CASE REPORT

CHILDHOOD SELLAR DERMOID CYST MIMICKING A CRANIO-PHARYNGIOMA: THE ADDED BENEFIT OF MRI SCAN

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ABSTRACT

BACKGROUND: Intrasellar brain dermoid cysts are extremely unusual cases with or without sudden symptoms. It is a congenital dysembryogenic lesion that is formed during the period of neural tube closure, by the trapping of embryonic ectoderm. This defect explains the propensity of dermoid cyst for the midline localization.The discovery of completely asymptomatic dermoid cysts in the pediatric population is exceedingly rare. Despite the advances in imaging modalities, it sometimes remains difficult to exclude the differential diagnosis of craniopharyngioma. **CASE REPORT:** We describe a case of a 07-year-old boy addressed for suspicion of craniopharyngioma diagnosed by decreased visual acuity and bitemporal hemianopia. Endocrine workup showed pituitary hormones within normal levels. CT scan was done which shows a large hypodense sellar lesion with intralesional calcifications. Despite the unusual localization and size of this lesion, an MRI brain was performed which showed a large abnormal signal intensity mass in the sella. It shows heterogenous signals on T2 weighted and FLAIR images, hyperintense signals on T1 weighted images without post contrast enhancement. The diagnosis of craniopharyngioma was ruled out in favour of a dermoid cyst. **CONCLUSION:** The reported case reveals an unusual localization of a dermoid cyst, which caused a diagnostic challenge. In the sellar area, it is often not fruitful to differentiate craniopharyngioma and dermoid cyst as they may have very similar CT findings however multisequential MRI imaging thus helps and gives an added benefit in formulating the diagnosis of sellar dermoid by their signal changes in different sequences of MRI.
Keywords: Craniopharyngioma; Dermoid cyst; MRI; Sellar.

Introduction

Intracranial dermoid cysts are rare congenital conditions that account for 0.04 - 0.6% of all intracranial tumors. Derived from ectopic cell remnants that are incorporated into the neural tube, they are usually located along the midline, in cisternal spaces and parasellar location with characteristic imaging appearances. Only 12 cases of sellar extending upto supratentorial dermoid cysts have been described so far in children; most of them were associated with a dermal sinus. Usually, slow growth is observed with insinuation of the lesion into the adjacent structures without invasion, therefore the symptoms usually arise by mass effect and compression over adjacent vital structures. The main leading symptoms for diagnosis are epileptic seizures, meningitis, focal neurological deficit, hydrocephalus, visual disturbances or sudden death. Their diagnosis may be challenging due to their rarity and variability of images.[1,2]

On computed tomography (CT) scan, typically dermoid cysts appear as well defined, low attenuating (-20 to -140) due to their lipid content. The sebaceous lipid

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material that simulate fat giving a characteristic hypodense appearance on CT, and predominantly T1-weighted (T1-W) hyperintense on MRI. Calcifications may be seen in the wall. Enhancement after intravenous gadolinium administration is rare. Although not pathognomonic, this classic imaging appearance is usually consistent with the diagnosis of a dermoid cyst. The differential diagnoses to consider are craniopharyngioma, epidermoid cyst, arachnoid cyst, teratoma, and lipoma. The treatment of choice is total surgical resection with careful dissection between the cyst capsule and the surrounding neurovascular structure. Dermoid cysts rarely have intracranial positions and to best of our knowledge, only two cases in literature extending from sellar to suprasellar region. Associated with a dermal sinus have been described in literature. Here, we describe an unusual case of large sellar dermoid cyst mimicking a craniopharyngioma with no dermal sinus associated.[3,4,5,6]

Case Report

We describe a case of a 07-year-old boy who was admitted from another centre after a presumed diagnosis of craniopharyngioma, diagnosed by decreased visual acuity and bitemporal hemianopia. Endocrine workup showed pituitary hormones within normal levels.CT scan was done which shows a large hypodense sellar lesion with intralesional calcifications measuring approximately 4.3 x 3.8 x 3.5 cms (AP x TS x CC dimensions). Despite the unusual localization and size of this lesion, an MRI brain was performed which showed a large abnormal signal intensity mass in the sella extending upto suprasellar region. It shows heterogenous signals on T2 weighted and FLAIR images, hyperintense signals on T1 weighted images without post contrast enhancement. No signs of rupture or cisternal dissemination was noted. The diagnosis of craniopharyngioma was ruled out in favour of a dermoid cyst.No dermal sinus was identified on clinical examination and images. (Fig.1,2 & 3). Neurosurgery department was taken on board which did an open craniotomy. The intraoperative findings revealed sellar demoid with extension into the suprasellar component. Off-white colored sebum was extracted after careful removal of the sebaceous cystic component, few hair follicles were also identified

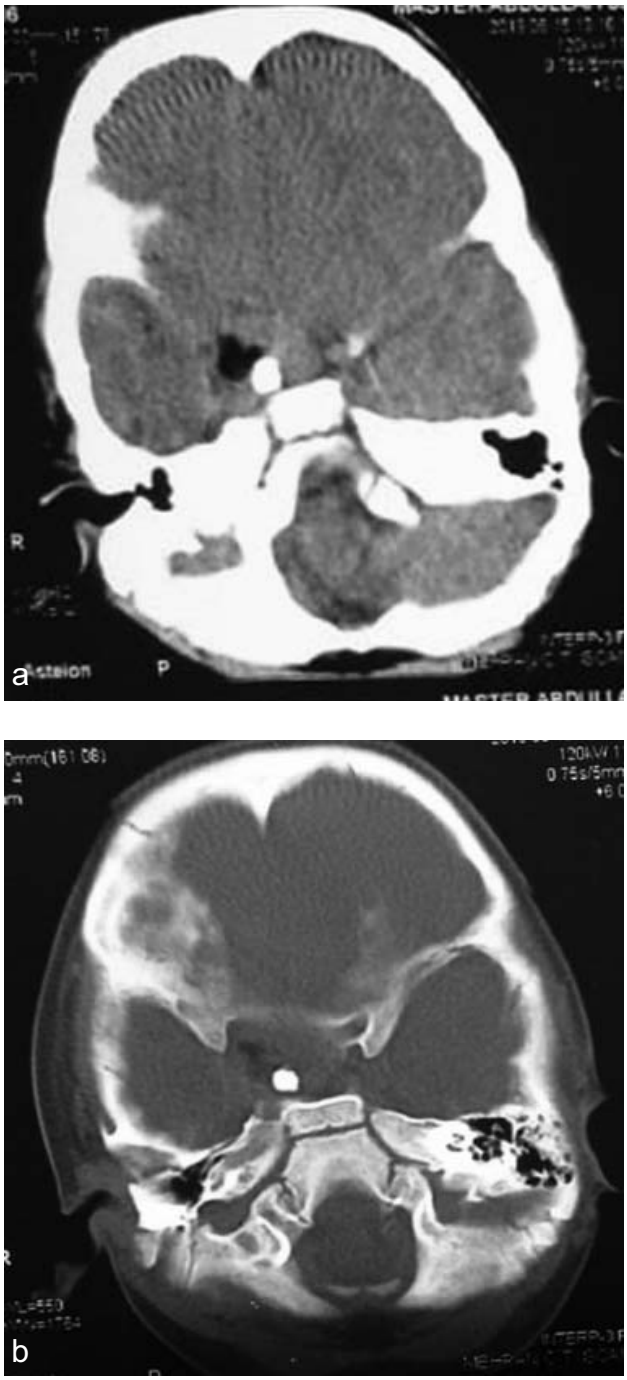
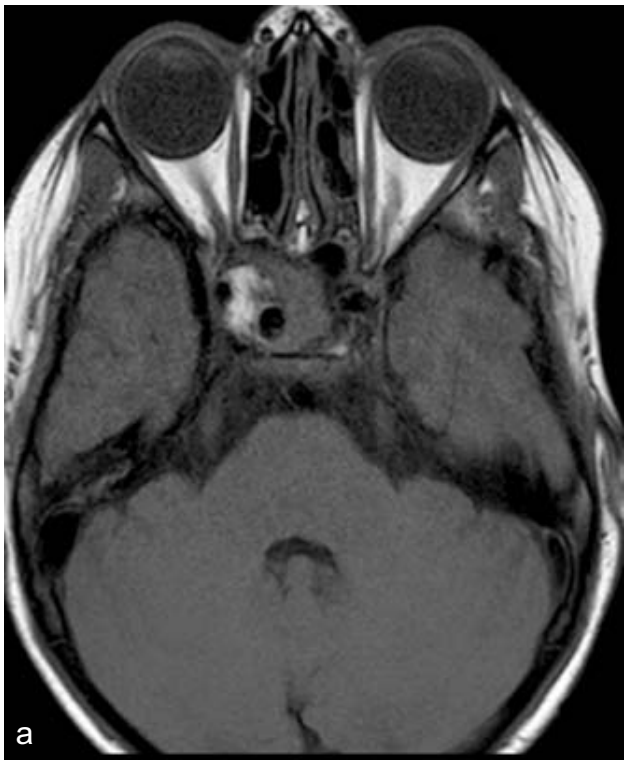


Figure 1: CT Scan Brain Axial view: A solid hypodense mass is seen involving the sellar extending to suprasellar region. It shows a calcifying component with a small fat density within it. It approximately measures 4.3 x 3.8 x 3.5 cm (APxTSxCC). Findings represent dermoid, possibility of craniopharyngioma was also kept in differential.



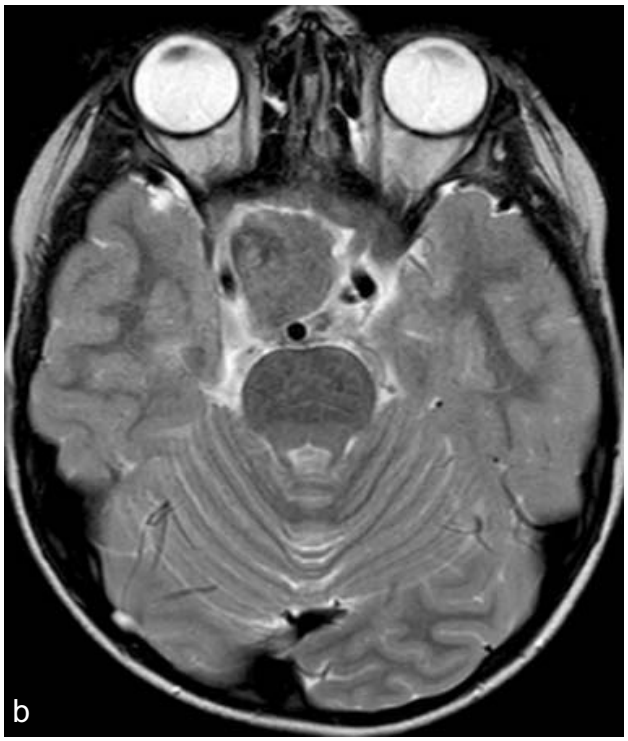
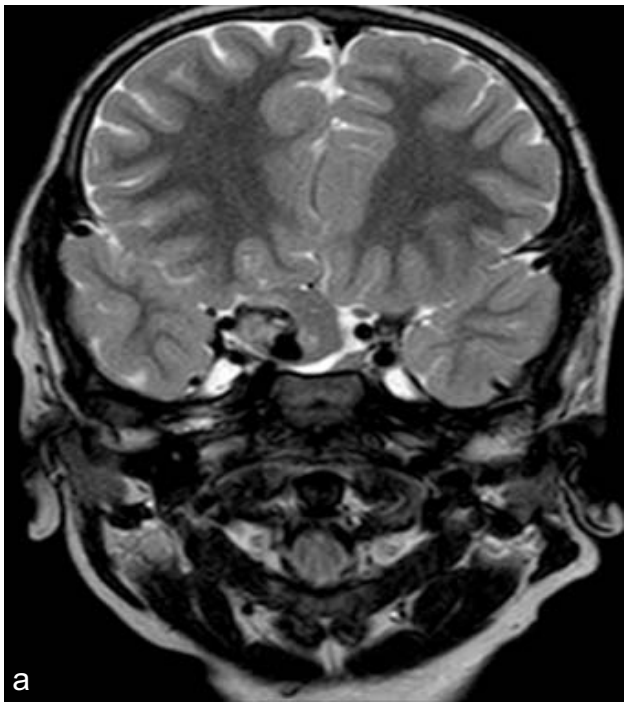


Figure 2: MRI Scan Brain Axial view: An abnormal signal intensity mass with similar measurement is again seen extending from sellar to suprasellar region. It shows high signals on T1 and heterogenous signals on T2 weighted images, suggestive of calcifying and fat component.



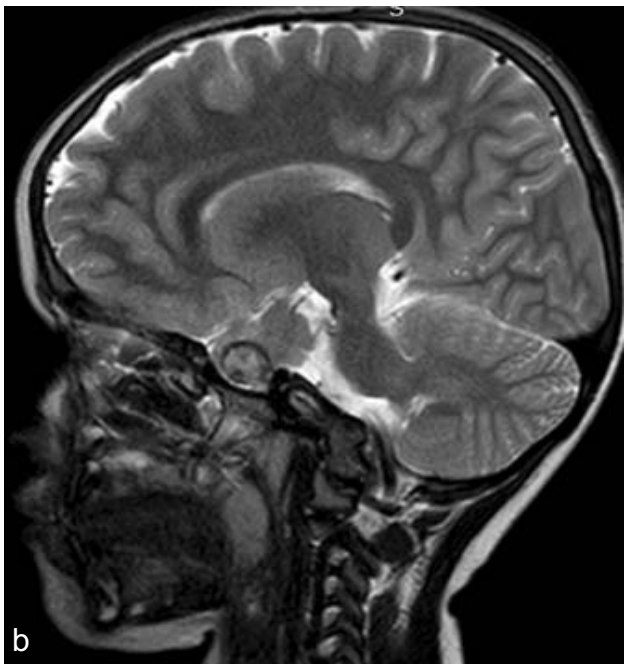


Figure 3: MRI Scan Brain Coronal and Sagittal views: The mass is seen to abut the optic chiasma and internal carotid arteries without evidence of invasion. Pituitary gland was separately visualized which appeared normal. Hence the diagnosis of sellar/suprasellar dermoid cyst was confirmed on MR imaging.

and removed adjacent to it. The patient showed signs of improvement in the post-surgery phase and was discharged home with follow-up MRI Brain scan in the pediatric OPD.

Imaging Diagnosis

The main common CT finding with dermoid cysts/ craniopharyngioma is the capsular and intralesional calcification, which makes it very attractive to perform brain CT scan as a first line investigation. The vast majority of craniopharyngiomas (95%) have a topologically suprasellar component, whereas dermoids due to their earlier formation are mostly located in the intrasellar regionin or close to the midline. Similarly, to craniopharyngioma and our MRI finding, literature refers to dermoids as mostly hyper-intense on T1-weighted images and hypointense to heterogenouson T2-weighted images due to a higher density of cholesterol within the tumor. Consideringthe fact of uncommon localization and the initial radiological characteristics that the diagnosis of this suprasellar lesion was misled for a craniopharyngioma. Differential diagnosis between two is cumbersome on CT scan so MRI is the modality of choice at this point in time. Dermoid cyst usually presentswith high signal on T1 weighted images due to the sebaceous secretions that accumulate inside the cyst. They generally do not enhance in opposition to cranio-pharyngiomas that normally do. Indeed, the solid components of the dermoid lead to a restriction in diffusion within the tumor in comparison to purely cystic and epidermoid lesions or craniopharyngioma. Apart from that, in comparison to craniopharyngioma, MRI scan also shows high signals in DWI images in cases of dermoid.[1,7,8]

Discussion

Intracranial dermoid cysts are rare benign neoplasms that are commonly located in midline and are caused by embryological malformation during the development of the neural tube between the 3rd and 5th weeks of gestation. These cysts are lined by squamous epithelium and contain skin appendages such as hair follicles, sebaceous glands, nails, and teeth. The lesion enlarges as a result of its increased content of glandular secretions and epithelial desquamation, and as it grows, many symptoms result secondary to the compressed neural structures.[9]

The rare CT hyper density in combination with T2 weighted hypo intensity on MRI of dermoid is thought to be due to combination of saponification of lipid/keratinized debris with secondary microcalcification in suspension, partially liquefied cholesterol, high protein content and hemosiderin or iron calcium complexes relating to previous episodes of haemorrhage within the cyst. In our cases, the lesion was hyperdense on CT scan and hypointense on T2 weighted MRI with only minimal fat density visualized in one of the cases.[5]

On MRI sequences, typically, dermoid cyst appears T1 hyperintense (due to cholesterol components) and is heterogeneous on T2-W images. On CT, typically, they are hypodense with attenuation values equal to that of fat density. However, this may not always be the reality. By virtue of their pathologic contents, rarely, dermoid cyst can show atypical or unusual appearance. It is important to familiarize with this unusual appearance of dermoid cysts described in our casefor the correct preoperative diagnosis and better surgical planning. T1 hyperintense (fat) droplets in the subarachnoid spaces may be visible if there is rupture of dermoid cyst. Such rupture can either be spontaneous or can occur at surgery resulting in chemical meningitis which may be severe leading to vasospasm, infarction, and death.[10]

Craniopharyngioma is an aggressive lesion often found in midline including the intra- and suprasellar region with serious skull-based erosion, a frequent finding on CT images. In children (adamantinomatous type), usually presents a solid (isodense) mass with cystic (hypodense) component (sometimes multi cystic); nodular calcifications and nodular or rim enhancement. In adults (papillary type), lesion appears isodense with nodular enhancement.

Common MR findings for the solid component are heterogeneous signal on T1and T2 weighted images with heterogeneous enhancement; cystic components are variably hyperintense on T2-w while signal varies with the cyst contents on DWI and T1-w images (short T1 due to high protein content), cyst wall shows strong post contrast enhancement. Dermoid cyst showed minimal or no enhancement.[11]

CT and MR images have been relevant for correct diagnosis that allows the most appropriate choosing for a better surgical procedure case by case.

Conclusion

The reported case reveals an unusual localization of a dermoid cyst, which caused a diagnostic challenge. In the sellar area, With CT imaging is not often fruitful in differentiating between craniopharyngioma and dermoid cyst as they may have very similar findings. Multisequential MRI imaging thus helps and gives an added benefit in formulating the diagnosis of by virtue of their intrinsic signal characteristics onvarious sequences also for preoperative planning. Identification of typical MRI imaging findings are crucial for Radiologist for correct interpretation of the lesion and may impact on prognostic potential.

Conflict of interest: Declared None.

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