INTRASPINAL EPIDERMOIDS

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EPIDERMOID tumors involving the spinal cord and its roots have long aroused the curiosity of physicians encountering them in clinical practice. These tumors occur infrequently in the central nervous system; yet the older literature indicates considerable speculation regarding their pathogenesis and their relationship to two types of allied tumors, the dermoids and the teratomas, with which they commonly have been confused. We also have found this confusion to exist in cases reported as recently as 1958. For this reason, we wish to describe briefly the differences between these tumors.

DIFFERENCES BETWEEN EPIDERMOIDS, DERMOIDS AND TERATOMAS

Histologic Appearance. An epidermoid is a mass of desquamated cells containing kerato-hyalin, encased within a capsule of well-differentiated stratified squamous epithelium. The earlier descriptions concerned chiefly the gross appearance; nothing was mentioned about specificity of cells.

Dermoids, although similar to epidermoids in their epithelial structure, contain hair, sebaceous glands, and other appendages of skin.

Teratomas represent heterotopic elements from all three germ layers, and can be distinguished from dermoids by the presence of cartilage, muscle, and other mesodermal tissues.

Pathogenesis and Historical Data. Various investigators differed regarding the etiology of epidermoids; and this probably led to the early confusion associated with the nomenclature. The first name, merely a descriptive term, was Cruveilhier’s “tumeurs perlées.” Not until 1838 was the tumor investigated more thoroughly by Müller. He undertook its histologic study and described the laminated structure, absence of blood vessels, and epithelial capsule made up of polygonal cells without nuclei. Considering cholesterol as a constituent of this tumor, he likened it to similar masses in the chronically inflamed middle ear, and named it “cholesteatoma.” Virchow did not consider cholesterol essential to this tumor and retained the name “Perlgeschwulst” (pearl tumor).

Although von Remak was the first to report on misplaced epithelial tissue giving rise to epidermoid tumors, most credit is given to Bostroem for his investigations on the pathogenesis of epidermoids, dermoids, and dermal sinuses. He furnished the concept and now accepted theory of displacement of ectodermal cells during embryogenesis of the central nervous system. These cells, that is, the cutaneous portion resulting from the differentiation of primitive ectoderm into skin and neural ectoderm, inadvertently participate in closure of the neural groove and become trapped within or near the newly formed neural tube, thereby giving origin to epidermoid and dermoid tumors. Should this phenomenon occur early in life, a dermoid will result, since the tumor will contain completely potential skin-forming cells; if it occurs later, the cells already will have become differentially unipotent and capable of producing only the structure-limited epidermoid.

Holmdahl, in experimenting on the embryogenesis of the vertebrates, distinguished between what he termed “primary and secondary body development.” He showed that during the primary phase, the neuraxis develops in the orthodox manner. However, during the secondary phase, an
undifferentiated mass of cells, caudal to primitive streak, forms the lower thoracic and entire lumbosacral portions of the spinal cord. These cells, which do not belong to any single germ layer, form a solid cord (the anlage of the medullary tube) apart from the superficial epithelium; this becomes canalized in a closed fashion and forms the neural canal. In conclusion Holmdahl wrote that one should expect that in this region the epidermoids develop either primarily inside or outside the neural canal, stressing that an inclusion from the overlying superficial (cutaneous) epithelium cannot take place.

List suggested that since this mass of cells is totipotential, the finding of epidermoids, dermoids, or teratomas in this region is not incompatible with Holmdahl’s propositions. “By disorderly differentiation of the primitive matrix,” List continued, “any of the three tumors can be formed.” That these tumors can arise from without seems most unlikely, since they contain residues of more than one germ-cell layer. However, an epidermoid occurring in this manner is not without some credulence. The method relating to this possibility is mentioned later.

Incidence. As mentioned before, epidermoid tumors rarely occur in the central nervous system. Their incidence among intracranial tumors in most large series is variously estimated at 0.2 to 1 per cent; in the spinal canal the incidence is even less. In a recent collection of 44 epidermoids of the central nervous system identified in patients 15 years of age or older, before December, 1955, Maccarty and his associates of the Mayo Clinic found only 3 to be intraspinal.

REPORT OF CASES

The following 2 cases, in which epidermoid tumors involved the cauda equina, represent new additions to the complete list of intraspinal epidermoids reported in the available literature on this subject. In the series, these 2 reports occur as Cases 37 and 45.

Case 37. A 46-year-old traveling salesman was seen at the Mayo Clinic on March 21, 1960, complaining of pain low in the back and in the left leg and left thigh of 3 years’ duration, intensified by coughing and sneezing, and accompanied by paresthesia over the left thigh of 4 years’ duration. He had had a brief bout of pain in the sacral region after a spinal anesthetic for appendectomy in 1957.

General neurological examinations gave normal results, save for minimal limitation to straight-leg raising on the left.

Laboratory studies, including determinations of hemoglobin, erythrocyte sedimentation rate, and leukocyte count, gave normal results as did urinalysis and the Kline flocculation test. Studies of the cerebrospinal fluid showed 85 mg. of protein and 7 lymphocytes per 100 cc.

Pantopaque myelography disclosed a well-circumscribed defect in the upper posterior portion of the 4th lumbar vertebra.

On March 23, 1960, bilateral laminectomy of the 4th and 5th lumbar vertebrae was performed by one of us (A. U.), and a pearly mass was found attached to the rootlets of the cauda equina on the left. The mass was removed, carefully, without any sacrifice of nerve roots. However, it was necessary to strip the arachnoid away from the rootlets of the cauda equina because this membrane had become thickened and had caused these nerves to be matted together. Histologically the mass proved to be a typical epidermoid cyst.

After operation the patient’s symptoms disappeared and he was dismissed on the 11th postoperative day.

Case 45. A 2½-year-old boy was seen at the Mayo Clinic on Feb. 3, 1955, because of a draining sinus in his back and weakness of the right leg, presumed to be a residuum of an attack of purulent meningitis some 3 months before his registration.

He had appeared normal at birth except for the lumbosacral dermal sinus, from which clear fluid had drained when he was 3 months of age. Two bouts of purulent meningitis followed; both times the patient responded to antibiotic therapy. After the second attack, weakness of the right leg was apparent. Physical therapy and exercise of muscles were unsuccessful. Further investigation was deemed necessary and the parents were referred to the Mayo Clinic.

On physical examination a lumbosacral dermal sinus was noted, as well as weakness of the anterior tibial and peroneal muscles of the right leg. Slight atrophy of the muscles of thigh and calf also was evident. Laboratory determinations for hemoglobin and leukocytes gave normal values, as did urinalysis. Roentgenograms of the thorax, skull, and lumbar portion of the spinal column did not show evidence of abnormality.

On Feb. 10, 1955, the dermal sinus was excised and sacrolumbar laminectomy was performed. The sinus extended through a sacral spina bifida occulta and was continuous with a large epider-