



Diploic epidermoid cyst associated with focal brain pseudohypertrophy

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Abstract

We present an 82-year-old woman, admitted for generalized status epilepticus, showing an osteolytic lesion of the right parietal bone associated with underlying focal brain pseudohypertrophy. A radiological diagnosis of diploic epidermoid cyst based on MRI characteristics was made. The aspect of focal brain pseudohypertrophy was probably caused by direct involvement of the underlying meningeal structures preventing the brain parenchyma locally from being retracted (i.e. retraction due to age-related brain atrophy). This aspect of focal brain pseudohypertrophy can potentially be seen in all chronic primary cranial bone lesions associated with meningeal involvement.

Case report

An 82-year-old woman without medical history of seizures presented with a generalized status epilepticus. Clinical examination, one hour after intravenously administered lorazepam (4 mg) and a bolus of valproic acid (15 mg/kg), showed no abnormalities apart of a small subcutaneous painless parietal scalp swelling covered with normal skin. The patient had no head trauma during the status epilepticus. Unenhanced brain CT showed a 5.5 cm large osteolytic lesion involving the inner table of the right parietal bone with adjacent tissue isointense to normal brain parenchyma, together with generalized brain atrophy (Fig. A and B). On brain MRI (Fig. C-J), the osteolytic lesion was hypointense on T1-weighted and hyperintense on T2-weighted imaging, and hyperintense relative to CSF on FLAIR sequences. The osteolytic lesion showed restricted diffusion, a hyperintense signal on diffusion-weighted imaging (DWI) and a hypointense signal on the ADC-map. Gadolinium-enhanced T1-weighted imaging revealed a (probably meningeal) enhancement of the inferior rim of the osteolytic lesion. A radiological diagnosis of diploic epidermoid cyst was

made. Tissue adjacent to the cranial vault lesion was in continuity with the underlying brain and isointense to normal brain parenchyma on all sequences, giving the aspect of focal brain hypertrophy. Focal brain pseudohypertrophy was probably explained by the attachment of the epidermoid lesion to the inner layers of the meninges preventing the underlying brain parenchyma from being retracted (in contrast to the rest of the brain showing – probably age-related – brain atrophy and retraction). Interictal electroencephalogram showed the presence of some right-sided parietal epileptiform discharges, although interpretation was difficult because of the partial parietal skull deficit. Seizures may have been provoked by traction on the involved brain parenchyma and/or by direct involvement of the underlying cortex by the epidermoid lesion. Since brain MRI was stable six months later and seizures did not recur under anticonvulsant treatment (valproic acid 500 mg bid), surgical intervention was not performed.

Discussion

To our knowledge, this is the first report on diploic epidermoid cyst associated with an aspect of focal brain pseudohypertrophy. The combination of both radiological abnormalities in our patient was probably caused by direct involvement of the underlying meningeal structures preventing the brain parenchyma locally from being retracted (i.e. retraction due to age-related brain atrophy). Differential diagnosis of an osteolytic cranial vault lesion includes primary bone tumour (e.g. osteoid osteoma, eosinophilic granuloma, lymphoma), meningioma, metastases, hemangioma, Paget's disease, and hypertrophic pachionian granulation. Restricted diffusion on diffusion-weighted imaging is typical for epidermoid lesions, and is not seen in these other lesions (Hoe *et al.*, 2004; Nambu *et al.*, 2006).

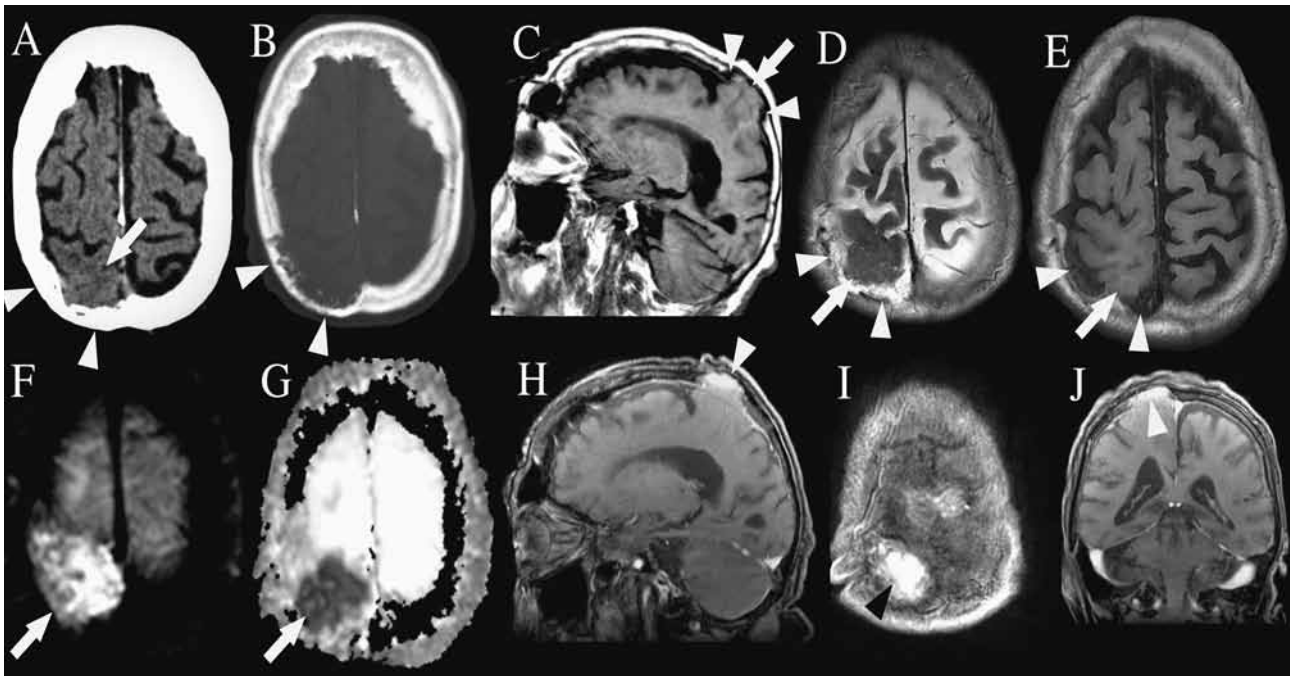


FIG. 1. — Brain CT (A and B) showing a 5.5 cm large osteolytic lesion involving the inner table of the right parietal bone (arrowheads) with adjacent tissue isointense to normal brain parenchyma (arrow). On MRI, the osteolytic lesion is hypointense on T1-weighted imaging (C, arrowheads), hyperintense on T2-weighted imaging (D, arrowheads), and hyperintense relative to CSF on FLAIR sequences (E, arrowheads). On the sagittal T1 sequences (C), the small parietal head protrusion seen on clinical examination can be noticed. On the uppermost axial sequences (resulting in an axial view of the bone lesion), the osteolytic lesion is strongly hyperintense on DWI (F, arrow) and hypointense on ADC-map (G, arrow). Gadolinium-enhanced T1-weighted imaging showing strong enhancement of inferior part of the osteolytic lesion and/or adjacent meningeal structures (arrowheads on H, I, and J corresponding to sagittal, axial, and coronal sequences). Underlying focal enlargement of the lateral ventricle can be seen on coronal sequences (J).

Hypertrophic pacchionian granulation can mimic an osteolytic lesion of the calvaria, and attachment to the arachnoid membrane has been reported (Celli *et al.*, 1999; Kuroiwa *et al.*, 1996; Beatty *et al.*, 1989). However, almost all reports of hypertrophic pacchionian granulation affect the midline or the adjacent bone within 2 cm of the midline, although exceptions have been reported. Primary brain tumours or meningioma can provoke osteolytic bone lesion to the adjacent cranial bone structures. In our patient, brain parenchyma did not show radiological signal changes, and contrast enhancement was only seen in the osteolytic and/or meningeal lesion (and not in the brain parenchyma itself), making a bone lesion secondary to a primary brain tumour unlikely. Tissue adjacent to the cranial vault lesion was in continuity with the underlying brain and isointense to normal brain parenchyma on all sequences, giving the aspect of focal brain hypertrophy. Seizure onset in the 9th decade of life, the absence of radiological signs of cortical dysplasia (e.g. increased cortical

thickness, poor differentiation from the white matter, polygyria, agyria, microgyria), an underlying focal enlarged (rather than reduced) lateral ventricle, and the presence of an associated bone lesion was against the diagnosis of focal megalencephaly (Abdel Razek *et al.*, 2009; Foldvary-Schaefer *et al.*, 2004).

Diploic epidermoid cysts are most frequently located in the parietal bone (Arana *et al.*, 1996). The most common symptom is a painless subcutaneous scalp swelling covered with normal skin. Often both tables of the cranial vault are involved. However, isolated inner (as in our patient) or outer table involvement is frequently reported, with a slight predilection for the inner table. Seizures have been described in some patients with intradiploic epidermoid cyst (Cho *et al.*, 2007). In the largest series of patients with diploic epidermoid cysts, 78% had sclerotic borders and 22% non-sclerotic borders (Arana *et al.*, 1996). This aspect of focal brain pseudohypertrophy can potentially be seen in all chronic primary cranial bone lesions associated with meningeal involvement.

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