A rare case of temporoparietal dermoid with intracranial communication

Abstract
Dermoid cysts are benign, slow growing tumors derived from ectopic inclusions of epithelial cells during closure of neural tube. Preoperative diagnosis of these lesions through characteristic computed tomography (CT) and magnetic resonance imaging (MRI) appearances is generally possible. However, the radiologic features are uncommon and the lesion is usually misdiagnosed with other tumors. Clinical, radiological and histopathological features of a dermoid cyst are reviewed. We report a case of young female with an unusual dumbbell dermoid cyst that was unexpectedly located in the temporoparietal region of the skull at a site where epidermoids are usually found. The imaging features were characteristic, of this type of lesion.

Keywords: dermoid cyst, brain, magnetic resonance imaging

Introduction
Dermoid cysts are benign dysembryogenic tumors originating from ectopic inclusions of epithelial cells during closure of neural tube. They account for about 0.04 to 0.7% of all intracranial tumors with a predilection in the cranial midline, the parasellar and sylvian cisterns. Dermoid cysts have characteristic CT and MRI features making their preoperative diagnosis straightforward possible.

Case report
A 11-year-old girl presented with a gradually increasing swelling along left side of face. Starting at 5 years of age, the mass slowly progressed from small, soft, tender mass to a firm and nontender swelling. On physical examination, a 8x5cm mass was palpated in temporal region on left side, and a small pit on the overlying skin. A CT scan of the skull (Figure 1) showed a large mass in left temporal region and groove in the skull vault. On MRI, a large well defined cystic lesion was seen with areas of fat intensity at places. No area of diffusion restriction was seen and post contrast enhancement of soft tissues was noted. During surgery, incision was followed by white, cheesy material exuding from the defect in the skull bone. There was no attachment of the soft tissue mass with the underlying dura and the entire swelling could be removed en masse. Histologic examination of the surgical specimen (Figure 2) revealed findings consistent with ruptured dermoid cyst along with presence of a small number of eccrine glands and hair follicles beneath the epithelium. Granulation tissue was surrounding the cyst, and there were evidence of periosteal reaction with new bone formation in the sections of the bone examined.

Figure 1 NCCT of the patient showing a large well defined rounded extra cranial lesion showing areas of fluid and fat attenuation with vertical groove in the inner surface of the calvarium with slightly sclerotic borders.

Figure 2 T1W, T2W and T2W FS images of the patient showing extracranial well defined lesion in the temporo-parietal region on left side with areas of cystic (blue arrowhead) and fat intensity (red arrowhead) within.
Discussion

Congenital dermal cysts of the scalp and calvarium present as either dermoid or epidermal cysts. Histologically, dermoid cysts are described to have a well-developed lining of stratified squamous epithelium and contain elements of mesodermal origin, like hair follicles, sebaceous glands. They can be located anywhere but generally in the periorbital region, over the anterior fontanelle, or in the occipital area. Cutaneous changes and intracranial extension of periorbital dermoid cysts is infrequently reported in literature.\(^1\)\(^2\) Dermal cysts of the anterior fontanelle are commonly described in the subgaleal space over the anterior fontanelle. They cause flattening and depression of the outer table of the calvarium.\(^3\)\(^4\) Occipital dermoid cysts and sinuses are generally seen in the midline of the occipital bone, and dermal sinus extends intracranially through the bone defect located here.\(^5\)\(^6\)

Epidermal cysts of the diploe, contrary to the appearance of dermoid cysts do not contain hair follicles or eccrine glands, instead are filled with cholesterol crystals and keratinized epithelial debris. They are reported in the lateral aspect of the frontal and parietal bones, adjacent to the coronal suture.\(^7\)\(^8\)\(^9\) Less common sites include squamosa of the temporal bone, occipital bone, and, rarely, in the frontal region. In skull radiographs, epidermoid cysts appear as round or oval radiolucent defects with defined sclerotic margins and variable involvement of skull vault. Involvement of the dura with occasional compression of the brain has also been less frequently described.\(^10\)\(^11\)\(^12\) During early gestation, dermal cysts in the midline originate from ectodermal rests that become pinched-off during closure of the neural groove. In the case of dermoid cysts that are not in the midline, onset is later in life and is postulated to be due to an incomplete separation of the neural tube from the surface ectoderm. To sum up, epidermoid cyst contains only ectodermal elements, whereas both ectodermal and mesodermal elements results in formation of a dermoid cyst. The young female reported in this paper had a ruptured dermoid cyst in the temporoparietal region of the skull. The diagnosis was made on the basis of the glandular structures present in the cyst wall. The cyst contained both epicranial and epidural component, and a small defect in the skull bone was the bridge between the two parts.

Only two such dumbbell dermoid cysts in the temporoparietal region of the skull have been described in literature,\(^13\) an area of the skull that is more typical for epidermal cyst of the diploe than for true dermoid cyst. However, an overlap in location is not inconceivable as the two lesions are indeed closely related developmentally. A nearly vertical groove in the inner table of skull vault with defined borders is a most unusual and characteristic feature of this type of lesion.

Acknowledgments

None.

Conflicts of interest

The authors declare that there is no conflicts of interest.

References

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