Supratentorial dermoid cysts

PIERPAOLO LUNARDI, M.D., AND PAOLO MISSORI, M.D.

Department of Neurological Sciences, Neurosurgery, University "La Sapienza," Rome, Italy

Supratentorial dermoid cysts are rare lesions. In eight cases presented here, the lack of recurrence after subtotal removal of the capsule and the good long-term prognosis are emphasized. This finding is in agreement with the literature. The frequent relationship of these lesions with the cavernous sinus suggests a vascular genesis in the development of intracranial dermoid cysts.

KEY WORDS • cavernous sinus • cyst, dermoid • cyst, epidermoid

Intradural dermoid cysts represent one of the rarest forms of benign intracranial lesion, their frequency ranging from 0.04% to 0.6% of all intracranial tumors. Because of their common congenital origin they have frequently been confused with epidermoid cysts, although intradural dermoid cysts are four to nine times less common than epidermoid lesions. Many reports indicate that the midline below the tentorium is the preferential localization of intradural dermoid cysts, often associated with the occipital dermal sinus. More recent series show a higher frequency of supratentorial than of infratentorial dermoid cysts. The significant differences between dermoid and epidermoid cysts and between infratentorial and supratentorial dermoid cysts prompted us to review our series of supratentorial dermoid lesions. This report examines eight cases of these unusual and very interesting non-neoplastic lesions.

Summary of Cases

Patient Population

Eight patients with intracranial intradural dermoid cysts were surgically treated in our department between 1952 and 1989. They ranged in age from 4 to 48 years (mean 25.5 years) without a marked preponderance in any one decade. Five were female with a mean age of 18.4 years and three were male with a mean age of 37.3 years. Two patients presented suprasellar dermoid cysts and six had paramedian lesions: frontal and/or temporal.

Symptoms and Signs

The length of the clinical history was related to the lesion site: the mean duration was 3 months in suprasellar dermoid cysts and 12 years in frontotemporal lesions. In the majority of cases, epileptic seizures were the only symptom, occurring for many years; however, two children complained of visual disturbances and in one case chemical meningitis was the first symptom of the dermoid cyst. Neurological examination was entirely normal in six cases (75%), while reduced visual acuity with field defects was detected in two children.

Neuroradiological Investigations

The first two patients in the series were subjected to conventional plain x-ray films, pneumoencephalography, and angiography. The last six patients were easily evaluated by computerized tomography (CT) and/or magnetic resonance (MR) imaging.

Operative Features

All of the dermoid cysts were approached by a frontal or pterional route. After exposure and opening of the whitish capsule, removal of the semifluid yellowish substance was easy and allowed sharp dissection of fibrous bands that were loosely adherent between the capsule and neurovascular structures. A microsurgical technique facilitated the excision, but small portions of the capsule firmly adherent to vessels, nerves, or brain were left in situ in five cases; cerebrospinal fluid (CSF) filled the residual cavities.

Neuroradiographic Results

The conventional neuroradiological investigations showed only nonspecific signs of a space-occupying lesion. In the six patients in whom CT was performed, the scan displayed a uniformly hypodense roundish lesion (from ~20 to ~40 Hounsfield units (HU)), with capsular calcification in two. A slight mass effect without capsular enhancement or edema was detectable.
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Fig. 1. Computerized tomography scan showing a large roundish frontotemporal lesion (~40 HU) with slight mass effect. Fat droplets (arrow) are scattered throughout the subarachnoid spaces. The point of attachment with the cavernous sinus is not displayed in this axial plane.

(Fig. 1). Coronal and sagittal reconstruction performed in the last three patients clearly showed a point of attachment between the dermoid cyst and the cavernous sinus (Fig. 2). In four patients, fatty material was floating in the cisterns, probably due to spontaneous rupture of the cyst (Figs. 1 and 2 right).

In two patients, T1- and T2-weighted MR images confirmed the fatty nature of the lesion but did not reveal a tendency to spread along the subarachnoid spaces, which is typical of epidermoid cysts. In both patients, free fat globules appeared in the subarachnoid spaces (Fig. 3).

Pathological Findings

No abnormalities of the skin or bone defect were detected at surgery. The external surface of the cyst appeared roundish and lobulated with a pearly sheen. Foci of calcification were found in one case. The cyst wall usually varied in thickness, depending on the amount of epithelial and connective tissues incorporated in it. The typical dural, granular, fibrous, and cellular layers were represented in the ectodermal epithelial tissue. Connective tissue containing mesodermal elements, such as sweat or sebaceous glands, hair follicles, adipose cells, elastic fibers, and capillaries, was observed only in small areas (Fig. 4).

Contrary to epidermoid cysts, which are filled with waxy concentric lamellae of cholesterol crystals, the dermoid cysts contained yellowish-brown material resembling fat degenerated as a result of sebaceous gland secretion and progressive desquamation of the epithelium. Small hairs scattered throughout the fatty substance were detected in five cases.

Operative Outcome

There was no operative mortality. The postoperative

Fig. 2. Computerized tomography (CT) scans. Left: Coronal CT scan showing the point of attachment of the temporal dermoid cyst (~20 HU) to the cavernous sinus. Right: Axial CT scan showing the oblong temporal dermoid cyst originating close to the cavernous sinus. Fat globules (arrow) appear in the chiasmatic cistern.

Fig. 3. Coronal magnetic resonance image (TR 500 msec) delineating the hyperintense dermoid cyst projecting into the temporal lobe, with its attachment to the cavernous sinus.

Fig. 4. Photomicrograph of a typical dermoid cyst. Numerous sebaceous glands are present. Note the characteristic hair follicle (straight arrow) and thin layer of epithelium with stratum granulosum and accumulated keratin (curved arrow). H & E × 65.
course was complicated by mild chemical meningitis in the first three patients in the series. Systemic cortisone therapy ensured swift and uneventful recovery in the last five patients. One woman received a ventriculoperitoneal shunt 9 years after operation for nonresorptive hydrocephalus. With a mean follow-up period of 10.8 years (range 1 to 32 years), all of the patients are in good health without clinical or CT evidence of recurrence. All are independent and working full-time.

Discussion

Pathogenesis

Dermoid cysts are most frequently seen in the midline below the tentorium. At this site a tuck of skin may be retained when the dura mater invaginates to form the tentorium, causing the development of a dermoid cyst.\(^9\) The high incidence of associated dermal sinuses\(^2.3.7.8\) confirms the dysembryogenetic theory.

In all likelihood, supratentorial dermoid cysts arise due to misplacement of embryonic inclusions in the vicinity of the developing neural tube up to the 3rd week of life,\(^7\) during Carnegie stages 8 to 10, when the neural groove begins to close.\(^9,40\) Cranial abnormalities such as bone defects, dermal sinuses, or meningoencephaloceles are not associated with this development.

In our experience, intracranial intradural dermoid cysts appear near or adherent to large venous structures: the cavernous sinus for supratentorial dermoid cysts, and the torcular herophili for infratentorial dermoid cysts. Indeed, either dermal structures or venous channels derive from the mesoderm. This relationship leads us to suppose that these lesions originate from mesodermal nests dysembryogenetically adherent to primitive veins. It will be recalled that in an embryo of about 5 mm the venous drainage of the cranial region is supplied by three superficial pial plexuses surrounding the primary cerebral vessels.\(^20,24,41\) Its formation always precedes the intrinsic vascularization of any region.\(^32\) Endothelial pseudopods and pial vessels in the adjacent glia then perforate and penetrate into the nervous tissue, allowing the formation of new intraneural vessels. We think it reasonable to suppose that this vascular migration is responsible for the development of dermoid cysts through the entrapment of mesodermal nonvascular cells, a hypothesis that calls for further research.

Location of Supratentorial Dermoid Cysts

Unlike infratentorial dermoid cysts, most supratentorial dermoid tumors are located on the skull base near the midline,\(^7,12,19,23,29,31,44,49,51\) suprasellar,\(^7,11,19\) or pineal dermoid tumors\(^11,22,38\) are very rare. With lesions at this site, according to our theory, the point of attachment may be the great vein of Galen.

Clinical Course

Age at onset varies greatly, but it is noteworthy that the greatest incidence of posterior fossa dermoid cysts is in the first decade, whereas supratentorial dermoid lesions appear in the 20- to 30-year age group.\(^7\) The duration of symptoms is related to the location of the lesion. With frontobasal or temporobasal lesions the symptoms are of more than 10 years' duration and epilepsy is the only complaint reported by patients. Suprasellar lesions reveal themselves early with visual disturbances or headache due to hydrocephalus.

An intracranial dermoid cyst may rupture spontaneously, producing sudden chemical meningitis,\(^8,21,44\) numbness of the legs,\(^11,17\) acute hemiparesis,\(^46\) or seizures.\(^2,19,23,47\) Arachnoiditis or chronic ventriculitis due to repeated leakage of the cyst contents into subarachnoid or ventricular spaces may produce hydrocephalus or mental deterioration.\(^34,37\) For these reasons, we believe that all supratentorial dermoid cysts rupture sooner or later, especially when they become large. Because the capsule is soft, spontaneous rupture is facilitated by movements of the head and even more by brain pulsations, of which the hammer-like action causes a cleft and leakage from the cyst into the subarachnoid or ventricular system.

Clinical examination is always unrevealing in patients with paramedian supratentorial dermoid cysts. All of our patients with these lesions were free from deficits. In fact, frontobasal or temporobasal lesions can reach a considerable size without causing any neurological sign. On the other hand, the mass effect created by suprasellar dermoid cysts produces a chiasmal syndrome.

Radiographic Findings

Currently, dermoid cysts can be diagnosed early and safely with the aid of CT and MR imaging. Computerized tomography scans demonstrate a marked hypodense area with values ranging from 0 to -150 HU;\(^2,23,19,21,23,29,31,44,47,49\) this area is not enhanced by contrast medium, although one enhancing infratentorial dermoid cyst has been reported.\(^51\) The cysts appear as well-defined roundish lesions, with regular margins, which enlarge like a balloon, with no flow along the CSF pathways. Very frequently, coronal sections show involvement of the skull base but no skeletal or other anomalies. A calcified rim of high absorption around the wall has been commonly reported,\(^2,9,17,19,21,44,47,51\) but it is more characteristic to find fatty material scattered throughout the subarachnoid cisterns in ruptured dermoid cysts,\(^2,17,13,7,21,44,47,49\) as in four of our six patients. The MR appearance includes high signal intensity on T\(_2\)-weighted images and lower signal intensity on the T\(_1\)-weighted images.\(^11,17,22,47\) A thin rim of signal void area represents calcification.\(^22\) As with CT scanning, free fat globules in the subarachnoid spaces may reveal a ruptured dermoid cyst.\(^17,47\) Differential diagnosis includes epidermoid cysts, arachnoid cysts, and cystic craniopharyngiomas. These lesions all closely resemble dermoid cysts on CT and MR studies. Several factors such as patient age, clinical history, location of the lesion, presence of calcifications, and lower absorp-
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tion values in dermoid tumors may be helpful in the preoperative diagnosis.

Treatment

Dermoid cysts are benign intracranial lesions; carcinomatous transformation has been reported in only one case.14 The primary aim of surgical treatment in dermoid cysts is removal of the cyst contents, which is easily effected by aspiration. It is unwise in many cases to attempt removal of the capsule, which frequently adheres to cranial nerves and arteries. In such cases, the operating microscope allows accurate dissection but, if the risk for the patient is too high, conservative treatment affords long survival with very good quality of life.

The absence of recurrence in our series, despite incomplete removal of the capsule in all but three patients, is in agreement with the literature; we could find only two instances of recurrence after subtotal removal of a supratentorial dermoid cyst.2,3 This result is remarkable, especially in light of the not-infrequent recurrence of epidermoid cysts after partial excision.8,23-44 The reason for the difference is unclear.

Conclusions

Supratentorial dermoid cysts are too rare for any conclusions to be drawn. Their relationship with the cavernous sinus requires further evaluation in order to assess their etiopathogenetic nature more accurately. On the other hand, the frequent spontaneous rupture and lack of recurrence in spite of subtotal removal of the capsule in our series led us to sharply distinguish dermoid from epidermoid cysts. The biological behavior of the latter appears related to morbidity and mortality rates that are not inconsiderable.

References

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P. Lunardi and P. Missori

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Address reprint requests to: Paolo Missori, M.D., Neurochirurgia, Viale dell’Università 30 a, 00185 Roma, Italy.