan, and MacCarty (1964) the lesions were situated in the conus terminalis. In general the site of these tumours may be extradural as well as extra- or intramedullary. They are characterized by an extremely slow expansion, thus allowing time for compensatory measures. The clinical course is protracted with long remissions so that at the time the correct

diagnosis is established most of these cases are already hopeless and the tumours inoperable (Naffziger and Jones, 1935).

Radiological findings in intraspinal dermoids and epidermoids consist mostly in a more or less conspicuous excavation of the bony spinal canal. Calcified deposits are usually absent or minute, but massive calcific shadows are occasionally seen in the widened spinal canal, for example, in the case reported by Moore and Walker (1951).

The following case is presented on account of the exceptional clinical and radiological findings.

T.J., a 62-year-old man, without anything remarkable in his family history, was first seen at the Neurological Clinic in 1958. Fifteen years before he had begun to suffer from slight weakness and numbness of the left leg together with mild pains in the joints of the upper and lower extremities. About six weeks before admission increasing weakness and clumsiness of both legs appeared, more pronounced on the left, with painful spasms in the left calf.

There was normal mobility and no tenderness of the spine. There was hyporeflexia C5-8 bilaterally, the abdominal and cremasteric reflexes were absent, and the knee and ankle jerks were hyperactive. There was moderate spasticity of the lower extremities, more on the left, and Babinski's sign was positive bilaterally. There was slight tactile hypaesthesia in the lower lumbar and sacral dermatomes, otherwise sensation was undisturbed.

Lumbar puncture disclosed an incomplete subarachnoid block and slightly increased protein content in the spinal fluid; the colloidal gold curve was 01113310.

Radiographs of the axial skeleton disclosed basilar impression and atlas assimilation; the tip of the dens was situated as high as 20 mm. above Chamberlain's line. Otherwise only osteochondrosis at C5-6 and some spondylotic lipping were encountered. Gas myelography of the cervical portion performed by lumbar insufflation showed normal findings on the intraspinal structures. The passage of air through the spinal canal was free.

There was distinct objective and subjective improvement in the neurological deficit under routine symptomatic therapy so that the patient refused the proposed operation of the basilar impression.

The patient appeared again six years later in a much worsened condition which was induced by a fall two years

FIG. 1.

FIG. 2.

FIG. 3a.

FIG. 3b.

FIG. 1. Conical intraspinal shadow of calcific density at the level of D11.

FIG. 2. Tomogram showing no gas in the thoracic portion of the spinal canal. Dorsal aspects of the vertebral bodies D8-L1 are flattened and even slightly excavated. The calcified shell at D11 is more clearly visible.

FIG. 3. Operative specimen and radiograph of the specimen showing granular calcifications.

after the previous period in hospital, and the disease had progressed steadily since that time. Walking was now scarcely possible even with two crutches. Two years previously there appeared urgency of micturition and loss of libido as well as some sensory loss and frequent tonic spasms of both lower extremities.

There were no neurological abnormalities referable to the cranial nerves and upper extremities except hyporeflexia at C5-8 bilaterally. The lower limbs were weak and his gait was severely spastic. The knee and ankle jerks were very brisk with bilateral Babinski's sign. Position and vibratory appreciation were defective in the lower extremities. Perception of pinprick and touch was absent below the level of D11 segment. There were trophic changes on the lower extremities and over the sacral area.

Laboratory studies, including determinations of the haemoglobin, erythrocyte sedimentation rate, and leucocyte count, gave normal results as did urine analysis. The Queckenstedt test gave evidence of a complete block of the spinal subarachnoid space: the response to abdominal pressure was prompt. The spinal fluid was xanthochromic, coagulated spontaneously, and contained a marked increase in protein content.

Radiographs of the dorsal spine showed a conical calcified shadow at the level of D11 (Fig. 1), suggesting at first sight a mere summation effect from the pleura or the lungs. In tomograms, however, performed during gas myelography (Fig. 2) the intraspinal position of the shadow looking like a calcified shell could be clearly seen. The air insufflated by the lumbar route did not penetrate into the thoracic portion of the spinal canal. In the antero-posterior projection the calcific deposits were faintly visible. The medial aspects of the pedicles D10 were flattened and slightly divergent distally so that the interpediculate space was widened. The dorsal aspects of the vertebral bodies D8-11 were flattened and even slightly excavated. In a lateral tomogram through the level of the left-sided pedicles abnormal reduction in size of the intervertebral foramen D8-9 was noted.

On the basis of the clinical findings, the excavation of the spinal canal, together with intraspinal calcification, anomalous shape of an intervertebral foramen and basilar impression, the diagnosis of a developmental intraspinal lesion in the lower thoracic region was suggested.

A laminectomy was performed, exposing the tense dura from the eighth to the twelfth thoracic vertebrae. After the dura was incised a blue-brownish fusiform widening

of the cord was found. At first it seemed to be caused by an intramedullary gliomatous tumour. After medullectomy, however, yellowish-white cheesy material containing many long and short hairs protruded from the wound. The cord was reduced to a thin envelope associated intimately with the dermoid. The entire content of the dermoid was removed, together with a part of its capsule (Fig. 3a and b), the complete removal of which was impracticable. In the lower part of the huge cavity left after removal of the dermoid there were firm calcified deposits involving also the dural sac. The pathological report confirmed the diagnosis of dermoid.

The post-operative course was stormy. The neurological deficit increased, there appeared paraplegia, trophic ulcers, thrombophlebitis of the left lower extremity, and chronic urinary infection. The patient died at home five months after the operation. Necropsy was not performed.

This case appears interesting on account of the less common site of the lesion, the instructive clinical course, and, especially, in the much less conspicuous radiological picture than that known from the literature.

SUMMARY

A case of intramedullary dermoid in a 62-year-old man with interesting clinical and radiological findings is reported.

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