THE CONSTELLATION OF CRANIOFACIAL AND CNS ANOMALIES IN A SINGLE PATIENT

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ABSTRACT

We report an unusual case of a 2 year child who presented to the civil hospital with congenital facial deformity and delayed developmental milestones. His MRI and CT scan demonstrated peri-callosal lipoma with callosal dysgenesis, right parasagittal arachnoid cyst, left para-sellar dermoid with internal carotid artery encasement, falx calcification, right nasal dermoid causing hypertelorism, with ipsilateral nasal cleft, frontal and cribriform plate defects. To the best of author's knowledge, this unique combination of cranio-facial and CNS anomalies has not been described earlier in the literature.

Key words: Dermoid Cyst, Peri-Callosal Lipoma, Callosal Dysgenesis, Nasal Dermoid, Falx Calcification.

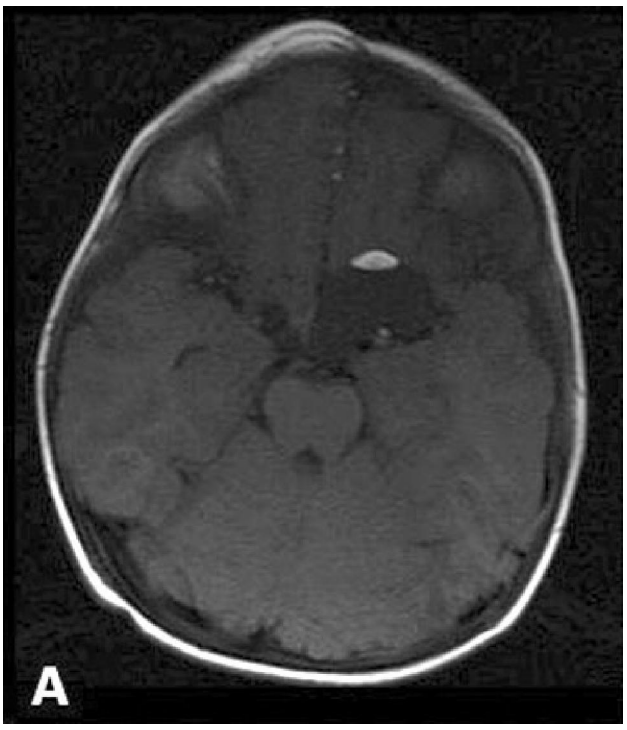
Introduction

Craniofacial syndromes include developmental malformations of the face and skull that are associated with central nervous system malformations. These syndromes are often evaluated with CT and MR imaging, which allow detailed anatomic description for surgical planning and detection of associated intracranial lesions.[1] We present the multi-modality imaging features of craniofacial and CNS anomalies of different embryologic origin with special focus on atypical appearance of dermoid cyst and review of the literature.

Case Summary

A 2 year old child was presented to the civil hospital with congenital facial deformity and delayed developmental milestones. According to his mother, he was born with pre-auricular tags, prominent forehead and a nasal cleft but there was no cleft lip or palate. He was born via normal vaginal delivery. There was no significant family history and her mother had no history of drug usage or radiation exposure.

He was further evaluated with brain MRI and head CT scan.



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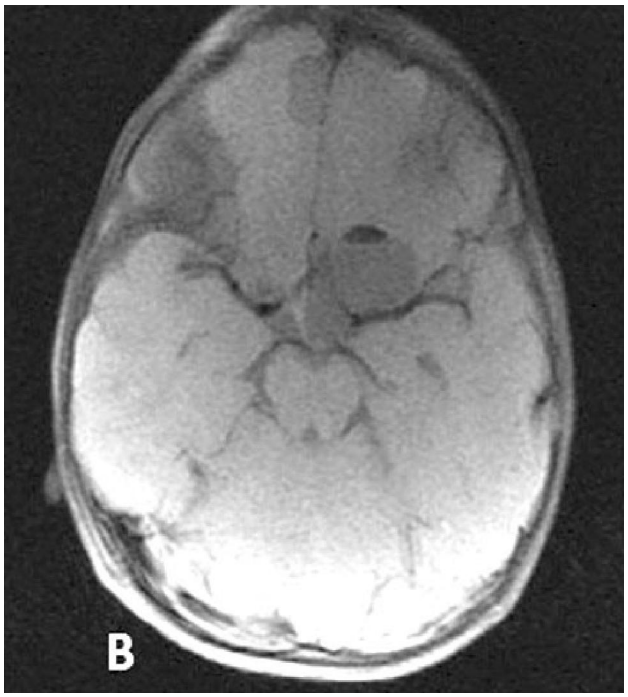


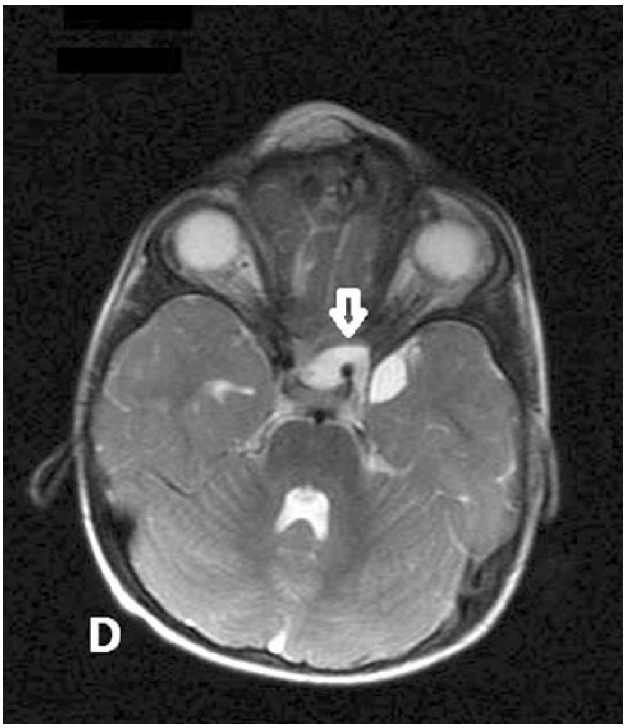
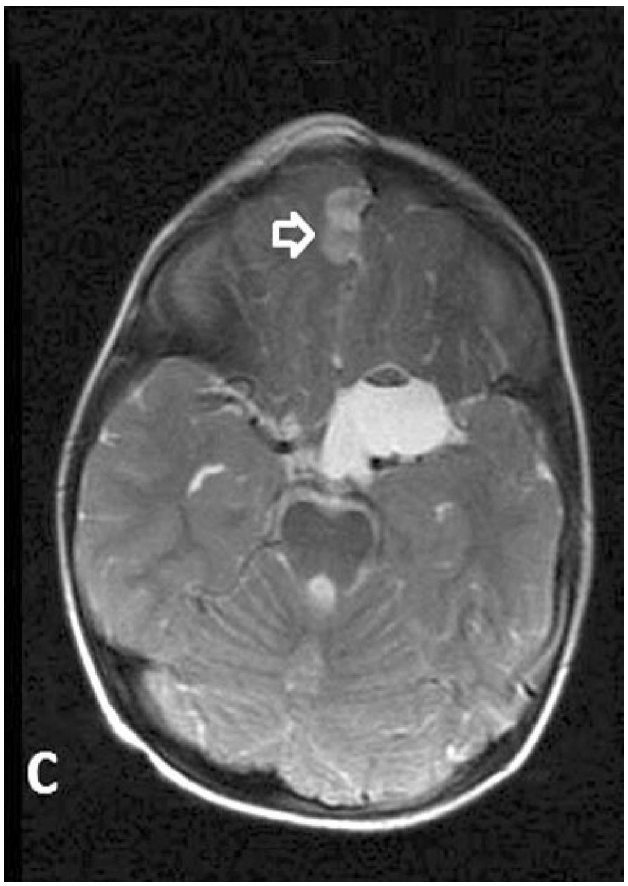
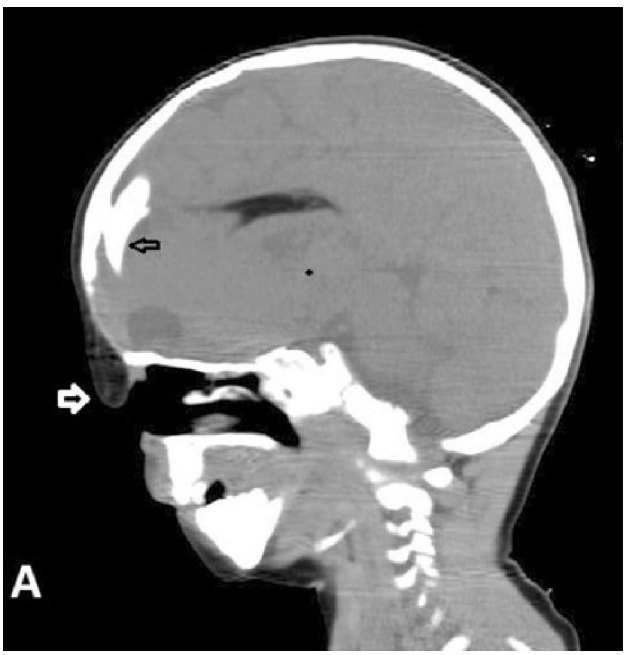


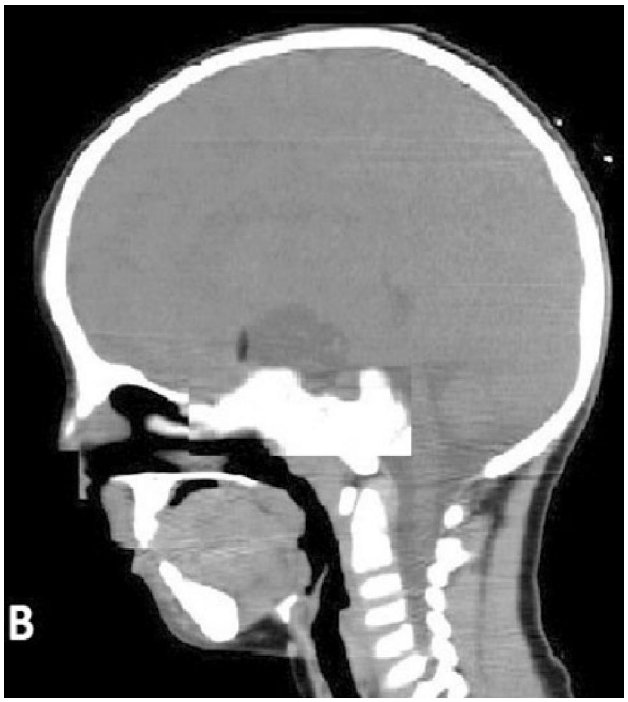
Figure 1(A-D): Contagious axial T1W MRI **(A)** with fat saturation **(B)** showing fat fluid level, with ICA encasement on T2W images (arrow in **D**)

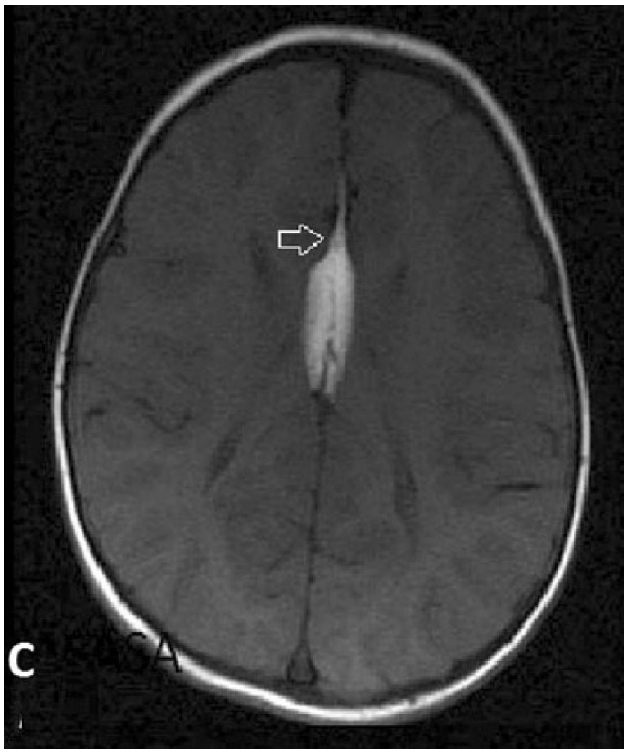


Magnetic Resonance Imaging was performed on GE Health Care Signa HDxt 1.5 Tesla scanner using the head coil with slice thickness of 4 mm with inter slice width of 1 mm. Multi-planar, multi-sequential images in T1W, T2W fast spin echo sequence, FLAIR and



post contrast T1W sequence were obtained. Plain brain CT was performed with 16 slice Toshiba Spiral CT and flat-panel volume technique. The scanning parameters selected were 120 KV, 150mA and 1 mm slice thickness.





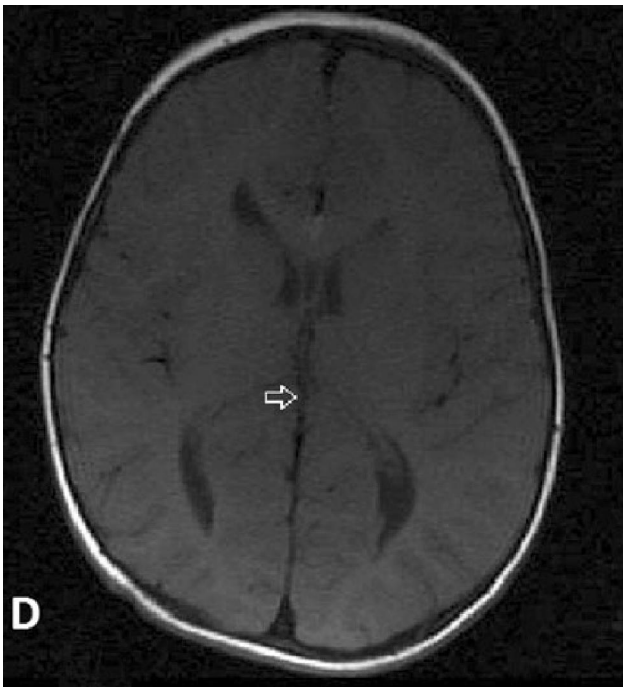


Figure 2(A-D): Sagittal and parasagittal CT sections showing falxcerbri calcification (white arrow in A), nasal dermoid (black arrow in B) and left parasellardermoid (D).Contagious axial T1W MRI images showing peri-callosal lipoma & callosal dysgenesis (arrows in Fig. C & D).

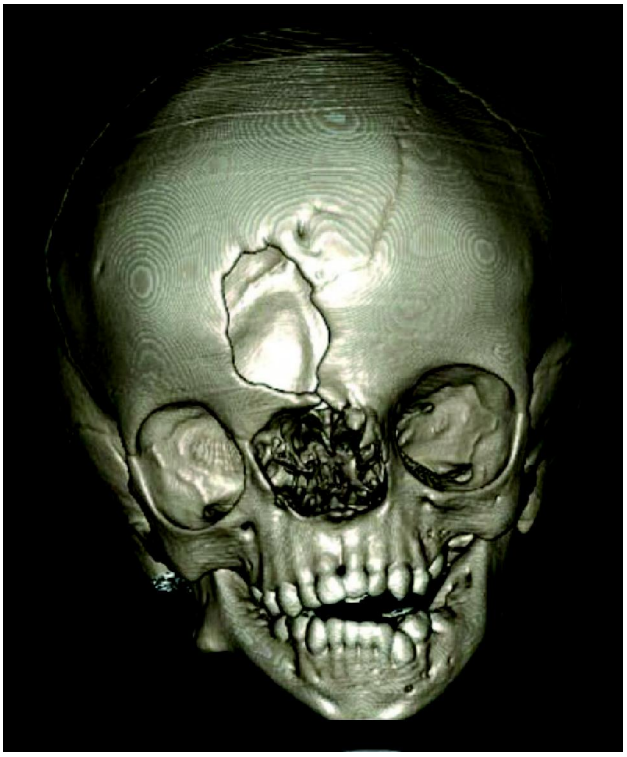


Figure 3: Volume rendered-3D CT of the skull showing right frontal and nasal defects

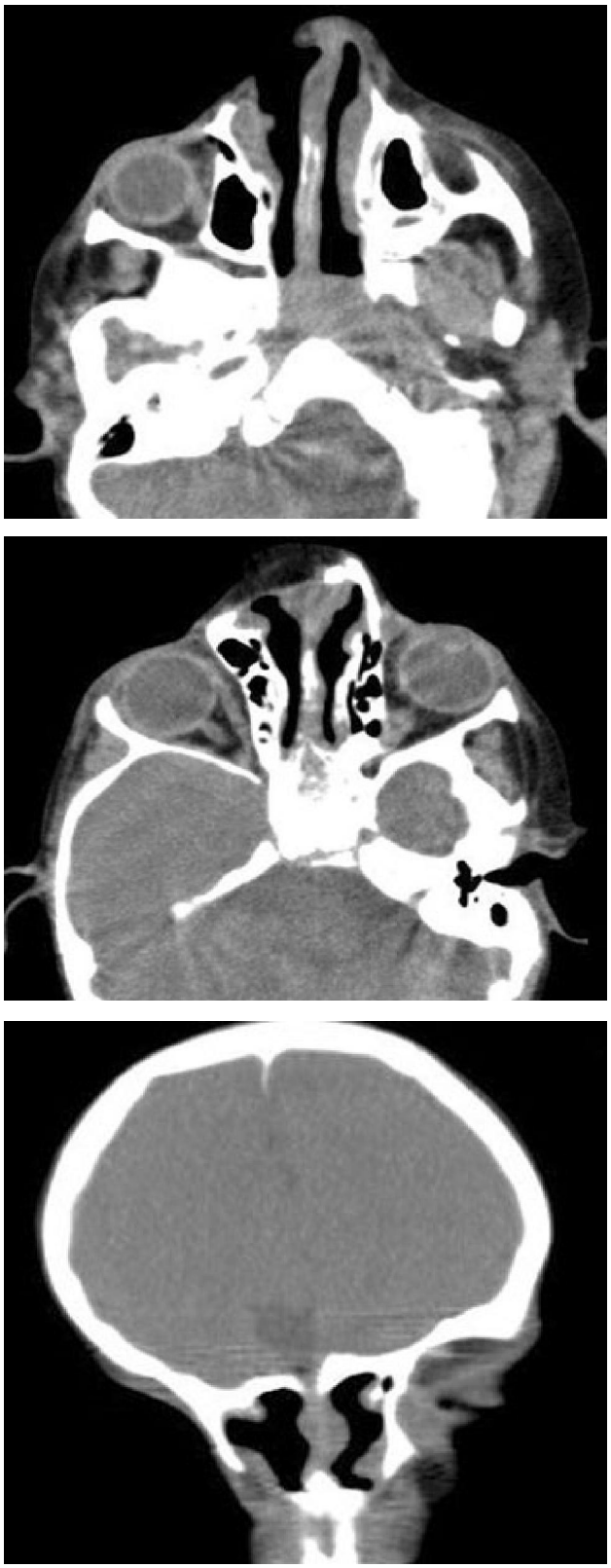


Figure 4: Contagious axial sections showing cutaneous osseo-cartilagenous nasal defect; Coronal CT showing right cribiform plate defect and para-sagittal arachnoid cyst.

There was a 26 x 30 x 23 mm (CC x TV x AP) well defined non-enhancing lesion showing fat fluid level in left para-sellar region with supra-sellar and extension and encasement of supra-clinoid and cavernous segments of the ipsilateral internal carotid artery (seen on T2W sequence). The fatty component was hyper intense on T1W, hypo intense on T2W and showing suppression on fat sat sequence with the average attenuation value of -90 Hounsfield units on plain CT (Fig. 1). It is mildly displacing the fronto-temporal lobe and basal ganglia.

There wasan 11 x 36 mm peri-callosal lipoma and dysgenesis of the corpus callosum involving its splenium and body. There was a small 10 x 11 mm right extra-axial arachnoid cyst along falxcerebri measuring with extensive calcification of the anterior falxcerbri (Fig. 2).

A 32 x 21 mm right frontal osseous defect leading to mild anterior frontal lobe herniation was noted with ipsilateral cribiform plate agenesis. There was a right sided full thickness facial cutaneous osseo-cartilagenous defect 13 mm in width, extending from nasal root to apex with nasal bone agenesis and 16 x 11 mm right nasal dermoid causing hypertelorism (Fig. 3-4).

Discussion

Congenital midline naso-frontal masses are the result of faulty regression of the embryologic dural diverticulum from the prenasal space and occur in one of every 20,000 - 40,000 births. They are rare but important anomalies that almost always present in childhood. Their rate of occurrence is approximately one in every 20,000 - 40,000 live births. These include nasal encephaloceles, nasal gliomas and nasal dermoid/epidermoid. The type of mass is determined by the nature of the faulty regression.[1-2] The mass may be intranasal, extra-nasal or a combination of the two. Extra nasal glabellar masses are due to extension of the diverticulum through the foramen cecum and fonticulus frontalis. Dermoid and epidermoid cysts occur when skin elements are pulled into the prenasal space along with the regressing dural diverticulum.

In 1993, Chakrabortty S et al[3] reported an unusual case of a congenital frontal bone defect compatible to our case, with intact overlying scalp and intact

underlying dura mater which was later on repaired by cranioplasty for protective and cosmetic purposes. Intracranial dermoid cysts are rare tumors constituting less than 1% of all intracranial tumors. They are considered to be the product of a defective closure of the neural tube, which occurs between the third and fifth gestational week. Endo H et al in 2011,[4] reported a patient with an extradural dermoid cyst of the right parasellar region, causing right visual disturbance. CT revealed a hypodense mass lesion with rimlike calcification at the right parasellar region, accompanying marked erosion of the adjacent skull base. To our knowledge there has been only one reported case ofun ruptured intracranial epidermoid with a fluid level documented by Cornell SH et al in 1977.[5]

Intracranial dermoids are most commonly seen below the tentorium. These may also become symptomatic due to rupture which results in spillage ofcontents into the subarachnoid space and the ventricles, resulting in an aseptic chemical meningitis, vasospasm, infarction, or increased intracranial pressure.[6]

Dermoid cysts confined to the cavernous sinus have seldom been reported in the literature. Rato RM et al in 2012 reported the first incidental case of a cavernous sinus dermoid cyst in a 21-year-old woman.[7] They are usually developed between the two layers of its lateral dural wall and are classified as interdural. They present heterogeneous signal due to its heterogeneous content, high signal intensity on T1-W and low or mixed signal intensity on T2-WI due to the presence of fat[8] - compatible to our case. They displace medially the cavernous ICA without narrowing it; however, our case demonstrated ICA encasement by the cyst.

Craniofacial syndromes include developmental malformations of the face and skull that are associated with CNS malformations. We present the multi-modality imaging features of multiple cranio-facial and CNS anomalies in unique combination, not been described earlier in the literatureto the best of author's knowledge. The presence of central nervous system anomalies is extremely important in these syndromes because they often affect IQ and prognosis and influence outcome.The multi-disciplinary approach was taken for our patient with involvement of Neurology, Neurosurgery and Plastic surgical department and initially nasal cleft repair was performed with later on cranioplasty repair for frontal bone and follow up neurosurgery visits was decided.

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