Pontine pathology: narrowing the radiological differential

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Learning objectives

1. Identify common pathologies that affect the pons.
2. Describe the magnetic resonance imaging (MRI) features of these conditions.

Background

The pons, part of the metencephalon of the hindbrain, bridges the two other parts of the brainstem: the medulla oblongata and the midbrain. The pons contains tracts of white matter fibers carrying signals between the spinal cord and the rest of the brain. It also houses the motor and sensory nuclei of the trigeminal nerve (CN V), the motor nucleus of the abducens nerve (CN VI), the nuclei of the facial nerve (CN VII) and vestibular nerve (CN VIII). It also contains other important structures such as the reticular formation responsible for breathing rhythms.

A range of pathologies can affect the adult pons including neoplasia, inflammatory disease, infection, vascular abnormalities and neurodegenerative disease. Clues regarding the underlying aetiology can be obtained by detailed imaging assessment and identification of specific features.

The MRI characteristics of pontine pathologies that commonly affect the adult pons scanned at a large tertiary neuroscience centre are reviewed in combination with the clinical details.

Findings and procedure details

NEOPLASIA

· Glioblastoma multiforme (GBM)

GBM is a WHO grade IV lesion; the most aggressive glioma. It is the most common primary brain tumour, representing between 12-15% of all intracranial neoplasms. GBM commonly affects cerebral hemispheres and rarely affects the brainstem alone. Radiological appearance of GBM is of a poorly marginated mass with mixed intensity
on T1WI. T2/FLAIR show heterogeneous hyperintensity with indistinct tumor margins and surrounding vasogenic edema. Necrosis, cysts, hemorrhage at various stage of evolution, fluid/debris levels and "flow voids" from extensive neovascularity may be seen. T2* imaging often shows foci of susceptibility artifact and T1 post gadolinium images show inhomogeneous enhancement (Fig. 1 on page 7, Fig. 2 on page 8, Fig. 3 on page 9, Fig. 4 on page 10, Fig. 5 on page 11).

In contrast to adults, diffuse pontine gliomas (DPG) in the pediatric age group are more common, constituting between 10-15% of all childhood brain tumors and are the main cause of death in this group. DPG tend to infiltrate and expand the pons giving a "fattened" pontine appearance. They appear hyperintense on T2WI/FLAIR. Any enhancement is suspicious for anaplasia (Fig. 6 on page 12, Fig. 7 on page 13).

INFLAMMATORY DISEASE

• Langerhans cell histiocytosis (LCH)

LCH is a non-neoplastic granuloma-like lesion of unknown origin; it is characterized by proliferation of reticulohistiocytic structures, polynuclear eosinophils, neutrophils, lymphocytes, plasma cells, multinucleate giant cells and Langerhans cells. Cerebellar involvement in LCH is infrequent compared to the more common enhancing granulomatous infiltrates that typically affect the hypothalamus and pituitary stalk. Secondary cerebellar degeneration can occur in 25% of cases and is seen as confluent T2/FLAIR hyperintensities with varying enhancement (Fig. 8 on page 14, Fig. 9 on page 15). A differential for this cerebellar pattern would include Juvenile Xanthogranuloma but this condition preferentially affects young children.

• Chronic Lymphocytic Inflammation with Pontine Perivascular Enhancement Responsive to Steroids (CLIPPERS)

CLIPPERS is a recently defined inflammatory central nervous system (CNS) disorder, predominantly involving the brainstem and in particular the pons. The condition features a combination of clinical symptoms essentially referable to brainstem pathology and a characteristic magnetic resonance imaging appearance with punctuate and curvilinear gadolinium enhancement "peppering" the pons (Fig. 10 on page 16, Fig. 11 on page 17, Fig. 12 on page 18, Fig. 13 on page 19, Fig. 14 on page 20).

• Behçet Disease (BD)

Behçet Disease is a chronic idiopathic, relapsing-remitting, multisystem, inflammatory vascular disease that is characterised mainly by mucocutaneous recurrent oral and
genital ulcers, aphthous stomatitis, ophthalmologic lesions such as uveitis and iridocyclitis and multiple arthralgia. The central nervous system is involved in 20-25% of patients. When BD occurs in the CNS, it is termed neuro-Behçet disease (NBD). Although any part of the CNS can be affected, brainstem involvement, especially of the cerebral peduncles, is typical and occurs in 50% of cases. Typical MR findings are small circular, linear, crescent-shaped, or irregular foci of T2/FLAIR hyperintensity in the midbrain. During the acute phase, the pons can be enlarged with diffuse signal abnormality, resembling neoplasia. Ill-defined patchy enhancement is common; strong uniform enhancement is rare (Fig. 15 on page 22, Fig. 16 on page 22, Fig. 17 on page 23). There may be radiological improvement following therapy. Long term sequelae of the condition can include neuronal loss and atrophy (Fig. 18 on page 24).

- **Neuromyelitis optica (NMO)**

Neuromyelitis Optica (NMO) previously known as Devic's disease is a severe demyelinating condition typically characterised by relapses of optic neuritis +/- myelitis. This may be coupled with lesions in specific areas of the brain, mainly the hypothalamus, periventricular nucleus, and brainstem. With pontine involvement the appearance is generally one of ill-defined T2 signal hypertense foci also incorporating the middle cerebellar peduncles (Fig. 19 on page 25, Fig. 20 on page 26, Fig. 21 on page 27). Concomitant involvement of the optic nerve or a contiguous spinal cord lesion (> 3 vertebral segments) aids the diagnosis. Aquaporin-4/NMO-IgG seropositivity should be investigated.

- **Multiple sclerosis (MS)**

MS is the most frequent demyelinating pathology of the CNS and it is characterized histopathologically by multiple inflammatory demyelinating foci called "plaques". MS is a complex, multifactorial disease whose precise pathogenesis remains unknown. Most MS plaques are supratentorial (although infratentorial lesions are relatively more common in children) and vary in size and number. Onset typically occurs in young to middle-aged adults from 20-40 years of age. Clinical presentation varies with heterogenous neurological manifestations, evolution and disability. Over 95% of patients with clinically definite MS have a positive finding on MR imaging. The most recent revised McDonald criteria for MS diagnosis rely on MR imaging to demonstrate dissemination in both space and time (see below).

**REVISED McDonald Criteria for MS Diagnosis**

**Dissemination in Space**

- #1 T2 hyperteintense lesion(s)
In at least 2 of the following 4 areas

1. Priveentricular
2. Juxtacortical
3. Infratentorial
4. Spinal cord

**Dissemination in Time**

- *Either* new T2 Gd-enhancing lesion(s) on follow-up MR
- *Or* simultaneous presence of
  1. Asymptomatic Gd-enhancing and
  2. Nonenhancing lesions at any time

With pontine involvement, lesions vary from indistinct T2 hyperintensity change possibly incorporating the middle cerebellar peduncles to focal lesions with precise symptomatology (Fig. 22 on page 28).

**INFECTION**

Brainstem abscess is rare and can carry high mortality. In most cases the suppurative process results from direct extension from a contiguous focus of infection or haematogenous spread from a distant source. Clinical presentation includes lethargy, headaches, vomiting and gait disturbance. Typical MRI appearances are of a (or multiple) ring enhancing lesion with central restricted diffusion (Fig. 23 on page 29, Fig. 24 on page 30, Fig. 25 on page 31, Fig. 26 on page 32).

**VASCULAR ABNORMALITIES**

- **Acute bilateral pontine infarct**

Pontine infarcts are one form of brainstem infarction involving the posterior circulation. Hypertension, diabetes and atrial fibrillation are the major risk factors. Regarding the lesion boundaries, there are five mains clinical presentations that depends on the territories of intrinsic pontine arteries: anteromedial pontine syndrome (motor deficit with
dysarthria and ataxia), anterolateral pontine syndrome (motor and sensory deficits), tegmental pontine syndrome (mild motor deficits and associated with sensory syndromes, eye movement disorders and vestibular symptoms including vertigo, dizziness and ataxia), bilateral pontine syndrome (transient consciousness loss, tetraparesis, and acute pseudobulbar syndrome), and unilateral multiple pontine infarcts associated with sensory-motor deficits and tegmental signs.

Due to the vascular anatomy, pontine infarcts will obey the midline allowing them to be differentiated from neoplastic, inflammatory or infective aetiology (Fig. 27 on page 33, Fig. 28 on page 34).

**Pontine small vessel disease**

Small vessel disease is a common aging phenomenon that is exacerbated by hypertension, diabetes mellitus, atherosclerosis and atrial fibrillation. It is associated with significant vascular risk factors leading to stroke, dementia, motor impairment and possibly Parkinsonism. Small vessel disease appears as ill-defined T2 signal intensity change often without a set pattern. It is uncommon to have isolated pontine disease - concomitant supratentorial involvement and/or focal infraction allows differentiation from mimicking conditions such as central pontine myelinolysis (Fig. 29 on page 35, Fig. 30 on page 36, Fig. 31 on page 37).

**Pontine cerebral cavernoma malformations**

Cerebral cavernomous malformations (CCM) are clusters of abnormal blood vessels mainly found in the brain and spinal cord. They account for 2-17% of CNS vascular malformations and are characterised by repeated intralesional haemorrhage into thin-walled, angiogenically immature, blood-filled locules called "caverns". Cavernomas are often asymptomatic and when symptoms are present they depend on the location and the size of the lesion. The classical CCM appearance on MRI is a discrete reticulated or "popcorn ball" lesion caused by blood products contained within the variably sized "caverns" or "locules". Fluid-fluid levels of different signal intensities are common. The mixed signal core is surrounded by a complete hemosiderin rim on T2WI that "blooms" on T2* sequences. CCMs with subacute haemorrhage are hyperintense on T1WI and mixed hyper-/hypointense on T2WI. They are frequently associated with a developmental venous anomaly (Fig. 32 on page 38, Fig. 33 on page 39, Fig. 34 on page 40).

**DEGENERATIVE AND METABOLIC**

**Multiple system atrophy (MSA)**

MSA is a sporadic neurodegenerative Parkinson-plus disorder characterised clinically by any combination of Parkinsonism, dysautonomia and cerebellar dysfunction, and
pathophysiologically by cell loss, gliosis, and glial cytoplasmic inclusions in several CNS structures. The typical appearance of the "hot cross bun" sign, referring to a cruciform hyperintensity in the pons found on a T2-weighted MRI is highly suggestive of MSA. Additional imaging features such as hyperintensity of putamen rim, putamen hypointensity, putamen atrophy can improve sensitivity and specificity (Fig. 35 on page 41, Fig. 36 on page 42).

• Central pontine myelinolysis (CPM)

CPM is a demyelinating disease that affects the central portion of the base of the pons. CPM occurs as a result of severe underlying disorders. It is most frequently caused by rapid correction of hyponatriemia and can be associated with chronic alcoholism, hepatic cirrhosis, tumour disease, cerebral infarction, acute porphyria and AIDS. Clinical manifestations can include lethargy, hypotension, spastic paraparesis, tetraparesis of various degrees of severity, ataxia, psychiatric changes, pseudobulbar palsy and lock-in syndrome.

Nowadays, the clinical tentative diagnosis is usually confirmed by MRI, which is superior to CT especially in early diagnosis. MRI shows a classic trident shape area of prolonged T1 and T2 in the central pons (Fig. 37 on page 43).

Images for this section:
Fig. 1: Axial T2 of a 23 year old gentleman with pontine GBM and showing pontine expansion with heterogenous T2 hyperintense signal abnormality extending into the middle cerebellar peduncles, predominantly on the left.

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Fig. 2: Axial T1 showing intrinsic hyperintense focus consistent with tumour haemorrhage.

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**Fig. 3:** B1000 DWI showing hyperintense signal within the tumour with corresponding low ADC signal (Fig 4) indicating foci of restricted diffusion - implying cellular hyperdensity and higher tumour grade.

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Fig. 4: ADC map showing restricted diffusion as low intensity foci

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Fig. 5: Axial T1 post gadolinium demonstrating intratumoural enhancement implying high grade neoplasia.

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Fig. 6: Axial T2 of a 3 year old boy showing diffuse signal hyperintensity and expansion of the pons reflecting DPG.

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Fig. 7: Sagittal T1 post gadolinium revealing inhomogeneous enhancement implying higher grade components to the DPG.

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Fig. 8: Ax T2 of a 19 year old male with LCH showing diffuse symmetric signal hyperintensity change in the cerebellar hemispheres and extending to involve the middle cerebellar peduncles and pons.

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Fig. 9: Ax T1 post gadolinium showing minimal patchy enhancement.

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**Fig. 10:** Axial T2 of a 23 year old female with CLIPPERS showing diffuse pontine signal abnormality but with additional involvement of the middle cerebellar peduncles, especially on the right.

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Fig. 11: B1000 DWI demonstrates a few foci of restricted diffusion, particularly in the right middle cerebellar peduncle.

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**Fig. 12:** ADC map shows low signal corresponding to restricted diffusion hyperintense foci on B1000.

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**Fig. 13:** Axial T1 volume post gadolinium reveals several foci of abnormal enhancement.

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**Fig. 14:** Sagittal T2 through the cervical and upper thoracic spine showing abnormal pons but normal cord, allowing some differentiation from neuromyelitis optica.

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**Fig. 15:** Axial FLAIR of a 43 year old female with an acute presentation of neuro-BD showing diffuse pontine swelling with involvement of the middle cerebellar peduncles.

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Fig. 16: Axial T1 post gadolinium showing inhomogeneous enhancement within the pons and middle cerebellar peduncles.

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Fig. 17: Axial FLAIR through the cerebral cortex of the same patient with neuro-BD revealing multiple punctate white matter lesions reflecting the spectrum of neuro-BD.

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**Fig. 18:** Axial T2 of the same patient shown in Figures 15-17 following high dose steroid therapy showing residual signal abnormality and parenchymal volume loss.

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**Fig. 19:** Axial T2 of a 38 year old female with NMO showing ill-defined signal intensity change in the pons and middle cerebellar peduncles, which alone is non-specific but when investigated with orbital and spinal assessment shows the typical findings (Fig 20-21).

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**Fig. 20:** Coronal T1 fat saturated post gadolinium through the orbits showing abnormal expansion and enhancement of the left optic nerve in keeping with neuritis.

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**Fig. 21:** Sagittal T2 cervicothoracic spine reveals extensive cord signal change extending over multiple segments and reflecting myelitis.

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Fig. 22: Axial T2 of a 29 year old male with trigeminal neuralgia showing a demyelinating plaque at the right trigeminal nerve root entry zone.

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Fig. 23: Axial T2 of a 19 year old male with infective endocarditis showing two focal lesions - pons and left cerebellum - with accompanying vasogenic oedema.

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**Fig. 24:** Axial T1 post gadolinium demonstrates ring enhancement of the lesions and DWI (Fig 25-26) shows central restricted diffusion typical for pyogenic abscesses.

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Fig. 25: B1000 DWI showing high signal centrally within the lesions.

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**Fig. 26:** ADC map showing corresponding central diffusion restriction.

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**Fig. 27:** B1000 DWI of an 81 year old male showing bilateral acute pontine perforator infarcts with corresponding low ADC (Fig 28) indicating restricted diffusion. Note how the pontine midline is maintained.

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Fig. 28: ADC showing diffusion restriction in acute bilateral pontine infarction.

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**Fig. 29:** Axial T2 of a 74 year old male showing diffuse pontine signal abnormality without pontine expansion.

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**Fig. 30:** Axial T2 of the same patient in Fig 29 showing more focal ischaemic change in the pons and multiple bilateral cerebellar infarcts indicating that the patient has chronic cerebrovascular disease.

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**Fig. 31:** Axial T2 of the cerebrum of the same patient in Fig 29-30 revealing the typical supratentorial pattern of small vessel disease with early confluent and punctate T2 signal abnormality in the cerebral white matter.

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**Fig. 32:** Axial T2 of a 34 year old male with trigeminal neuralgia showing a well-defined mixed intensity lesion consistent with cavernoma in the pons at the level of the right trigeminal nerve root entry zone.

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Fig. 33: Axial T1 show the lesion to be bright indicating subacute haemorrhage.

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Fig. 34: Axial susceptibility weighted image (SWI) reveals signal intensity reduction in keeping with haemorrhage within the lesion in addition to an associated developmental venous anomaly, seen as serpiginous vessels leading away from the lesions posterior edge.

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**Fig. 35:** Axial T2 of a 64 year male with multiple system atrophy showing the classic, although not pathognomonic, "hot cross bun" sign within the pons.

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Fig. 36: Axial T2 of the same patient in Fig 35 reveals volume loss in the putamina bilaterally with bilateral putaminal signal intensity reduction posterolaterally and a thin rim of T2 hyperintensity change immediately lateral to the putamina.

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Fig. 37: Axial T2 of a 49 year old alcoholic showing diffuse central hyperintensity change with mild pontine expansion consistent with central pontine myelinolysis.

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Conclusion

Radiological assessment with MRI is fundamental to the investigation of pathologies within the pons but the imaging findings can be diverse and lead to uncertainty regarding the underlying diagnosis. Familiarization with the radiological appearances of common pontine pathologies and with the associated symptomatology will increase diagnostic confidence and permit timely treatment stratification.

Personal information

References