Intramedullary Epidermoid Cyst Associated with a Dermal Sinus

Case Report

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An intramedullary epidermoid with a dermal sinus is an unusual association. We have been unable to find a previously reported coincidence of these malformations although there are many definitive papers on these uncommon entities.  

Crueilhier as early as 1835 coined the eponym, “Perless Tumeur,” while Muller suggested the term “cholesteatoma” a little later in the 19th Century. Seventy years ago Bostroem discussed the pathogenesis of epidermoids much as we understand it today, as an error of medullary fold closure during the third to fifth fetal week with inclusion of maturing epidermis deep to the surface. The earlier literature, in general agreement, is replete with flowery description such as “a dysrafism,” “disembrionic malformation,” and “pseudotumor of epithelial grafts.”

The pathogenesis of dermal sinuses is similar to that of epidermoids in that they represent a malformation in the lines of closure of the medullary folds. Holmdahl’s observations on the formation of the spinal cord suggested that the lower portion of the cord formed from a solid mass of germinal cells not belonging to a specific germ layer. He felt this unique formation of the caudal medullary tube was an explanation for the propensity of both dermal sinuses and dermoid tumors to develop in the lumbosacral area. Other authors in the earlier literature suggested that dermal sinuses arose from downgrowths of epithelium to the neuraxis. Neither of these views is as commonly accepted today as the medullary fold malformation theory. The position of the epidermoid in a spectrum of differentiation of cell rests and multipotential cells with dermoids and teratomas has been discussed by French and Peyton, Rewcastle and Francœur, Reeves, and others.

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Morphological Review

An epidermoid cyst is a mass of desquamated squamous cells encased in a collagenous capsule lined by well-differentiated stratified squamous epithelium. The dermoid cyst is similar but also contains hair and has sebaceous glands and other skin appendages in its wall. The contents of the epidermoid cyst is characteristically a soft homogenous white material thought to be a keratin breakdown product. The material in the dermoid is a buttery, yellow sebaceous secretion mixed with a variable number of hairs. Both tumors may have capsular calcification occasionally to the degree of x-ray visualization. Russell and Rubinstein consider these as separate entities despite their similarities and do not recommend assimilating them under the title of “pearly tumors.”

Dermal sinuses are a midline opening in the skin surface, lined occasionally by stratified squamous epithelium but more commonly by an inflammatory exudate extending to the dura and occasionally into the subarachnoid space. They may communicate with intraspinal epidermoids, dermoids, and teratomas. The common site for these sinuses is at the sacral or lumbosacral area, but they have been reported over the entire midline surface of the posterior trunk, neck, and scalp.

Case Report

This patient was a 48-year-old white woman with a progressive history of stress incontinence and right foot drop of 12 years’ duration. She had a recognized lumbosacral dermal sinus, and her grandmother and cousin had cervical and thoracic dermal sinuses, respectively. The patient had meningitis at the age of 35 years at which time she had a lumbar puncture for diagnosis. The spinal fluid had been described as opalescent, containing 850 lymphocytes per high power field and protein of 92 mg%. Spinal
fluid culture was sterile. She had been treated with antibiotics and recovered rapidly without apparent sequelae.

Examination. The patient had a right foot drop, saddle hypesthesis, anesthesia over the dorsum of the right foot and a non-draining, non-infected, hair-bearing, lumbosacral dermal sinus. Intravenous pyelogram and cystogram were interpreted as normal. Plain x-ray films of the spine showed a sacral spina bifida occulta. A cisternal myelogram revealed an intramedullary mass at the level of the second lumbar vertebra.

Operation. At laminectomy, an intramedullary epidermoid of the low-lying conus at the L-2, L-3 level was evacuated. The dermal sinus ended blindly in the dura at the L-4 level without communicating with the tumor. The patient did well postoperatively. Over the 4-month period since surgery, bladder function has returned to normal, and the right foot drop has shown slow but steady improvement.

Discussion

In reporting this case, we felt that the significant aspect was the association of a dermal sinus with an intramedullary epidermoid. We are well aware that there are a number of reported extramedullary epidermoids with associated sinuses; however, we have been unable to find a previously recorded intramedullary epidermoid with this association. Sachs and Horrax included one extramedullary epidermoid associated with a dermal sinus. The series of Manno, et al., contains six children with extramedullary epidermoids and dermal sinuses. Fager’s review of intramedullary epidermoids records no associated dermal sinuses.

Intramedullary epidermoids appear to be a condition seen primarily in early middle age; whereas the association of an epidermoid with a sinus is seen primarily in children.

Undoubtedly, these tumors are of two origins, congenital and iatrogenic. With the latter, it has been well established that implanted skin fragments, most commonly by lumbar puncture, may be a nidus for the development of a dermoid or epidermoid. Van Gilder and Schwartz implanted keratin-free minced fragments of skin in the cranial and spinal subarachnoid space by craniotomy or laminectomy in rats. Sixteen of the 18 animals developed tumors within 171 days. Similar work by Oblu obtained similar results. These tumors are commonly extramedullary and usually dermoids.

The significance of intrathecal drugs plus the presence of meningitis in tumor development has been debated. Blockey and Schorstein have pointed out that the important distinction between the congenital and acquired epidermoids is the presence or absence of associated spinal anomalies; there is a significant incidence of associated spinal malformations in the congenital series which is noticeably lacking in those cases thought to be secondary to implanted skin fragments. Our patient, however, in addition to the dermal sinus, had an occult sacral spina bifida. The low-lying conus is important in this regard as well.

Manno, et al., and Black and German have noted cord anomalies of syringomyelia and diastomatomyelia. We found two other reported intramedullary epidermoids, both of the thoracic area, with a past history of lumbar puncture; no doubt these were unrelated coincidences. But in our case with the tumor in a low-lying conus, the lumbar puncture could have been a factor. We suggest that in the presence of a suspected conus medullaris or cauda equina epidermoid that the myelography by cisternal tap is preferable both to avoid cerebrospinal fluid contamination with punctured cyst contents and to prevent conus injury.

This patient’s meningitis could have been of bacterial or chemical origin; the opalescent CSF is very suggestive of the latter. The work of Matson and Jerva plus that of Clifton and Rydell, Moïse, and Stammers has now established the importance of a dermal sinus as an avenue for meningeal contamination. But it was not too long ago that authors doubted the significance of a few drops of clear fluid from the dermal sinus or that the sinus could be related to meningitis. Kooistra has pointed out that the lower sinuses are usually complicated by sepsis, whereas the higher ones are more commonly associated with tumors.

Verbiest, in 1935, first suggested the importance of a meningeal reaction and chemical arachnoiditis secondary to cyst content spillage. He felt the significant materials in