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AN INTRADURAL EPIDERMOID CYST IN THE LUMBAR REGION

Case Report and Review of Current Aetiological Theories

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Dumèril (1807) was the first to report an epidermoid tumour, but Cruveihier (1829) published a more comprehensive account and coined the term "tumeur perlee" to describe the gross appearance of this neoplasm. Epidermoid tumours of the central nervous system are rare and represent less than one per cent of all brain tumours (Fleming & Botterell 1959), but their intraspinal appearance is even less common: in forty-four patients with epidermoid tumours involving the central nervous system, MacCarty et al. (1959) found only three in the spinal cord. After searching the literature thoroughly, Manno et al. (1962) found and classified only ninety epidermoid tumours of the spine. More recently Reeves (1967) and others have reported isolated examples.

CASE REPORT

In October 1971 a woman aged forty-seven years complained of low back pain of two years duration. The pain began suddenly when lifting a heavy weight and affected the lower lumbar region on both sides, radiating down the posterior and lateral aspects of the right thigh. Coughing, sneezing and straining aggravated the pain, that was continuous in nature. Before attending our clinic the patient had been given numerous forms of treatment: bed rest, daily spinal traction for two weeks, back exercises and two caudal blocks. These were of no benefit to her. On the contrary, the caudal blocks that were performed in May and August 1971 aggravated her symptoms.

Examination

Examination of the vertebral column revealed sacrospinalis spasm and limited spinal flexion in the lumbar region: the lumbar lordosis was not obliterated and

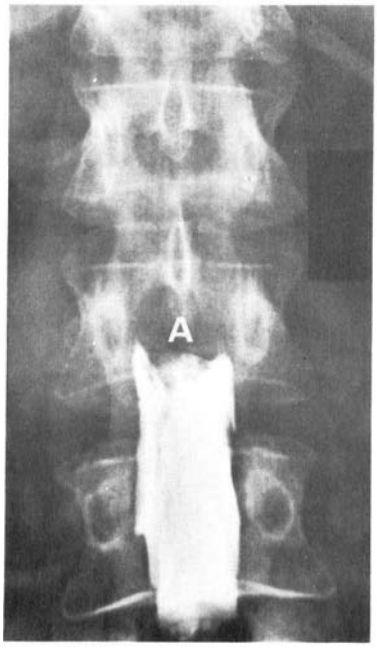

Figure 1. A lumbar myelogram showing a block to the flow of myodil of site A opposite L 2.

the finger tips reached only to within ten inches of the floor. A postural scoliosis convex to the left became more apparent. Spinal extension was full. Lateral flexion and lateral rotation were diminished on movement to the right. O'Connell's femoral stretch sign was present on the right side and there was some impairment of sensibility to pin prick on the lateral aspect of the right foot. Straight leg raising on the left and right sides was 70° and Lasegue's sciatic stretch sign was absent. There was weakness of dorsiflexion and eversion in the right ankle and foot. All reflexes were normal and there was a flexor plantar response.

Investigations

Further investigations revealed a normal full blood count, ESR and electrolytes. Radiographs of the lumbo-sacral spine were normal. A lumbar puncture revealed a pressure of 60 mm of CSF recorded with free rise and fall on jugular vein compression. Microscopy of the CSF demonstrated less than one polymorph per mm^3, less than one lymphocyte per mm^3, and five RBC's per mm^3. No micro-organisms could be demonstrated on Gram stain and culture was sterile. CSF protein was 345 mg per 100 ml, and a Lange curve of 0011000000 was recorded. Glucose and chloride levels in the CSF were not determined. A lumbar myelogram showed a complete block to the cranial flow of myodil in both prone and supine positions at the level of the first and second lumbar vertebrae (Figure 1). A provisional diagnosis of an intradural space occupying lesion was made.

Operation

At operation an intradural tumour, intimately entangled with the nerve roots of the cauda equina was carefully dissected and removed intact. The tumour was located opposite the first and second lumbar vertebrae and was oval in shape, measuring approximately 1 cm by 2 cm.

Progress

Post-operatively the patient made an uneventful recovery and was discharged on the fourteenth post-operative day. At routine out-patient follow-up at one month and two months, the patient was well and without symptoms.

Histology

Histological examination of the specimen revealed fragments of epidermis covered with numerous keratin squames with no skin appendages; a pathological diagnosis of Epidermoid Cyst was made. Bacteriological culture was sterile.

AETIOLOGY OF INTRASPINAL EPIDERMOID TUMOURS

It is stated by Reeves (1967) that epidermoid, dermoid, teratoid and teratomatous tumours arise from ectopic cell rests. The cell rests may be deposited in an ectopic position by some defect in embryo formation and the tumours which develop are considered to be congenital. Alternatively, the cell rests may be introduced into an intraspinal position by surgical intervention and the resultant tumours are then termed iatrogenic in origin.

The ultimate nature of the congenital tumours appears to depend upon the state of development and nature of the cell rests; ectopic implantation in the embryo of primitive undifferentiated cells derived from all three germ layers may lead to teratoma formation. Similarly, ectopic cells from two germ layers may produce teratoid tumours (Reeves 1967). It is currently accepted that congenital epidermoid tumours are formed by the investment of cutaneous epithelium during the closure of the medullary folds in the third to fifth week of embryonic life (Bostroem 1897).

However, in the case of teratomatous cysts Rewcastle & Francoeur (1964) dispute the aetiology of simple misplacement of normally developing cells and state that these neoplasms are a result of teratomatous development. This suggestion is well substantiated by a demonstration of female sex chromatin in teratomatous cysts removed from male patients. As yet, similar findings in epidermoid cysts have not been reported. Holmdahl (1933) considers the caudal end of the cord to develop from a solid mass of germinal cells of no specific embryolog-

ical layer and gives this as a reason for the preponderance of epidermoid, dermoid, teratoid and teratomatous tumours in the lumbosacral region. A combination of the Holmdahl (1933) and the Rewcastle & Francoeur (1964) theories, that is, teratogenesis within a mass of caudally placed germinal cells, would appear to be a reasonable hypothesis to explain the origin, in congenital terms, of each of these tumours, but experimental proof would be difficult to obtain.

Both clinical and experimental exidence for the production of iatrogenic tumours is vast and now unquestioned.

Van Gilder & Schwarz (1967) have shown convincingly that central nervous system dermoids and epidermoids can be produced by skin implantation along the neuroaxis in albino rats and similar experiments by Oblu et al. (1967) have led to the same conclusion.

Blockley & Schorstein (1961), Choremis et al. (1956), Oeconomos & Caracolos (1957), De Rougemont et al. (1962) each have described young children with subarachnoid extramedullary epidermoid cysts, sometimes multiple, of the lumbar region and in all cases the children have received multiple (three to many) lumbar punctures during the treatment of meningitis. Because the cysts were approximately at the sites of the lumbar punctures and because of their subarachnoid position and multiplicity, it is strongly suggestive that they arose from viable fragments of epidermis carried in by the lumbar puncture needles. In this connection it is interesting to note that Gibson & Norris (1958) showed that in sixty per cent of skin punctures, a small piece of skin becomes detached and remains in the needle. Since lumbar puncture needles carry a stilette, the possibility of introducing a fragment of skin into the subarachnoid space must be high.

The growth rate of intraspinal epidermoid tumours is variable depending upon the aetiology. Simple congenital epidermoid tumours, not associated with dermal sinuses, grow slowly, as indicated by the twenty to twenty-five year average latent period, before the onset of symptoms (List 1941). Gross (1934) in twenty patients with simple intraspinal epidermoid and dermoid tumours found the duration of symptoms to be ten years and the average age at operation thirty to forty years. Congenital epidermoid tumours in association with dermal sinuses have a shorter latency (List 1941) indicating a more rapid rate of growth. In those patients reported by Manno et al. (1962) with both congenital epidermoid tumours, and dermal sinuses, the average age at operation was five years. Where these two defects are apparent in the same patient, the embryological malformation is likely to be of a

marked degree and the initial cell rest may be of considerable size. Early diagnosis, prompted by the existence of a dermal sinus, plus an initially large epithelial cell rest may give a misleading impression of a more rapid growth rate. Epidermoid cysts of iatrogenic origin, produced by implantation, have been shown by Manno et al. (1962) to have a relatively short latency and a mean period of 6.9 years between the initial surgical procedure and operative removal of a tumour.

DISCUSSION

The aetiology of the epidermoid tumour presented in this case report is undoubtedly congenital. Although two caudal blocks, performed twelve months before the operation, may have been responsible for the introduction of skin fragments, albeit at a lower level, these could not have attained, according to the statistics quoted by Manno et al. (1962), the size of the tumour removed at operation. It is interesting to note that this congenital epidermoid tumour was unaccompanied by associated anomalies such as spina bifida or dermal sinus.

In a small series of patients with intraspinal epidermoid tumour collected by List (1941) a male predominance was noticed but the larger series of Manno et al. (1962) showed sex incidence to be equal. By adding those patients who have subsequently appeared in Western literature to those figures quoted by Manno et al. (1962) the distribution becomes forty-six females to fifty-three males, including the case presented in this communication.

Gross (1934) in a series of twenty intraspinal epidermoid tumours found the average duration of symptoms to be about ten years and Reeves (1967), although stating no precise period, suggests that the prolonged and slow progression of symptoms is an important guide in the pre-operative diagnosis of the tumour. The patient presented here showed an unusually short history of only two years. The relatively early diagnosis was made possible by recognition of a small but significant group of clinical signs and confirmation was secured by prompt myelographic investigation. The straight X-ray of the lumbo-sacral spine in iatrogenic and simple congenital epidermoid cysts (in the absence of spina bifida occulta) appears to be of limited value for making an early diagnosis in that direct pressure producing sufficient vertebral erosion to appear on a plain X-ray could be maintained only by a large epidermoid tumour. The X-ray changes produced by spinal tumours are well documented by Elsberg & Dyke (1934) and Hannan

& Giest (1950) have described particularly those changes produced by spinal teratomatous tumours, stating that only fifty per cent of proven cases show plain X-ray changes. Calcified deposits are generally minimal or absent but a case reported by Moore & Walker (1951) showed massive intraspinal calcific shadows. In the patient reported here, myelographic studies undoubtedly led to an early diagnosis. The outcome is a testimony to the value of myelography in the presence of progressive symptoms and signs associated with a space occupying lesion in the spinal canal.

At operation, total cyst excision was technically possible and preferred to cyst evacuation. The latter method allows affected nerves to be decompressed, but Hullay et al. (1962) have shown it to be followed by an early recurrence rate. Although dissection of the tumour from the cauda equina proceeded without difficulty in this patient, it is important to note that Van Gilder (1967), using animals, has shown the pericystic collagen tissue to be locally invasive: subarachnoid implants have been demonstrated to invade nerve roots. However, a long remission may occur even with incompletely removed tumours (MacCarty et al. 1959), presumably because of their slow growth rate.

The removal of the intact cyst in the patient reported in this communication was probably a beneficial factor to her post-operative recovery. Verbiest (1939) postulated that cyst spillage can result in a meningeal reaction and arachnoiditis. Similarly, Van Gilder (1967) noted, on examining one of his animals sacrificed because of spasticity, that a cyst had ruptured and produced necrosis in the spinal cord nearby. Craig (1943) noted this phenomenon as a post-operative complication and thought it to be responsible for the poor operative results that he had obtained.

The present author is of the opinion that excision, when not complete, should be as wide as possible, but should not rupture the cyst or jeopardise apparently healthy nerve roots.

SUMMARY

An intradural epidermoid cyst in the lumbar region of a forty-seven year old woman is reported. The importance of myelographic examination in patients suffering gradual symptomatic deterioration coupled with progressive physical signs is emphasized. Current theories regarding the aetiology of epidermoid cysts are reviewed.

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