Developmental persistent falcine sinus with cranium bifidum occultum, craniosynostosis, and associated anomalies: A unique case

Arvinder Singh, Inderpreet Kaur

From Department of Radiodiagnosis, Government Medical College, Amritsar, Punjab, India

Correspondence to: Dr. Arvinder Singh, Department of Radiodiagnosis, Government Medical College, Amritsar, Punjab, India. E-mail: arvinderdr@rediffmail.com

Received - 13 January 2018 Initial Review - 09 February 2018 Published Online - 09 March 2018

ABSTRACT

The falcine sinus is an intrauterine anatomic structure located in the falx cerebri that is closed after birth and persistence of this embryologic falcine sinus is considered as a rare variation of the venous pathway which is associated with defect in the development of the straight sinus leading to formation of an alternate venous pathway served by the persistent falcine sinus. Cranium bifidum occultum is another rare skull ossification disorder referred to as the Catlin mark characterized by ossification defects in the parietal bones. Many other associated anomalies can be seen with persistent falcine sinus apart from cranium bifidum occultum which include absent or dysplastic tentorium cerebelli, agenesis of the corpus callosum, apert syndrome, atretic occipital/parietal encephalocele, vein of galen malformation, osteogenesis imperfecta, and chiari malformation Type II. We describe a rare case of a 10-year-old girl who presented with short stature, intermittent headache, and abnormal skull shape. On radiological examination, persistent falcine sinus was detected with large midline skull defect at the high parietal area. The straight sinus was absent, and there was dysplastic tentorium cerebelli. In addition to these, craniosynostosis was also present with many other associated anomalies. Persistent falcine sinus and cranium bifidum occultum are very rare, and when found, they are associated with many anomalies raising the possibility that they may represent the benign end of the same developmental spectrum. To the best of our knowledge, these constellations of anomalies have been reported in very few children.

Key words: *Associated anomalies, cranium bifidum occultum, persistent falcine sinus*

The falcine sinus is a venous channel in the falx cerebri, which is normally seen in the embryonic state. It connects the great vein of galen with the superior sagittal sinus and closes shortly after birth [1].

The persistent falcine sinus is the condition in which this venous channel remains patent even after birth forming an alternative pathway between deep and superficial venous system of brain [2]. It may be congenital or acquired in nature and may be associated with other abnormalities such as cranium bifidum, atretic encephalocele, dysplastic tentorium cerebelli, malformation of vein of galen, acrocephaly, absence of corpus callosum, apert syndrome, and Chiari II malformation [3]. We are hereby presenting a case of persistent falcine sinus associated with cranium bifidum occultum and craniosynostosis.

CASE REPORT

A 10-year-young female was referred to the department of radiodiagnosis for X-ray skull and cranial magnetic resonance imaging (MRI). She had a history of difficult in learning with low cognitive abilities. No cranial nerve abnormalities were detected. On physical examination, she was short stature. The skull was towering type with a palpable soft tissue swelling in the high parietal region in the midline. The antero-posterior diameter of skull was reduced with increased biparietal measurements. The nasal bridge and malar bones were depressed with evidence of hypertelorism, with a protruding lower jaw (Fig. 1). No evidence of polydactyly or syndactyly was noted.

On plain X-ray, the skull was towering type, fused suture planes (*acrocephaly*). A large central bony defect was noted in the high parietal region with a sharp bony central spike. The frontal bone was flattened, nasal bridge depressed, hypoplastic malar bones, and causing false protruding of lower jaw (*pseudo prognathism*) (Fig. 2a and b).

The computed tomography (CT) scan of the skull was done which confirmed the finding of the plain X-ray skull, showing a large midline defect with a sharp central bony projection (*cranium bifidum occultum*). The overlying scalp tissue was slightly thinned out but was intact. A thin papery membranous tissue was noted between the underlying brain tissue and overlying scalp (*atretic encephalocele*). On volume CT scan, all the sutures were fused including coronal, sagittal, and lambdoid sutures with a large symmetrical bony defect as mentioned above. The biparietal diameter of skull vault was increased as compared to occipitofrontal diameter with a ratio of >92 (*ultra brachycephaly*) (Fig. 2c and d).

The malar bones and nasal bridge were depressed. A complete bony septum was noted passing through the cavity of the left maxillary antrum with a partial bony septum in relation to right maxillary antrum (*Underwood's septa*). The MRI brain T1W, T2W, and flair sequences were done in axial, coronal and sagittal planes to see the status of the intracranial contents. There was the absence of posterior midline falx causing interdigitations of the occipital gyri in the midline with downward herniations through the anterior part of the tentorium cerebelli. The frontal lobes were compressed by the flattened frontal bone causing their crowding of sulcogyral spaces (Fig. 3c and d). The corpus callosum was intact.

On T2W sagittal plane, a single large linear arch-shaped signal void structure was noted in the midline suggestive of a venous vascular channel replacing the deep cerebral venous system. This structure appears to be arising from the site of confluence of great vein of galen and was curving up, reaching superiorly draining into the superior sagittal sinus, just below the site of bony skull defect. The straight sinus was dysplastic in nature with poorly formed torcula high up as compare to normal site (Fig. 3a). In magnetic resonance venography (MRV), the venous sinuses on the left side were hypoplastic, and most of the venous drainage was from right-sided venous channels into the right jugular bulb (Fig. 3b).

The posterior cranial fossa was funnel-shaped with both cerebelli appeared pointed in nature posteriorly with dilated cerebrospinal fluid spaces. The brain stem including pons and medulla was unremarkable.

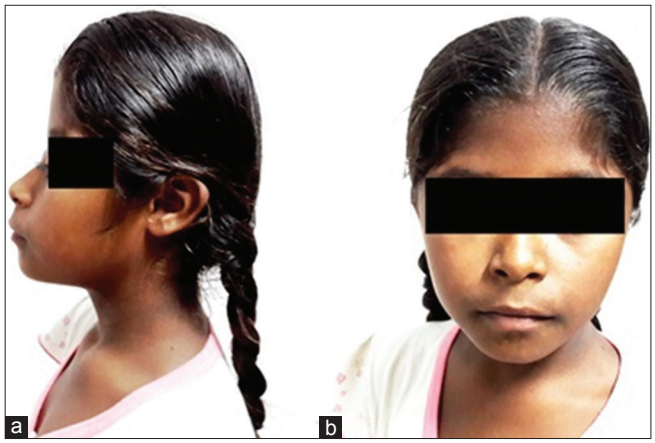

Figure 1: (a and b) Photograph-lateral and frontal views of a 10-year-old female showing towering skull (acrocephaly), depressed nasal bridge, hypertelorism, and pseudo prognathism due to malar bone hypoplasia

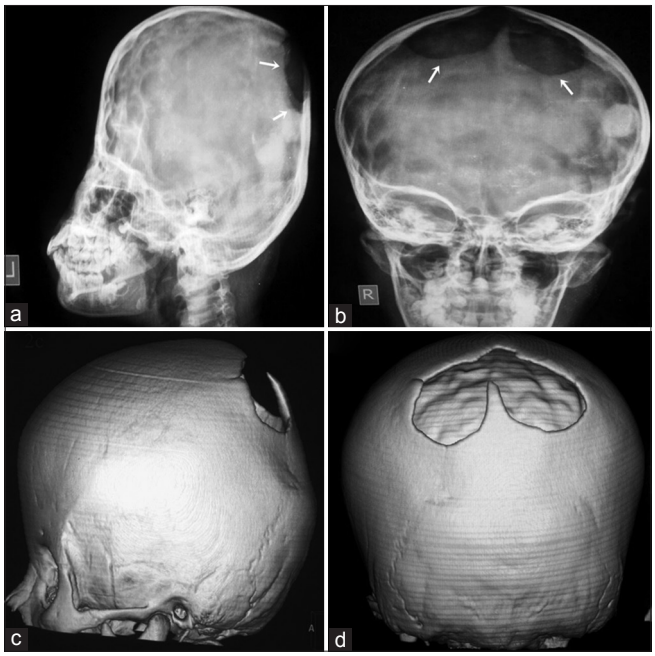

Figure 2: An 10-year-old girl who had a persistent falcine sinus (2a and b): Plain radiograph of the skull lateral and AP views showing a large well defined bony defect in the higher parietal region with a central bony spicule-cranium bifidum occultum (arrow). (2c and d): Volume computed tomography scans showing a large bony defect in a high parietal midline region with fusion of all the suture planes (acro-craniosynostosis) causing towering skull

DISCUSSION

The falcine sinus is an intrauterine anatomic structure located in the region of falx cerebri that normally closes after birth. It is a venous structure which connects the vein of galen to the superior sagittal sinus [1]. A developmental defect of the straight sinus (atretic/hypoplastic sinus) leads to the formation of an alternative pathway from the sagittal plexus to shunt the blood from deep venous system to the superficial venous system known as the *congenital persistent falcine sinus* [2]. Alternately, the acquired form of falcine sinus can be visualized in the event of straight

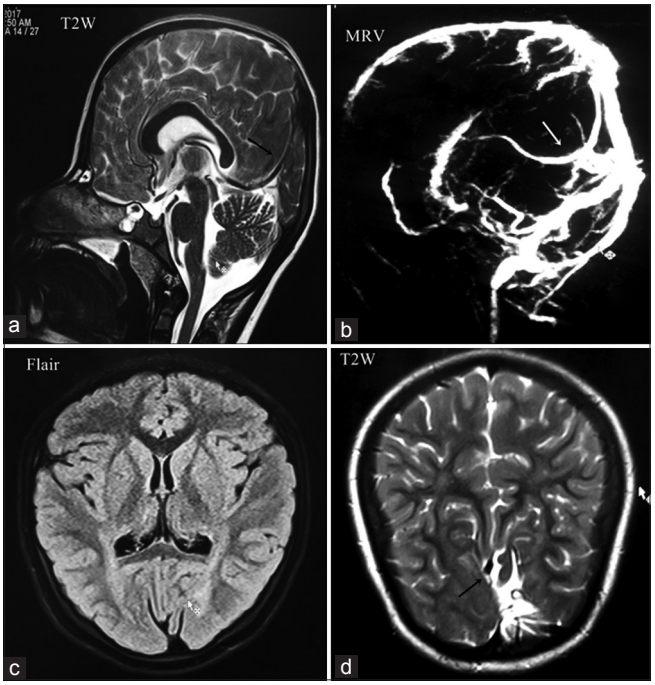

Figure 3: Magnetic resonance images of persistent falcine sinus, (a) sagittal T2-weighted image is demonstrating an abnormal flow void in the interhemispheric space (arrow) from the deep cerebral venous system and reaching up to the superior sagittal sinus, a low position and suspicious partial agenesis of the tentorium cerebelli. (b) magnetic resonance venography showing an absent straight sinus and persistent falcine sinus continuous with the internal cerebral vein (c and d) axial flair and T2-weighted image showing abnormal flow void in interhemispheric space

sinus thrombosis or obstruction leading to its recanalization. There are three types of falcine sinus, i.e., arch type, stick type, and bifurcated type, with an arch type, the most common presentation. The most of persistent falcine sinus are asymptomatic and are not associated with any significant congenital malformations [4].

We report a case of a 10-year-young female having an arch-like persistent falcine sinus with a large central midline bony skull defect at a high parietal region (cranium bifidum occultum) [5]. A small bulging soft tissue swelling could be noted in this region consistent with the atretic encephalocele. Such a midline defect broadly belongs to the neural tube defects due to abnormal neural tube closure. In contrast, a defect without herniation may be due to maldevelopment resulting in bony ossification defect [3].

In our case, the persistent falcine sinus with a large bony defect having soft tissue budge, dysplastic straight sinus, early closure of sutures causing extreme craniosynostosis in the form of acrocephaly indicative of a *developmental persistent falcine sinus* [5].

CONCLUSION

The falcine sinus is a common finding in antenatally, but its association with intracranial abnormalities such as a dysplastic/hypoplastic straight sinus and atretic encephalocele along with early cranial suture fusion (craniosynostosis) and cranial bifidum occultum makes it a rare condition. The various imaging modalities such as plain X-ray skull and CT scan helps in the diagnosis of bony defects associated with this condition. The MRI and MRV facilitates in early diagnosis of the congenital as well as acquired forms of persistent falcine sinus along with straight sinus abnormalities.

REFERENCES

1. Kim MS, Lee GJ. Two cases with persistent falcine sinus as congenital variation. J Korean Neurosurg Soc 2010;48:82-4.
2. Sener RN. Association of persistent falcine sinus with different clinicoradiologic conditions: MR imaging and MR angiography. Comput Med Imaging Graph 2000;24:343-8.
3. Reddy AT, Hedlund GL, Percy AK. Enlarged parietal foramina: Association with cerebral venous and cortical anomalies. Neurology 2000;54:1175-8.
4. Bartels RH, Merx JL, van Overbeeke JJ. Falcine sinus and occipital Encephalocele: A magnetic resonance venography study. J Neurosurg 1998;89:738-41.
5. Yang CA, Peng SS, Hsieh WS, Tsao PN, Chen CY, Chou HC, *et al*. Large parietal midline defect with unusual ridge-like structure at the rim and persistent falcine sinus. Childs Nerv Syst 2013;29:1069-72.

Funding: None; Conflict of Interest: None Stated.

How to cite this article: Singh A, Kaur I. Developmental persistent falcine sinus with cranium bifidum occultum, craniosynostosis, and associated anomalies: A unique case. Indian J Case Reports. 2018; 09 March 2018 4(1):50-52.