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Diffuse Calvarial Hemangiomatosis Associated with Hereditary Hemorrhagic Telangiectasia

Hereditary hemorrhagic telangiectasia or Rendu-Osler-Weber disease often is associated with CNS abnormalities [1, 2]. We describe a case in which a patient with hereditary hemorrhagic telangiectasia had multiple cranial nerve deficits associated with calvarial hemangiomatosis.

Case Report

MR, CT, and angiography were performed on a 49-year-old woman with hereditary hemorrhagic telangiectasia who had acute onset of left-sided hearing loss. The patient previously had had a left occipital craniotomy, and subsequently, diffuse calvarial hemangioma had been diagnosed. Physical examination at the time of admission revealed bilateral deficits of the seventh and eighth cranial nerves.

MR showed multiple lesions of abnormal signal intensity involving the diploic space of the calvarium in the temporal, parietal, and occipital bones (Fig. 1). Enhanced cranial CT showed bilateral expansile calvarial and skull base defects. Cerebral arteriography showed multiple irregular vascular lakes and pools fed by the external carotid arteries.

Discussion

CNS involvement is said to occur in 27–29% of patients who have hereditary hemorrhagic telangiectasia [3]. The association of calvarial hemangiomatosis with this disease appears to be a rare occurrence; it has been reported only once before [4]. Our example is unusual in that deficits of the seventh and eighth cranial nerves occurred as a result of involvement of the temporal bone.

Although the plain-film and arteriographic findings of calvarial hemangioma have been described before [5, 6], the MR appearance

has not. Ross et al. [7] recently reported on vertebral hemangiomas. All of their examples were of increased signal intensity relative to adjacent normal vertebral body on T1-weighted images. Also, every lesion in their study increased in signal with T2 weighting. Our case was similar; on T1-weighted images, the lesions were of increased signal relative to adjacent calvarium and progressively increased in signal with greater T2 weighting. This may be a characteristic MR pattern for osseous hemangioma. Differences in relative intensity between individual lesions on any given spin-echo sequence may be related to the constituents of the hemangioma.

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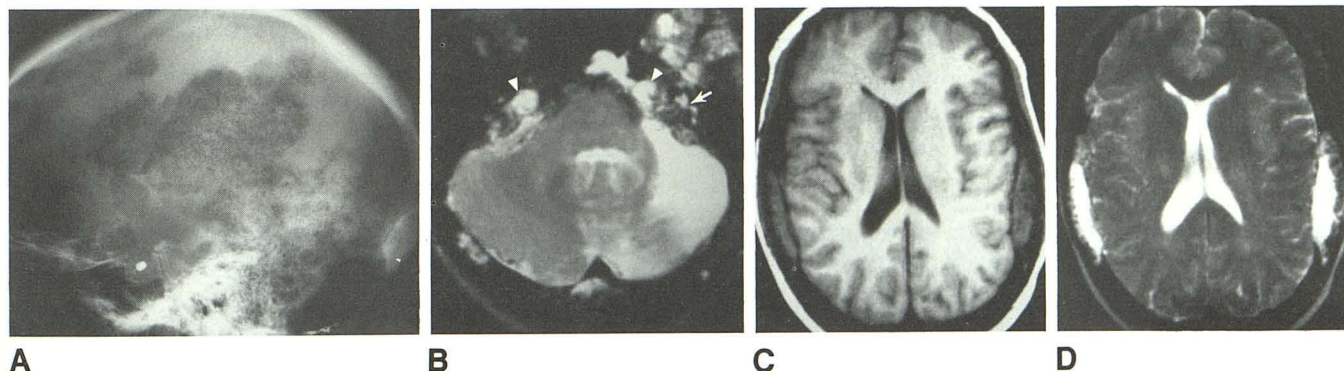


Fig. 1.—Diffuse calvarial hemangiomatosis associated with hereditary hemorrhagic telangiectasia.
A, Lateral skull film shows typical bony pattern of calvarial hemangioma consisting of geographic granular-appearing defects (multiple in this case).
B, T2-weighted spin-echo axial MR image, 2500/80, two excitations, shows abnormal increased signal intensity associated with hemangioma (arrowheads) at level of internal auditory canals (arrow is on left internal auditory canal). Increased CSF space in left posterior fossa is associated with previous surgery.
C, T1-weighted spin-echo axial MR image, 600/15, at level of lateral ventricles shows broad-based extradural masses arising from calvarium in both parietooccipital regions and mild compression of underlying brain tissue. Hemangioma has increased signal relative to adjacent calvarium.
D, T2-weighted spin-echo axial MR image, 2000/120, shows lesions increase markedly in signal with heavy T2 weighting.