



# Primary central nervous system vasculitis mimicking a brain tumor on conventional magnetic resonance imaging: the usefulness of perfusion-weighted imaging. A case report

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A 31-year-old woman presented with headache and subacute aphasia. Brain magnetic resonance imaging (MRI) (Fig. 1) showed a left temporal lobe intraaxial mass of  $2.6 \times 1.8 \times 2.5$  cm. The lesion was first identified on T2-weighted images where the peripheral, solid part of the mass was isointense to gray matter and the central part hyperintense, consistent with necrosis. Gadolinium-enhanced T1-weighted images showed a peripheral “ring-enhancement”. MR angiography was normal. Diffusion-weighted MRI with apparent diffusion coefficient map (DWI-ADC) showed low diffusion value ( $0.000870 \text{ mm}^2/\text{s}$ ) in the solid portion of the mass, and high diffusion value ( $0.00194 \text{ mm}^2/\text{s}$ ) in the central portion, confirming central necrosis. Dynamic susceptibility contrast perfusion-weighted MRI (DSC-PWI) demonstrated low relative cerebral blood volume (rCBV) in both lesion wall (mean value 0.76) and perilesional area, compared to normal white matter. Biopsy revealed ischemic necrosis without cellular atypia, coupled with perivascular and transmural infiltrate of lymphonuclear cells involving the vessels, which caused narrowing of their vascular lumen. History did not reveal any underlying disease, including immune disorders. Antinuclear antibodies, antineutrophil cytoplasmatic antibodies, and rheumatoid factor were normal. A diagnosis of primary

central nervous system vasculitis (PCNSV) was then established. Steroid therapy was immediately initiated. Two months later, clinical examination was negative and brain MRI showed gliotic-malacic evolution without new intracranial lesions.

PCNSV is a rare and poorly understood vascular inflammatory disease affecting the central nervous system [1]. Diagnosis can be difficult both clinically and neuroradiologically. Computed tomography and MRI findings are various and nonspecific, ranging from normal features to acute, subacute or chronic ischemic and/or hemorrhagic changes, leptomeningeal contrast-enhancement, and mass lesions as occurred in the patient presented herein. Hemorrhagic and/or ischemic acute presentation results in typical neuro-radiological findings. Subacute presentation may result in irregular contrast-enhancement and increased diffusion at DWI-ADC, mimicking a brain tumor [2]. Vasculitic changes in large- to medium-size arteries may be disclosed by vascular neuroimaging, with digital subtraction angiography being more sensitive, despite its invasiveness and radiation exposure. Vasculitic changes in small vessels may be seen at biopsy.

In our patient, MRI showed a tumor-like “ring-enhancing” lesion, associated with vasogenic edema, as can be observed in glioblastoma or metastasis. DSC-PWI is a non-invasive technique providing unique information about cerebral haemodynamics and is mainly dependent on the lesions microvasculature [3]. In intracranial lesions, T1 MR signal increase after gadolinium administration is associated with damage of the blood-brain barrier, whereas in high-grade gliomas, cerebral metastases, and lymphoma an increased rCBV is usually associated with increased neovascularization in both solid tumor components and peritumoural areas. DSC-PWI metrics can be also used to identify and quantify areas of high capillary density [3]. Conversely, small-vessel involvement in PCNSV is readily displayed by DSC-PWI as regional

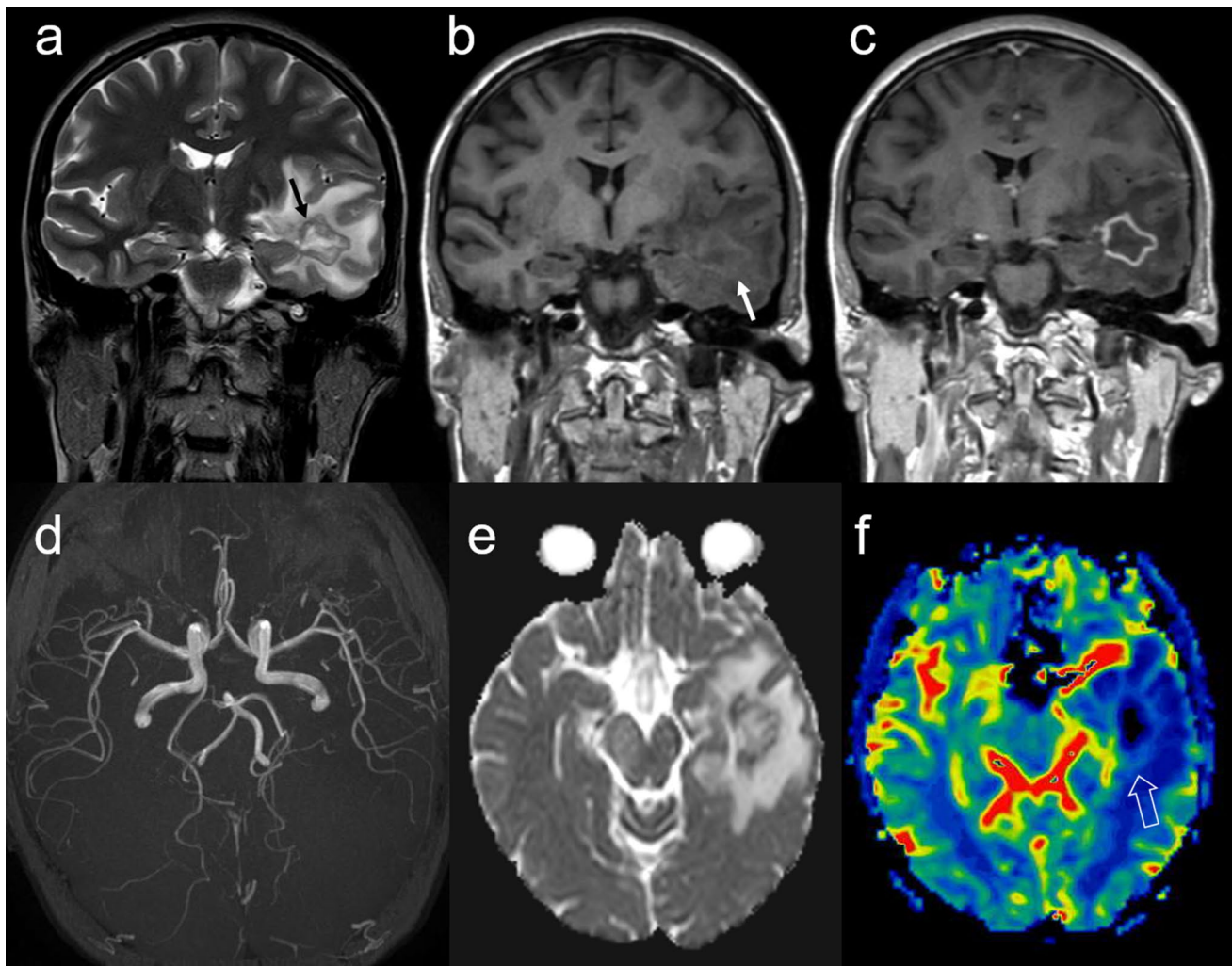
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**Fig. 1** Magnetic resonance (MR) imaging of the brain. T2- (a), unenhanced T1- (b), and gadolinium-enhanced T1-weighted (c) coronal MR images show a left temporal lobe necrotic mass lesion with a peripheral rim of T2-weighted hypointensity (black arrow) and T1-weighted iso-hyperintensity (white arrow), surrounded by vasogenic edema. Time-of-flight Intracranial MR angiography (d) is nega-

tive for large- to medium-size arteries abnormalities. Apparent diffusion coefficient map (e) shows restricted diffusion in the peripheral rim lesion and increased diffusion in its central part. Relative cerebral blood volume (rCBV) map (f) does not show an increased rCBV in both lesion wall and perilesional area (open arrow) when compared to normal white matter

hypoperfusion [4]. Yuh et al. [5] reported a patient with PCNSV who presented with a stroke-like lesion on conventional MRI and a negative angiogram, and had been diagnosed on the basis of DSC-PWI.

In conclusion, in lesions resembling brain tumors on conventional MRI, a regional hypoperfusion on DSC-PWI may address toward the correct diagnosis of PCNSV.

**Author contributions** All authors have approved the manuscript.

### Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflict of interest.

**Ethical approval** All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

**Informed consent** Informed consent was obtained from all individual participants included in the study.

## References

1. Greenan TJ, Grossman RI, Goldberg HI (1992) Cerebral vasculitis: MR imaging and angiographic correlation. *Radiology* 182:65–72
2. Yaldizli O, Kastrup O, Wanke I, Maschke M (2005) Basal ganglia infarction mimicking glioblastoma. *Eur J Med Res* 10:400–401
3. Muccio CF, Esposito G, Bartolini A, Cerase A (2008) Cerebral abscesses and necrotic cerebral tumours: differential diagnosis by perfusion-weighted magnetic resonance imaging. *Radiol Med* 113:747–757
4. Muccio CF, Di Blasi A, Esposito G, Brunese L, D'Arco F, Caranci F (2013) Perfusion and spectroscopy magnetic resonance imaging in a case of lymphocytic vasculitis mimicking brain tumor. *Pol J Radiol* 78:66–69
5. Yuh WTC, Ueda T, Maley JE (1999) Perfusion and diffusion: a potential tool for improved diagnosis of CNS vasculitis. *Am J Neuroradiol* 20:87–89