Pictorial Essay: Classic Signs in Pediatric Neuroradiology

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Abstract: Background: A number of metaphorical imaging signs have been described in the radiology literature.

Objective: These signs not only make the learning process easier but also increase diagnostic confidence. For this reason, these signs are quite popular among radiologists.

Conclusion: In this article, we have demonstrated classic signs described in pediatric neuroradiology with appropriate examples and illustrations.

Keywords: Pediatric, neuroradiology, CT, MRI, HRCT, pediatric neuroimaging.

1. INTRODUCTION

Classical signs in radiology, when found, bring the impression of an image to mind. Familiarity with these signs increases our diagnostic confidence. Pediatric neuroimaging is no exception and many classical signs have been described in the literature. Knowledge of these signs helps in making diagnosis for daily radiology practice. In this review, we have described various classic signs encountered in pediatric neuroradiology with illustrations. We have also described the underlying cause for the sign and any differential diagnosis of the sign.

2. ICE CREAM CONE SIGN

This sign is seen in High axial Resolution Computed Tomography (HRCT) of temporal bone, representing the normal appearance of incudomalleolar joint. A scoop of ice cream is formed by the head of malleus and cone is formed by the body of the incus (Fig. 1).

Significance:

a. Familiarity with this appearance helps in easy identification of Prussak’s space (lying between scutum and incudomalleolar joint) [1], which is a common site of cholesteatoma.

b. Identification is important to identify ossicular luxation, especially in cases of trauma.

3. HIGH HEEL FOOTPRINT SIGN

This sign is seen on axial Computed Tomography (CT) of skull base and helps in assessing complicated anatomy of the skull base. The anterior aspect of the high heel footprint represents the foramen ovale, and the posterior aspect (the heel itself) represents the foramen spinosum (Fig. 2) [2].

Significance: In cases where foramen spinosum is absent, this sign is not visible. It is noteworthy that absent foramen spinosum is associated with a persistent stapedial artery [3].

4. OMEGA SIGN

This sign is seen in axial Magnetic Resonance Imaging (MRI) of the brain and refers to appearance of central sulcus, resembling inverted omega or sigmoid cook (Fig. 3). This appearance results from a knob of precentral gyrus projecting posteriorly with indentation on central sulcus. This posterior projection or knob represent motor area for the hand [4, 5].

Significance:

a. This sign helps in anatomical identification of the central sulcus on imaging.

b. It also helps in lesion localization with respect to the motor cortex.
5. CORTICAL VEIN SIGN

The cortical vein sign refers to the presence of superficial cortical veins on contrast-enhanced MRI and CT (Fig. 4), seen traversing an enlarged subarachnoid space [6, 7]. In global cerebral atrophy or benign enlargement of the subarachnoid spaces in infancy, the cortical veins are still adjacent to the inner table of the calvaria and thus visible traversing the subarachnoid space. While in a subdural hygroma, the arachnoid membrane and subarachnoid space are displaced by the subdural fluid collection and thus the veins are displaced away from the inner table. Thus, if a cortical vein is seen traversing through a fluid containing collection/space, it is an enlarged subarachnoid space rather than subdural hygroma.

Significance:

a. Can differentiate cerebral atrophy from subdural hygroma [7].

b. This sign can differentiate benign enlargement of the subarachnoid spaces in infancy from subdural hygroma (which may be a sign associated with non-accidental injury).
6. MOLAR TOOTH SIGN

This sign is seen on axial Magnetic Resonance Imaging (MRI) or CT images due to abnormal orientation of superior cerebellar peduncles at the level of midbrain. This sign is seen in Joubert syndrome, in which superior cerebellar peduncles fail to cross in midline. As a result, there is an abnormal anteroposterior orientation of cerebellar peduncles, resembling the roots of molar tooth (Fig. 5) [1, 8].

Significance: This sign is considered as pathognomonic of Joubert syndrome and is seen in 85% of cases [7].

7. BATWING 4TH VENTRICLE SIGN

This sign is seen in the Joubert and associated syndromes. The hypoplastic vermis with apposed cerebellar hemispheres give the fourth ventricle the appearance of a bat with its wings outstretched (Fig. 6) [9, 10].

8. VIKING HELMET SIGN

This sign is seen on coronal CT and MRI images in cases of corpus callosum agenesis. The appearance results from a combination of high riding third ventricle and curved upward-pointed frontal horns of the lateral ventricle (Fig. 7). Medially frontal horns are indented by Probust bundles, which are myelinated tracts consisting of uncrossed commissural fibers [11].

Anomalies in the development of corpus callosum vary from partial, complete or atypical agenesis. They may occur in isolation or can be associated with neuronal migration

Fig. (4). Axial CECT image of brain showing a normal cortical vein (arrow) traversing through the subarachnoid space.

Fig. (5). A: Axial T2 weighted MRI showing abnormal shape of midbrain resembling a molar tooth. B: Magnified image with graphical representation showing roots of molar tooth formed by anteroposterior orientation of superior cerebellar peduncles.

Fig. (6). Axial T2 weighted MRI showing abnormal configuration of 4th ventricle resembling a bat with outstretched wings (arrow).

Fig. (7). (A,B): Coronal CT and MRI brain images showing a case of corpus callosum agenesis with curved upward pointed frontal horns (white arrow in A and B) with high riding third ventricle (black arrow in A and B). C: Magnified MRI image with graphical representation showing helmet is formed by the high riding third ventricle while its horns are formed by curved and upward pointing frontal horns of lateral ventricle.
disorders, Arnold Chiari and Dandy-Walker malformations, trisomy 18 and 13, Aicardi and Apert's syndrome and certain inborn errors of metabolism of lactate and pyruvate [12].

**9. DOUBLE CORTEX SIGN**

This sign is classically seen in gray matter heterotopia- a neuronal migration disorder. Heterotopia results from an early arrest of neuronal migration and consists of a smooth layer of the gray matter separate from overlying cortex. Three varieties of gray matter heterotopias are seen based on MR imaging: subependymal heterotopia, focal subcortical gray matter heterotopia and diffuse subcortical heterotopia [13]. Double cortex sign is seen in focal and diffuse subcortical heterotopias.

On MR imaging, the characteristic 3-layer cake (double cortex) appearance is seen with the cortex and subcortical layer of band heterotopia separated from each other by a thin white matter band (Fig. 8). The cortex may be relatively normal or pachygyric [14].

**10. LEMON SIGN**

This sign is seen on transverse sonograms of fetal skull obtained at the level of ventricles. It refers to the loss of normal convex contour of frontal bones, which appear flattened or scalloped (resembling a lemon) (Fig. 9). This sign is seen in cases of spina bifida and is commonly associated with hydrocephalus and Chiari II malformation. It has been proposed that decreased intracranial pressure in cases of spina bifida, leads to flattening or scalloping of vulnerable frontal bones. With fetal maturity, frontal bones become stronger thereby leading to the disappearance of lemon sign. Thus, with an increase in gestational age, lemon sign becomes less reliable in the diagnosis of spina bifida [15].

Significance: This sign is very useful for the diagnosis of spina bifida before 24 weeks of gestation. However, it should be noted that besides spin bifida, this sign has been seen in a number of conditions like encephalocele, Dandy-Walker malformation with encephalocele, thanatophoric dysplasia, cystic hygroma, diaphragmatic hernia, corpus callosum agenesis, etc [16].

**11. BANANA SIGN**

This sign is seen on axial imaging through the posterior fossa of the fetus. It is seen in Chiari II malformation and results from a tight wrapping of cerebellum around brainstem due to small posterior fossa [17]. The cisterna magna gets obliterated and the shape of the cerebellum bears resemblance to a banana (Fig. 10).
12. FROG EYE SIGN

This sign is seen in anencephaly, which is the commonest and most severe form of neural tube defect. Anencephaly is characterized by absent cranial vault and telencephalon. Remnant brain is covered by a vascular membrane. Anencephaly can be detected radiologically as early as at 11 weeks [18]. The “frog eye” appearance results from the absence of cranial bones and brain with bulging orbits and can be seen in coronal ultrasound or MRI image (Fig. 11).

![Fig. (11). Coronal gray scale ultrasound image in a 12 weeks fetus showing absent cranial vault with bulging orbits.](image)

13. TRANSMANTLE SIGN

This sign is seen on MRI, almost exclusively in type II focal cortical dysplasia. The key imaging feature is a high signal on T2 and Fluid Attenuated Inversion Recovery (FLAIR) images extending from the ventricle to the cortex (Fig. 12). The area of abnormal signal intensity is associated with focal dysplasia, which helps in differentiating it from radial band sign of tuberous sclerosis (which is associated with subependymal nodules and cortical tubers). This sign is seen in ~45% (range 21-72%) of patients with type II focal cortical dysplasia [18].

Significance: This sign is not only specific for type II focal cortical dysplasia, it is also associated with excellent post-surgical seizure-free outcomes [19].

14. MARTINI GLASS SIGN

This sign is seen in persistent hyperplastic primary vitreous, which is a congenital developmental malformation of the eye due to incomplete regression of the embryonic ocular blood supply. This manifests as retrolenticular tissue on MRI, resembling the shape of a martini glass (Fig. 13), hence the name [20]. Persistent hyperplastic primary vitreous lacks calcification, which helps to differentiate it from retinoblastoma (another important cause of leukocoria in pediatric population) [20, 21].

15. RADIAL BAND SIGN

This sign is seen on MRI in patients with tuberous sclerosis. Radial bands are best seen on T2 and FLAIR images as
linear or curvilinear areas with an abnormal signal intensity extending from the periventricular region to the subcortical region (Fig. 14). This sign is believed to be indicative of abnormal migration of dysplastic stem cells in patients with tuberous sclerosis complex. It is seen in association with other intracranial manifestations like subependymal nodules, subependymal giant cell astrocytoma, cortical tubers, and white matter abnormalities [22].

Significance: Visualization of radial bands is thought to be specific to tuberous sclerosis.

16. THE TRAM-TRACK SIGN

This sign is seen on skull radiographs or CT as gyriform, curvilinear, parallel calcifications (resembling tram track) in cases of Sturge-Weber syndrome. Sturge-Weber syndrome is a rare neurocutaneous syndrome that includes a facial port-wine stain and is associated with leptomeningeal angiomatosis [23]. The calcifications are often curvilinear and gyriform, commonly involving parietal and occipital lobes (Fig. 15). Pathologically, these calcifications are seen in layer 4 of cortex, adjacent to angiomas and usually seen after two years of age [24].

17. THE FACE OF GIANT PANDA SIGN

This sign is classically seen in Wilson disease on axial MRI images. Wilson disease, an inherited disorder of metabolism, in which defective biliary excretion of copper leads to its accumulation, particularly in the liver and brain [25].

The face of giant panda refers to the appearance of the midbrain, when the red nucleus and substantia nigra are surrounded by a high T2 signal. The preservation of normal signal in red nucleus forms ‘eyes’, preserved signal intensity in the lateral portion of pars reticulata of substantia nigra forms ‘ears’ and hypointensity in the superior colliculus forms the ‘mouth’ (Fig. 16) [25].

18. DAWSON FINGER

Dawson fingers are seen on brain MRI in patients with multiple sclerosis. This appearance results from characteristic perivenular demyelination in multiple sclerosis. The demyelinating plaques are seen as round or ovoid lesions perpendicular to the ventricles (Fig. 17) [8]. Dawson fingers have been named after the Scottish pathologist James Walker Dawson, who described this phenomenon on histopathological specimen [26].

19. OPEN RING SIGN

This sign is seen in contrast-enhanced MRI of the brain in cases of tumefactive demyelinating lesions and is helpful in distinguishing demyelination from other ring-enhancing lesions [8]. It is observed in patients with multiple sclerosis. Large demyelinating lesions show ring enhancement, which is often incomplete or open (Fig. 18). The open portion of
the ring abuts the grey matter of cortex or basal ganglia, as demyelination appears as a crescent involving only white matter [27].

20. WHITE CEREBELLUM SIGN

This sign is seen on CT images and is also called “reversal sign” or “dense cerebellum sign”. It is encountered when there is a diffusely decreased density of the supratentorial brain parenchyma, with relatively increased attenuation of the cerebellum, brainstem and thalami (Fig. 19) [1]. It is seen in severe head injury, birth asphyxia, drowning, status epilepticus, bacterial meningitis, and encephalitis and represents anoxic/ischemic cerebral injury. The possible mechanisms for this sign are distention of deep medullary veins due to obstruction by raised intracranial pressure (resulting in cerebral edema) and relatively preserved blood flow in posterior circulation [28]. This sign indicates irreversible brain damage and has a very poor prognosis. Thus, this striking sign must be remembered for the bleak prognosis it carries.

21. CAPUT MEDUSA SIGN

The caput medusa sign, also known as a palm tree sign, is seen in venous angioma or developmental venous anomaly. Venous angioma is the most common vascular anomaly of brain. The most common site is frontal lobe (40%), followed by posterior fossa (20%) and parietal lobe (15%) [29]. The
sign is seen on both contrast enhanced CT and MRI in which a number of medullary/subcortical veins are seen draining centrally towards a single dilated venous structure (Fig. 20). The appearance is reminiscent of Medusa, a gorgon of Greek mythology [30, 31]. She has been described as a winged human with living venomous snakes in her head in place of hair. Multiple smaller subcortical/medullary veins draining into single larger vein give the appearance of “snakes” converging towards a common point (the “head”), hence the name caput medusa.

![Fig. (19). Axial CT image in a child showing apparently increased attenuation of cerebellum (arrow) against a background of diffuse hypodensity involving bilateral cerebral hemispheres.](image)

![Fig. (20). A: Axial contrast enhanced MRI showing a dilated venous channel (arrow) in right parietal region with adjacent smaller cortical veins. B: Sagittal contrast enhanced MRI showing a large central vein (representing head) with smaller cortical veins converging towards it giving medusa head appearance.](image)

22. POPCORN SIGN

This sign is seen on MRI images in cases of cavernous haemangioma. It refers to a well-defined lobulated lesion with central heterogeneous signal intensity on T1 and T2 weighted images, resembling a popcorn. This sign is considered characteristic of cavernous haemangiomas, and can be seen in both cerebral and spinal lesions [32, 33]. The central heterogeneous area of mixed intensity is formed by thrombosis, calcification, fibrosis and blood breakdown products (Fig. 21). There is also a peripheral hypointense rim representing hemosiderin and iron deposition in adjacent brain parenchyma [32].

![Fig. (21). Axial T2 weighted MRI showing a well defined lobulated lesion in right temporal lobe (arrow) with central mixed heterogeneous signal and peripheral hypointense rim.](image)

23. IVY SIGN

The ivy sign is observed in MRI of patients with moyamoya disease. It can be seen in both the post-contrast T1 and FLAIR images. On FLAIR images, it is seen as hyperintensity in sulcal spaces due to a slow arterial flow in leptomeningeal collaterals. On post-contrast images, it manifests as diffuse leptomeningeal enhancement (Fig. 22), which represents fine neovascularization over the cortical surface [34]. This appearance resembles an ivy plant creeping on the stones, hence the name. However, leptomeningeal enhancement seen in moyamoya disease, should be distinguished from meningitis and meningeal carcinomatosis [34].

24. PERIPHERAL RIM SIGN

This sign has recently been described in cases of Dysembryoplastic Neuroepithelial Tumor (DNET) and is seen on FLAIR MRI images. It is characterized by a peripheral well-defined hyperintense rim on FLAIR images which may be complete or incomplete (Fig. 23). It has been suggested that this rim might represent loose neuroglial elements in the periphery [35]. This sign helps in differentiating DNET from other cortical- based tumors like ganglioglioma and pleomorphic xanthoastrocytoma.
25. BRACKET CALCIFICATION SIGN

This sign is seen on CT images in cases of pericallosal lipoma. Pericallosal lipomas are thought to result from abnormal persistence and differentiation of meninx primitive [36]. Pericallosal lipoma appears as a well-defined fat attenuation lesion on CT which may show peripheral curvilinear calcification resembling bracket symbol (Fig. 24) [37].

CONCLUSION

The identification of a classic sign in pediatric neuroradiology allows for a more confident diagnosis. The radiologist should be familiar not only with the appearance of these signs but also with basic pathophysiology behind these signs for better understanding. Radiologists are urged to read and remember these signs so that when encountered, they can be recalled and subsequently reported.
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