

49: Answer

49 **DIAGNOSIS** Low-grade astrocytoma (WHO grade 2).

IMAGING FINDINGS There is focal increased T2/FLAIR signal seen within the anterior medial left temporal lobe (49A) (i). This corresponds to hypointense T1 signal (49B) and shows no contrast enhancement (49C). No surrounding vasogenic oedema is seen. There is very little mass effect and no volume loss. No other lesions were seen.

DIFFERENTIAL DIAGNOSIS Primary differential diagnosis includes low-grade primary glial tumour, encephalitis, and infarction. Of the infectious causes, herpes encephalitis is the most worrisome and should always be considered when medial temporal abnormality is present (ii). Indeed, it is common to treat the patient empirically with anti-viral medication until serology and CSF studies can be definitively evaluated. The lack of diffusion restriction essentially rules out infarction. Metastatic disease is not seriously considered due to the lack of enhancement.

PATHOLOGY AND CLINICAL CORRELATION Low-grade astrocytomas (WHO grade 1 and 2) can be either focal or diffuse (iii). When focal, they can appear extremely benign. Though usually absent, minimal enhancement can occasionally be seen. Pathologically, all astrocytomas originate within white matter but these tumours can extend into the grey matter of the cortex or deep nuclei. Neoplastic cells can extend beyond the margins of MR signal abnormality. More worrisome, these tumours have the potential for malignant progression into anaplastic astrocytomas. Indeed, most cases of postresection recurrence are due to dedifferentiation.

Low-grade astrocytomas represent approximately 25% of all glial tumours. Presentation depends on the area of the brain involved. Approximately 2/3 of lesions are supratentorial while the remaining 1/3 are seen in the cerebellum and brainstem. Though they can affect any age group, patients tend to be younger than those affected by glioblastoma multiforme (WHO grade 4). Treatment is surgical resection. Adjuvant chemo- and radiation therapy is also often administered.

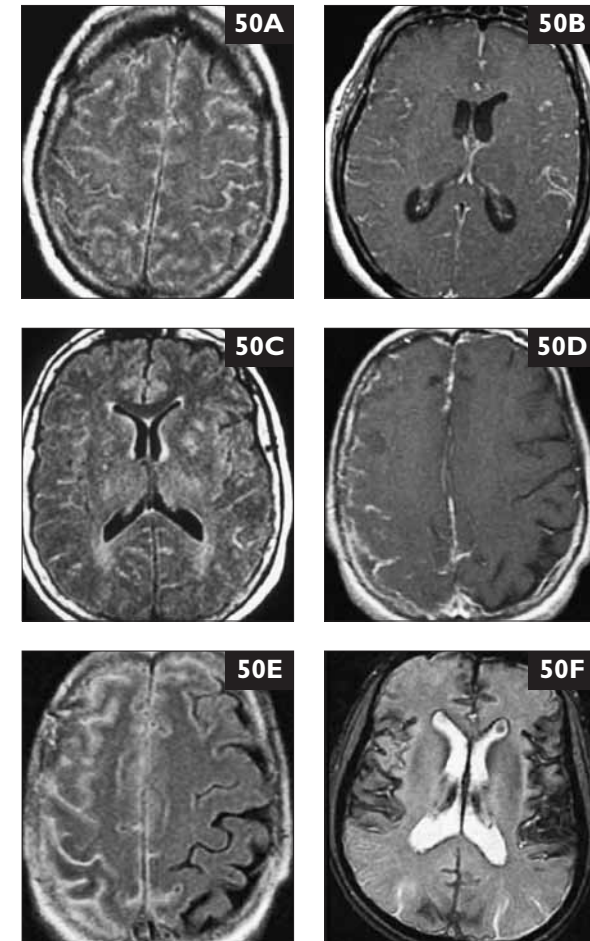
TEACHING PEARLS

- > Low-grade astrocytomas can be either focal or diffuse.
- > The lack of enhancement and surrounding oedema can make these lesions appear extremely benign.
- > Neoplastic cells extend beyond the margins of signal abnormality.

REFERENCES

- Burger PC, Scheithauer BW, Vogel FS (2002). *Surgical Pathology of the Nervous System and its Coverings*. The Brain: Tumours. 4th edn. Churchill Livingstone, Philadelphia, pp. 160–77.
- Kleihues P, Cavenee WK (2000). *Pathology and Genetics of Tumours of the Nervous System*. Diffuse astrocytoma. IARC Press, Lyon, pp. 22–6.
- Osborn AG, et al. (2004). Brain. Diffuse astrocytoma, low-grade. *AMIRSYS*, Salt Lake City, I-6 pp.8–11.
- Wessels PH, et al. (2003). Supratentorial grade II astrocytoma: biological features and clinical course. *Lancet Neurol* 2(7):395–403.

50: Question



50 A 23-year-old female presents with headache, fever, nuchal rigidity, and altered mental status. The following images are obtained (50A, B).

- Which sequences are shown?
- What is the pathological finding?
- What are typical complications of this disorder?
- What is the differential diagnosis?

DIFFERENTIAL DIAGNOSIS Images 50C–F.

50: Answer

50 **DIAGNOSIS** Pneumococcal meningitis.

IMAGING FINDINGS Images 50A, B (axial FLAIR, axial contrast-enhanced T1, respectively) (i) show hyperintensity and enhancement in the sulci and cisterns, representing exudate in a patient with pneumococcus meningitis (ii).

DIFFERENTIAL DIAGNOSIS The differential diagnosis (iv) of a diffuse leptomeningeal disease includes:

- Bacterial meningitis (e.g. *S. pneumoniae*) (50A, B)
- Viral meningitis.
- Carcinomatous meningitis.
- Neurosarcoïd.
- Fungal meningitis (e.g. coccidiomycosis) (50C, axial FLAIR).
- Rheumatoid meningitis (50D, E, axial contrast-enhanced T1 and FLAIR).
- Plus: see differential diagnosis for localized leptomeningeal disease (Question 33).

Mimics:

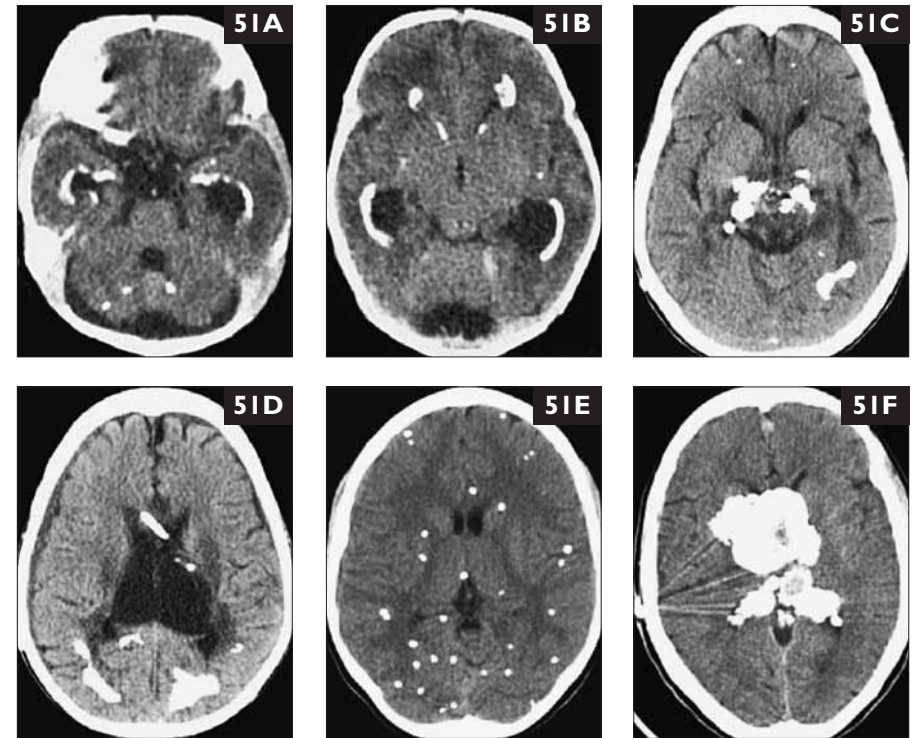
- Increased FLAIR signal in cerebrospinal fluid (CSF):
 - Subarachnoid haemorrhage.
 - 100% inspired oxygen.
 - Acute stroke (parenchymal oedema, congestion).
 - Gadolinium in CSF (dialysis-dependent patient).
 - Artifact.
- Superficial haemosiderosis (50F, T2*GRE).
- Moyamoya (see Question 74).

PATHOLOGY AND CLINICAL CORRELATION Meningitis is defined as inflammatory infiltration of the pia mater, arachnoid, and CSF, most commonly caused by haematogenous dissemination from a distant infection. Fungal and tuberculous meningitis are often basilar, nodular, and confluent. Meningitis is associated with three major types of complications; impaired CSF resorption may cause extraventricular obstructive hydrocephalus with increased intracranial pressure and perfusion alterations. Complications include empyema, ventriculitis, abscess, and vascular complications including ischaemia due to arterial spasm or infectious arteritis (iii).

TEACHING PEARLS

- *Imaging may be normal early in the disease process.*
- *Imaging findings are nonspecific, with intense leptomeningeal enhancement on contrast-enhanced T1 and hyperintense signal in sulci and cisterns on FLAIR and DWI sequences.*
- *Imaging delineates complications that occur in about 50% of adult patients.*
- *Despite effective anti-microbial agents, overall mortality rate is still 25%.*

51: Question



51 A 6-month-old male newborn presents with microcephaly and hearing loss. The following images are obtained (51A–D).

- What do the hyperdensities represent and where are they localized?
- What other cerebral and cerebellar pathologies would you expect in this disorder?
- What modality is usually used for neonatal screening?
- What is the differential diagnosis?

DIFFERENTIAL DIAGNOSIS Images 51E, F.

51: Answer

51 DIAGNOSIS Congenital cytomegalovirus (CMV).

IMAGING FINDINGS Images 51A–D (axial unenhanced CT) demonstrate multifocal coarse periventricular and subependymal calcifications involving the periventricular white matter (WM) (51A, B), the cerebellum (51A), the occipital lobe (51D) and deep grey nuclei (51C) (i). Dilation of the lateral ventricle (especially the body, atrium, and temporal horn) due to WM volume loss is best seen on 51D.

DIFFERENTIAL DIAGNOSIS The differential diagnosis (iv) of periventricular calcification includes:

In neonates:

- CMV (51A–D, axial NECT).
- Toxoplasmosis.
- Congenital HIV.
- Lymphocytic choriomeningitis (LCM).
- Rubella virus.

In any age-group:

- Tuberosus sclerosis (see Question 13).

Additional causes of parenchymal calcifications (not periventricular in distribution) are shown in images 51E, F. Image 51E demonstrates nodular calcified stage of neurocysticercosis with multiple calcified shrunken nodules in both hemispheres. Image 51F demonstrates an intensely calcified midline meningioma localized in the body, atrium, and septum pellucidum of the lateral ventricle bilaterally with extraventricular extension.

PATHOLOGY AND CLINICAL CORRELATION Congenital CMV infection is caused by transplacental transmission. Cranial sonography is used for neonatal screening (iii). Most newborns with central nervous system (CNS) involvement have major neurodevelopmental sequelae.

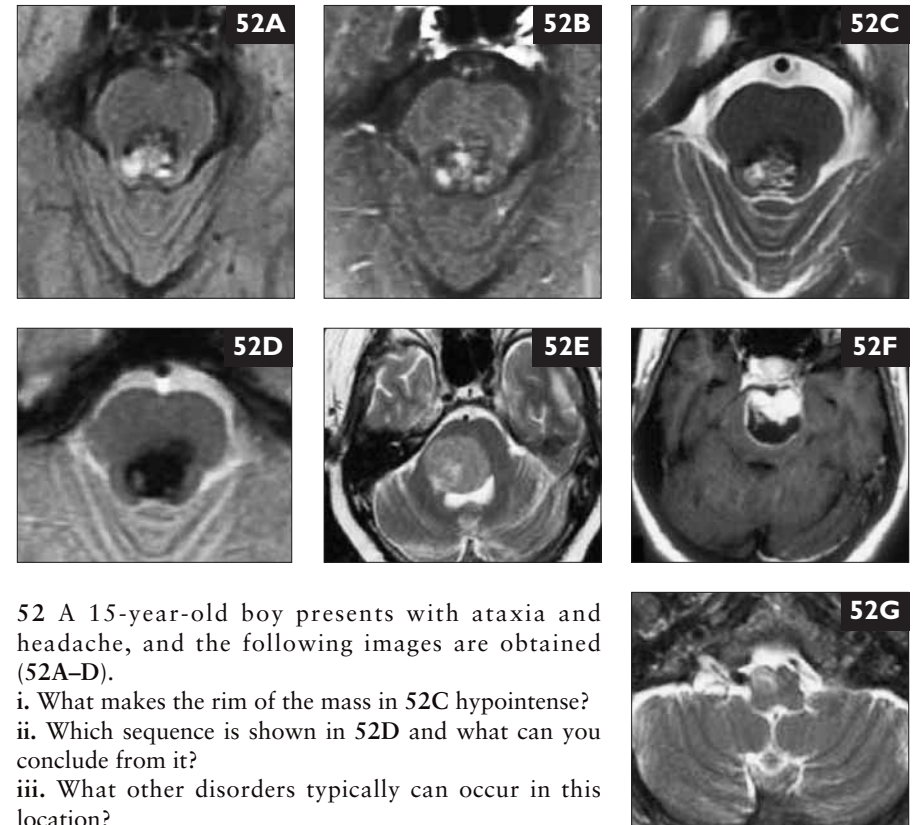
TEACHING PEARLS

- > CMV is the most common intrauterine infection affecting 1% of all newborns.
- > It presents with microcephaly, periventricular calcifications, cortical gyral abnormalities, and cerebellar hypoplasia (ii).
- > Clinically, think of CMV in a microcephalic, developmentally delayed infant with sensorineural hearing loss (SNHL).
- > However, most infected newborns appear normal.

REFERENCES

- Abe K, et al. (2004). Comparison of conventional and diffusion-weighted MRI and proton MR spectroscopy in patients with mitochondrial encephalomyopathy, lactic acidosis, and stroke-like events. *Neuroradiology* 46(2):113–7.
- Osborn AG, et al. (2004). *Brain*. Amirsys, Salt Lake City, Chapter I-10, pp. 28–31.

52: Question



52 A 15-year-old boy presents with ataxia and headache, and the following images are obtained (52A–D).

- What makes the rim of the mass in 52C hypointense?
- Which sequence is shown in 52D and what can you conclude from it?
- What other disorders typically can occur in this location?

DIFFERENTIAL DIAGNOSIS Images 52E–G.

52 **DIAGNOSIS** Cavernous malformation (CM).

IMAGING FINDINGS Images 52A–D (axial T1, axial T1 postgadolinium, axial T2, axial gradient echo, respectively) demonstrate a CM in the posterior pons compressing the 4th ventricle. The mass demonstrates no significant enhancement (compare 52A and 52B). Mixed signal characteristics suggest blood products in different stages of evolution, surrounded by a rim of hypointense hemosiderin (i) that causes susceptibility artifact on the gradient echo image shown in 52D (ii). No oedema or mass effect is visible.

DIFFERENTIAL DIAGNOSIS The differential diagnosis (iii) of a brainstem mass includes:

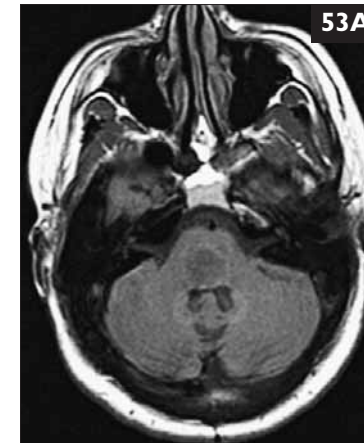
- Metastasis (52F; axial T1 postgadolinium).
- Pontine glioma (52E; axial T2).
- Pontine infarct.
- Multiple sclerosis/ acute disseminated encephalomyelitis.
- CM (52A–D).
- Neurofibromatosis type 1.
- Osmotic demyelination syndrome (see Question 65).
- Wernicke encephalopathy.
- Hypertrophic olivary degeneration (52G, axial T2).

The differential diagnosis of brainstem lesions is broad; accurate diagnosis requires consideration of patient age and specific imaging characteristics. Pontine glioma (52E) and metastasis (52F) represent the most common neoplastic lesions in children and adults, respectively. Focal areas of signal abnormality should prompt consideration of infarct or metabolic abnormality. Hypertrophic olivary degeneration (52G) is hypertrophy and T2 hyperintensity of the inferior olivary nucleus in response to an interruption of neuronal input along the triangle of Guillain–Mollaret, which runs from the dentate nucleus in the cerebellum via the red nucleus to the inferior olivary nucleus.

PATHOLOGY AND CLINICAL CORRELATION CM is a benign vascular hamartoma consisting of immature blood vessels with intralesional haemorrhages and lack of neural tissue. Within CM, 75% occur as a solitary, sporadic lesion and 25% as multiple, familial lesions with earlier presentation and higher risk of bleeding than the sporadic form. CMs might occur throughout the central nervous system, although the brain, especially the brainstem, is much more often affected than the spinal cord. CMs show a very variable prognosis, because lesions might regress, enlarge, or develop *de novo*. The typical clinical presentation is a 40–60-year-old patient with seizures after bleeding.

TEACHING PEARLS (CASE 52)

- > Think of CM in middle-aged patients with spontaneous intracranial haemorrhage.
- > CM is a round nonenhancing haemorrhagic mass (best seen in T2*GRE) with complete hemosiderin rim on T2.
- > It is the most common angiographically occult vascular malformation.
- > CM is most commonly located in brainstem or cerebral hemispheres.
- > Gradient echo images are useful to identify additional lesions.



53 An adult presents with right facial droop. The following images are obtained (53A–C).

- Is brainstem biopsy usually performed to confirm diagnosis?
- Define the anatomic boundaries of the brainstem.
- Name the age range most commonly affected by primary tumours of the brainstem.