Animals in the brain!

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Learning objectives

1. To discuss and illustrate the radiologic signs of a number of diverse conditions whose classic neuro-radiologic manifestations resemble various types of animals, insects and birds.

2. To improve understanding of the underlying causes for these amusing imaging signs.

Background

Neuroradiology remains one of the most intricate subspecialties of diagnostic radiology and the neuroradiology literature is replete with a variety of interesting radiologic signs.

The successful interpretation of neuroimaging studies requires the recognition and understanding of these distinguishing radiologic signs.

Certain neurologic conditions have characteristic radiologic manifestations that resemble various types of animals, birds and insects and their different body parts.

These "animal signs" are highly memorable and easily recognizable.

It is important that radiologists recognize these classic signs and thus clinch the apt diagnosis based solely on the imaging findings.

Imaging findings OR Procedure details

This educational exhibit describes several important "Animal" signs that are useful in diagnosing various diseases affecting the cranium, including:

1. Eye of the Tiger sign: *Hallervorden Spatz disease*
2. Face of the Giant panda: *Wilson's disease*
3. Giant panda cubs: *Wilson's disease*
4. Owls eye appearance: *Central pontine myelinolysis*
5. Face of the Piglet: *Central pontine myelinolysis*
6. Monkey sign: *Central pontine myelinolysis*
7. Butterfly sign: *Butterfly glioma (Glioblastoma multiforme)*
8. Bulls eye sign: Cerebral Toxoplamosis
9. Bulls eye or button sequestrum: Eosinophilic granulomas
10. Bat wing fourth ventricle: Joubert syndrome
11. Bat wing sylvian fissure: Glutaric aciduria type-1
12. Leopard skin or Tigroid pattern: Metachromatic leukodystrophy
13. Tiger skin pattern: Lhermitte Duclos disease
14. Tail sign: Meningioma
15. Horse shoe enhancement: Tumefactive demyelination
16. Steer horn ventricles: Corpus callosal agenesis
17. Tectal beaking: Arnold Chiari malformation
18. Zebra sign: Remote cerebellar hemorrhage
19. Rodent facies: Untreated Cooley's anemia

COMMENTARY

1) "Eye-of-the-Tiger" sign is most commonly described in Hallervorden-Spatz syndrome, and refers to symmetrical low signal intensity circumscribing a central region of high signal intensity in the globus pallidus on T2-weighted MR images (Fig. 1, 2). The ring of marked hypointensity involving the globus pallidus on T2-weighted MR images is due to excess iron accumulation and the central high signal intensity is attributed to gliosis, increased water content, and neuronal loss. This appearance, however, is not pathognomonic of Hallervorden-Spatz syndrome, and has been reported in other extrapyramidal parkinsonian disorders, including cortical-basal ganglionic degeneration, early onset levodopa-responsive parkinsonism, Steele-Richardson-Olszewski syndrome (progressive supranuclear palsy) and multiple system atrophy [1,2].

2-3) "Face of the Giant Panda" is an uncommon intracranial manifestation of Wilson's disease which was first described by Hitoshi et al in 1991. This appearance is caused by a combination of signal intensity changes at the level of midbrain on T2-weighted MR images. These include: high signal intensity in the tegmentum, normal signals in the red nuclei and lateral portion of the pars reticulata of the substantia nigra, and hypointensity of the superior colliculus (Fig. 3, 4). The exact pathogenesis of this constellation of MR findings is not known, but it is postulated that the paramagnetic effects of the deposition of heavy metals, such as iron and copper, may be responsible. It is believed that iron is assumed to play a more important role than copper in reducing the signal intensity of the superior colliculi on the T2-weighted scan. At times, signal alteration may also be encountered within the dorsal pons which has been popularly called as "Giant Panda Cubs" (Fig. 5, 6) [3].

4-6) Central pontine myelinolysis (CPM) is a non-inflammatory demyelinating disease of the white matter tracts traversing the pons. Since the peripheral pontine fibers are typically spared it is referred to as central pontine myelinolysis. The predominant
involvement of the transverse pontine fibres and relative sparing of the descending corticospinal tracts results in a characteristic "Owls-eye" appearance on axial T2-weighted MR imaging (Fig. 7, 8). If the areas of demyelination coalesce, axial T2-weighted MR images resemble "Face of the Piglet". This sign was first reported by Wagner et al. The pons with its characteristic appearance resembles the snout, while the internal carotid arteries and the fourth ventricle constitute the eyes and mouth of the piglet respectively (Fig. 9, 10). Corresponding T1-weighted MR images may show this characteristic pattern of signal alteration in the basal pons as resembling the face of a monkey - also referred to as the "Monkey sign" of CPM (Fig. 11) [4-7].

7) "Butterfly Glioma" refers to a high grade astrocytoma, usually a glioblastoma multiforme, which crosses the midline via the corpus callosum. The term butterfly refers to the symmetric wing like extensions across the midline (Fig. 12, 13). Mostly butterfly gliomas occur in the frontal lobes crossing via the genu of the corpus callosum, but posterior butterfly lesions (crossing through the splenium) are also encountered. The differential diagnoses include primary cerebral lymphoma and tumefactive demyelination [8].

8) The "Target sign" or "Bull's eye" is a favourite radiologic descriptor of everything from bowel ntussusception to cerebral tuberculoma. CT target sign, although uncommon, is considered nearly pathognomonic for cerebral tuberculoma when it is seen. The three-zone T2-weighted target (bull's eye) sign at MR imaging has not been described in any other condition other than Cerebral Toxoplasmosis [9]. The T2-weighted target or bull's eye sign is characterized by a hypointense core, an intermediate hyperintense region, and a peripheral hypointense rim (Fig. 14, 15).

9) The classic "Bull's eye" (or button) sequestrum sign is caused by a lucent bone lesion with a central opacity and is an uncommon manifestation of many diseases [10]. Initially described on radiographs, this sign can also be observed on CT scans. Originally, the button sequestrum sign was used to describe a radiographic manifestation of eosinophilic granuloma in the bone, which is the mildest form of Langerhans cell histiocytosis. The disease affects children and young adults and has a male predilection. However, this sign is not specific for a single disease. The central opacity or the 'button sequestrum' represents an island of dead bone and the adjacent lucent area may result from a number of processes. In cases of osteomyelitis, the lucent area is caused by infectious organisms that destroy the bone, which is then replaced by purulent material and granulation tissue. In cases of eosinophilic granuloma, the lucent lesion represents an erosive accumulation of histiocytes (Fig. 16, 17). This sign has also been described in neoplastic conditions such as fibrosarcoma (including desmoplastic fibroma and malignant fibrous histiocytoma), and lymphoma.
10) Joubert syndrome is an autosomal recessive disorder where there is agenesis of the cerebellar vermis to varying degrees. There is an absence of crossing of the fibers of the superior cerebellar peduncles and pyramidal tracts. Axial MR images of brain demonstrate absence of the cerebellar vermis, with apposition (but no fusion) of the cerebellar hemispheres. The characteristic imaging findings of Joubert syndrome include "Bat-wing fourth ventricle" (Fig. 18, 19) and prominent thickened elongated superior cerebellar peduncles giving rise to a 'molar tooth appearance'. The absence of the vermis is responsible for a triangular-shaped mid-fourth ventricle and a bat-wing shaped fourth ventricle superiorly [12].

11) "Batwings dilatation" of sylvian fissures and wide CSF spaces anterior to temporal lobes in a child should alert the radiologist of the possibility of Glutaric acidemia type-I. Glutaric acidemia type-I is a rare autosomal recessive disorder caused by the deficiency of a mitochondrial enzyme glutaryl-CoA dehydrogenase. This enzyme deficiency is responsible for the improper breakdown of the aminoacids resulting in an elevated plasma and urine level of glutaric acid. Glutaric acid accumulation in the body is responsible for neurotoxicity in the basal ganglia and fronto-temporal cortex. Neuroimaging serves as a useful tool, many a times providing the first clue to the diagnosis. The most striking imaging finding is the presence of very wide CSF spaces anterior to the temporal lobes and along the sylvian fissures which has been likened to 'wings of a bat' (Fig. 20, 21). Although the finding of wide opercula suggests glutaric acidemia, the finding is by no means specific, as this imaging appearance can also be encountered in severe developmental delay and idiopathic external hydrocephalus. But, if the 'Batwings dilatation' of sylvian fissure is associated with concomitant basal ganglia lesions, it is considered almost pathognomonic of glutaric acidemia type-I [13, 14].

12) Metachromatic leukodystrophy (MLD) is an autosomal recessive disorder caused by a deficiency of the lysosomal enzyme arylsulfatase. The decreased activity of arylsulfatase enzyme accounts for failure of myelin breakdown and reutilization, thus resulting in dysmyelination. At T2-weighted MR imaging, MLD manifests as symmetric confluent areas of high signal intensity in the periventricular white matter typically sparing the subcortical U fibers during the early stages. The sparing of the perivascular white matter within the periventricular white matter and centrum semiovale is responsible for the characteristic "Tigroid" and "Leopard skin" pattern (Fig. 22, 23). Besides MLD, tigroid and leopard skin pattern of dysmyelination can also be present in Pelizaeus#Merzbacher disease which usually manifests in the neonatal period or early infancy [15].

13) A "Tiger-striped" cerebellar foliar pattern that consists of alternating bands on T1- and T2-weighted images is considered almost pathognomonic of Lhermitte-Duclos disease. The maintenance of the overall cerebellar architecture in spite of the thickened, and hyperplastic folia is responsible for this characteristic imaging appearance (Fig. 24, 25). Lhermitte-Duclos disease (LDD) (also known as dysplastic gangliocytoma) is a
rare probably hamartomatous disorder involving the cerebellum. The debate, whether it represents a neoplastic, malformative or hamartomatous lesion, still continues. It usually manifests in adults, mostly in the third and fourth decades with worsening headache, unsteady gait, ataxia and cranial nerve palsy. Gross autopsy specimen's show expanded cerebellar folia usually confined to one side of the cerebellum. The abnormal tissue occasionally extends into the vermis and rarely into the contralateral cerebellar hemisphere. CT findings are mostly non-specific demonstrating a hypoattenuating or a mixed-attenuation cerebellar mass. MRI is the imaging modality of choice, usually sufficient for the diagnosis of this condition [16].

14) The **Dural "Tail" sign** is seen on contrast material-enhanced MR images as a thickening of the enhanced dura mater that resembles a tail extending from a mass (Fig. 26, 27). It was initially thought to result from direct invasion of the dura; however subsequent studies demonstrated it to be more a reactive process. It was described by Wilms and colleagues as a thin, tapering rim of dural enhancement, in continuity with a convexity meningioma. Goldsher and associates subsequently, in 1990, demonstrated the dural tail sign in 60% of their series of meningiomas and concluded that its presence, associated with an extraxial mass, was very specific for meningioma. The dural tail sign more recently has been described in patients having chloroma, lymphoma, sarcoidosis, acoustic neuroma, and metastatic tumors [17].

15) Up to 50 percent of tumefactive demyelinating lesions demonstrate abnormal contrast enhancement, often in the form of ring enhancement. Commonly the enhancement pattern is in the form of an **"open ring (Horse-shoe enhancement)"**, with the incomplete portion of the ring facing the cerebral cortex (Fig. 28, 29). The enhancing segment of the ring is thought to represent the zone of active demyelination accordingly favouring the white matter side of the lesion. The nonenhancing central part represents a more chronic phase of demyelination. Tumefactive demyelinating plaques are mostly well circumscribed and demonstrate high T2 and low T1 signal. Helpful diagnostic features include disproportionately less mass effect and vasogenic oedema for the size of the lesions [18, 19].

16) **"Steer-horn" lateral ventricles** refer to the abnormal shape of the frontal horns of the lateral ventricle on coronal MR images in patients with corpus callosum agenesis (Fig. 30, 31). The lack of supporting deep white matter fibers and associated redirection of longitudinal callosal fibers (Probst bundles), result in alteration in the configuration of the ventricles, with the frontal horns taking on a "steer or bull’s horn" appearance in the coronal plane [20].

17) **Tectal "Beaking"** refers to the triangular ("beaked") appearance of the tectum in Arnold Chiari type-II malformation (Fig. 32, 33). The tectum is abnormal in virtually all patients with Chiari -II malformation. The quadrigeminal plate is partially or completely
fused. The midbrain is elongated caudally and posteriorly to overlie the midline cerebellum and pons. Variable degrees of fusion of the colliculi and tectum result in a triangular ("beaked") tectum.

18) The "Zebra" sign refers to the streaky pattern of hemorrhage along the cerebellar folia in patients with remote cerebellar hemorrhage (RCH). Remote cerebellar hemorrhage or cerebellar hemorrhage distant from the site of surgery is a rare, usually benign, complication that most often occurs after supratentorial craniotomy. It is a rare but benign, self-limited complication of supratentorial craniotomies. RCH can be an unexpected finding on routine postoperative imaging studies and should not be mistaken for more ominous causes of bleeding such as coagulopathy, hemorrhagic infarction, or cortical vein occlusion. The cause of RCH is unknown, but it is thought to result from CSF volume depletion leading to cerebellar sagging, with consequent occlusion of the bridging veins and resultant hemorrhage. Remote cerebellar hemorrhage typically appears as streaky curvilinear areas of increased attenuation (on CT) or dark stripes (at T2-weighted or T2*gradient echo MR images) in the cerebellar sulci and folia, a finding that has been called the "zebra sign" (Fig. 34, 35) [22].

19) "Rodent facies" refers to the orofacial manifestations of thalassemia major which include abnormally prominent maxillary and cheek bones and a depressed nasal bridge (Fig. 36, 37). Erythroid hyperplasia and extramedullary haematopoiesis in thalassemic patients results in hypertrophy of osseous structures and a consequent prominence of the malar eminences. Proliferation of marrow within the frontal and facial bones impedes pneumatisation of the paranasal sinuses. Radiographs and CT examination show dilatation of marrow spaces with coarse osseous trabeculations. A generalised loss of bone density accompanies thinning of the cortex of maxillary and mandibular bones with obliterated paranasal sinuses [23].

20) "Penguin" sign on brain MRI is an interesting radiological sign seen in patients with progressive supranuclear palsy (PSP). It refers to atrophy of the midbrain tegmentum, with a relatively preserved pons on midsagittal T1-weighted images (Fig. 38, 39). The "penguin" sign can be helpful in distinguishing progressive supranuclear palsy from multisystem atrophy and Parkinson disease. Patients with Parkinson disease, multisystem atrophy & corticobasal degeneration have no midbrain atrophy & hence do not show this sign. The penguin sign is useful for establishing the diagnosis of PSP; and is reported to have a sensitivity of nearly 100% [24, 25].

Images for this section:
**Fig. 1:** Eye of the Tiger sign

**The Eye of the Tiger sign**

- This refers to symmetrical low signal intensity circumscribing a central region of high signal intensity in the globus pallidus on T2-w MRI [1, 2].

- The central hyperintensity is possibly due to tissue necrosis, and oedema.

- The surrounding hypointensity, due to abnormal iron deposition.

**Fig. 2:** Eye of the Tiger sign
Fig. 3: Face of the Giant panda

Fig. 4: Face of the Giant panda

Face of the Giant Panda sign

- This MR appearance is seen due to a combination of high signal intensity in the tegmentum (black arrow) with sparing of the red nuclei (dotted white arrow), pars reticulate (white arrow) and the superior colliculi (black arrowhead) giving rise to the typical ‘Face of the giant panda’ sign [3].
**Giant Panda cub:** Wilson’s disease

![Giant panda cub image](image)

**Fig. 5:** Giant panda cubs

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**Giant Panda cub**

- At times, signal alteration is also seen within the dorsal pons in patients with Wilson’s disease.

- This has been popularly called as ‘Giant panda cub’ [3].

- It is believed that iron is assumed to play a more important role than copper in reducing the signal intensity on the T2-weighted scan.

**Fig. 6:** Giant panda cubs
Fig. 7: Owls eye appearance

Fig. 8: Owls eye appearance

- Central Pontine Myelinolysis affects the basal pons with sparing of the descending corticospinal tracts as well as the peripheral pontine tissue.
- The corticospinal tracts may appear as preserved islands within a zone of hyperintense pontine demyelination giving an Owl's eye appearance [4].
**Face of the Piglet sign:**
Central Pontine Myelinolysis

**Fig. 9:** Face of the Piglet

The pons with its characteristic appearance resembles the snout, ICAs and the fourth ventricle constitute the eyes and mouth of the piglet respectively [5, 6].

**Fig. 10:** Face of the Piglet
**Monkey sign:** Central Pontine Myelinolysis

- Corresponding T1-weighted MR images may show this characteristic pattern of signal alteration in the basal pons as resembling the face of a monkey - also referred to as the ‘Monkey sign’ of CPM [7].

**Fig. 11:** Monkey sign

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**Butterfly pattern:** Glioblastoma Multiforme (GBM)

**Fig. 12:** Butterfly sign
**Butterfly pattern: GBM**

- GBMs typically cross white matter tracts (corpus callosum) to involve contralateral hemisphere.

- Thus displaying a characteristic bihemispheric involvement or a ‘butterfly-pattern’ - hence the term ‘butterfly glioma’.

**Fig. 13:** Butterfly sign

**Bulls eye sign: Cerebral toxoplasmosis**

**Fig. 14:** Bulls eye sign
**Bulls eye or Target sign:** Cerebral toxoplasmosis

- Bulls eye appearance (or target sign) is a three-zone target sign with a hypointense core, an intermediate hyperintense region, and a peripheral hypointense rim [9].

- The surrounding oedema helps to delineate the outermost iso to hypointense ring.

**Fig. 15:** Bulls eye sign

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**Button (Bulls eye) sequestrum:**
Eosinophilic Granuloma

**Fig. 16:** Bulls eye (button)sequestrum
**Button (Bulls eye) sequestrum:**

Eosinophilic Granuloma

- The button sequestrum sign is caused by a lucent bone lesion with a central opacity displaying a ‘Bulls eye appearance’ [10].

- The central opacity represents an island of dead bone.

- In cases of eosinophilic granuloma, the lucent lesion represents an erosive accumulation of histiocytes. In cases of osteomyelitis, the lucent area represents destroyed bone which is replaced by purulent material and granulation tissue [11].

**Fig. 17:** Bulls eye (button) sequestrum

**Bat wing fourth ventricle:**

Joubert syndrome

**Fig. 18:** Bat wing fourth ventricle
Bat wing fourth ventricle:
Joubert syndrome

- Joubert syndrome is an autosomal recessive disorder associated with agenesis of the cerebellar vermis to varying degrees.

- Absence of the vermis results in a bat-wing shaped fourth ventricle [12].

**Fig. 19:** Bat wing fourth ventricle

Bat wing sylvian fissure:
Glutaric acidemia type-I

**Fig. 20:** Bat wing sylvian fissure
**Bat wing sylvian fissure:**
Glutaric acidemia type-I

- Predominant frontal and temporal cerebral atrophy in Glutaric Acidemia Type-I result in widely dilated opercula which have been likened to 'wings of a bat' [13, 14].

**Fig. 21:** Bat wing sylvian fissure

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**Tigroid or Leopard skin pattern:**
Metachromatic Leukodystrophy

**Fig. 22:** Leopard skin or Tigroid pattern
Tigroid or Leopard skin pattern: Metachromatic Leukodystrophy

- The sparing of the perivascular white matter within the centrum semiovale and periventricular white matter is responsible for the characteristic ‘tigroid’ and ‘leopard skin’ pattern [15].

**Fig. 23:** Leopard skin or Tigroid pattern

Tiger striped foliar pattern: Lhermitte-Duclos disease

**Fig. 24:** Tiger skin foliar pattern
**Tiger striped foliar pattern:**
Lhermitte-Duclos disease

- A characteristic corduroy or tiger-striped cerebellar foliar pattern is typical of Lhermitte-Duclos disease [16].
- Maintenance of the overall cerebellar architecture in spite of the thickened, and hyperplastic folia is responsible for this characteristic and pathognomonic appearance.

**Fig. 25:** Tiger skin foliar pattern

**‘Tail’ sign:** Meningioma

**Fig. 26:** Tail sign
Dural Tail sign: Meningioma

- The dural tail sign is seen on contrast material–enhanced MR images as a thickening of the enhanced dura mater that resembles a tail extending from a mass [17].

Fig. 27: Tail sign

‘Horse shoe’ or Incomplete ring enhancement: Tumefactive Demyelination

Fig. 28: Horse shoe enhancement pattern
‘Horse shoe’ or Incomplete ring enhancement:
Tumefactive Demyelination

- The incomplete portion of the ring faces the cerebral cortex.
- The enhancing (horse-shoe) segment represents the zone of active demyelination, and therefore favours the white matter side [18, 19].

**Fig. 29:** Horse shoe enhancement pattern

**Steer horn ventricles:** Corpus Callosum Agenesis

**Fig. 30:** Steer horn ventricles
**Steer horn ventricles:** Corpus Callosum Agenesis

- Without the supporting deep white matter fibers, there is alteration in the configuration of the ventricles, with the frontal horns taking on a “steer-horn” appearance in the coronal plane [20].

**Fig. 31:** Steer horn ventricles

**Tectal beaking:** Arnold Chiari type-II

**Fig. 32:** Tectal beaking
**Tectal beaking:** Arnold Chiari type-II

- Variable degrees of fusion of the colliculi and tectum result in prominent beaking of the tectum and inferior displacement of the tectal plate [21].

**Fig. 33:** Tectal beaking

**Zebra sign:** Remote Cerebellar Hemorrhage

**Fig. 34:** Zebra sign
Zebra sign: Remote Cerebellar Hemorrhage

- Remote cerebellar hemorrhage typically appears as streaky curvilinear areas of increased attenuation (on CT) or dark stripes (at T2-w or T2*-GRE MR images) in the cerebellar sulci and folia, a finding that has been called the “zebra sign” [22].

Fig. 35: Zebra sign

Rodent facies: Thalassemia major

Fig. 36: Rodent facies
Rodent facies: Thalassemia major

- The best known of orofacial manifestations of thalassemia major are prominent maxillary and cheek bones with depression of the nasal bridge often referred to as ‘rodent facies’ [23].

Fig. 37: Rodent facies

Penguin sign: Progressive Supranuclear Palsy

Fig. 38: Penguin sign
**Penguin sign:** Progressive Supranuclear Palsy

- Sagittal T1-w image shows atrophy of the midbrain, with preservation of the volume of the pons. This appearance has been called the “penguin sign” [24, 25].
- There is also atrophy of the tectum, particularly the superior colliculi.

**Fig. 39:** Penguin sign
Conclusion

Knowledge of these amusing, attention-grabbing and easily decipherable 'animal' based signs not only helps to narrow down the elaborate list of differentials but can also facilitate the diagnosis on the basis of imaging findings alone.

Personal Information

References