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MR of Xanthogranuloma of the Choroid Plexus

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Summary: We present a case of a xanthogranuloma of the lateral ventricle choroid plexus in association with focal areas of abnormal T2 signal in the tegmentum of the pons as well as within the middle cerebellar peduncles. The characteristic MR appearance of this rare entity is described along with a pathologic basis suggesting an association with posterior fossa lesions.

Index term: Choroid plexus, neoplasms

Xanthogranulomas are uncommon lesions of the choroid plexus in the lateral ventricle. The lesions are usually asymptomatic and encountered at autopsies; their frequency varies from 1.6% to 7% (1, 2). Most are tiny plaques, but, rarely, they occur as masses of significant size. Previous reports of the neuroradiologic findings of symptomatic cases with surgical intervention have been infrequent (3–7). We present a confirmed case in which magnetic resonance (MR) imaging provided detailed definition of a xanthogranuloma of the lateral ventricle choroid plexus accompanied by focal areas of hyperintensity on T2-weighted images in the posterior fossa.

Case Report

A 43-year-old man was first admitted in June 1992 with complaints of dysarthria and gait disturbance for the previous 2 years. Admission laboratory data revealed elevated serum triglyceride of 3.52 mmol/L in the absence of any increase in serum cholesterol level. Neurologic examination disclosed slurred speech and cerebellar ataxia. There were no associated systemic findings, such as superficial cutaneous xanthomas. Plain skull films were normal. Computed tomography (CT) revealed a round area with slightly increased density and homogeneous contrast enhancement (Fig 1A and B). Four-vessel studies did not contribute to the diagnosis of the lesion.

MR examination was performed on a 1.5-T imager. T1-weighted (500/15/2 [repetition time/echo time/excitations]) and T2-weighted (3000/90/1) images were obtained before and after administration of gadopentetate dimeglumine.

A well-defined nodule measuring 2.0 × 1.7 × 1.8 cm was delineated in the trigone of the left lateral ventricle protruding into the medial cerebral parenchyma, and the posterior part of the left lateral ventricle was slightly dilated. The lesion showed homogeneous isointensity on T1-weighted images and hypointensity on T2-weighted images (Fig 1C and D). Several foci of increased intensity in the tegmentum of the pons as well as within the middle cerebellar peduncles, which were not detected on CT scans, were evident on T2-weighted images (Fig 1E). The lesion showed homogeneous contrast enhancement (Fig 1F).

At occipital craniotomy, an elastic, hard, pale yellow tumor with a smooth surface was found in the trigone of the left lateral ventricle. Pathologic examination revealed proliferation of lipid-laden foamy cells associated with a mixed population of lymphocytes, histiocytes, giant cells and chronic inflammatory cells, and diffuse deposition of hemosiderin (Fig 1G). The histopathologic diagnosis was xanthogranuloma of the choroid plexus.

The clinical course after surgery was uneventful. No additional neurologic deficit developed, and no significant postoperative change in clinical symptoms has been noted.

Discussion

As to the pathogenesis of the choroid plexus xanthogranuloma, Shuangshoti et al (8) suggested that desquamative epithelium of the choroid plexus enters the interstitium at the sites of disruption of basal lamina, where the lipid content of epithelial cells accumulates as they degenerate. Disintegration of the lipid-laden cells releases additional lipid, which provokes a further xanthomatous response. Some are associated with degenerative changes with resultant demyelination of neuronal tissues, whereas others present as masses despite their

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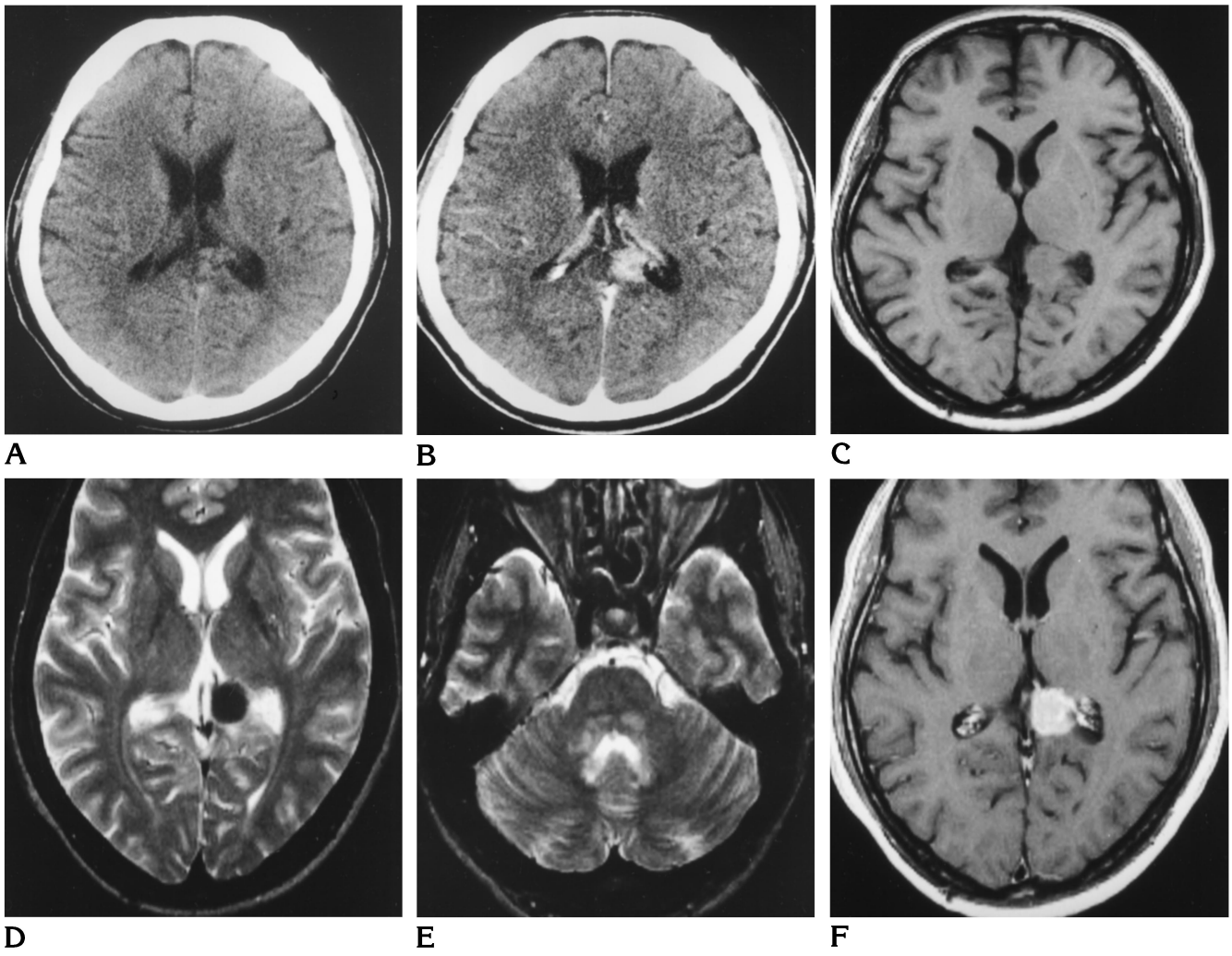

Fig 1. Forty-three-year-old man with xanthogranuloma of the lateral ventricle choroid plexus.

A, Noncontrast CT scan shows an area of slight hyperdensity in the trigone of the left lateral ventricle.

B, Contrast-enhanced CT scan reveals homogeneous enhancement.

C, Noncontrast axial T1-weighted MR image reveals an isointense mass in the trigone of the left lateral ventricle protruding into medial cerebral parenchyma.

D, Noncontrast axial T2-weighted image shows a hypointense tumor.

E, Focal lesions of hyperintensity are seen in the tegmentum of the pons and within the middle cerebellar peduncles on this T2-weighted image.

F, Contrast-enhanced axial T1-weighted image shows homogeneous enhancement of the tumor.

G, Photomicrograph of histopathologic section of the choroid plexus xanthogranuloma shows lipid-laden foamy cells, giant cells, and inflammatory cells in a fibrotic granulation tissue (hematoxylin-eosin, original magnification ×40).

nonneoplastic character. Shuangshoti et al considered hemorrhage as a factor involved in the formation of xanthogranulomas.

Brück et al (6) described MR findings of a choroid plexus xanthogranuloma in the lateral ventricle. MR features in our case resembled those described by these authors except that in their case the lesion was hyperintense on T1-weighted images. This difference in signal intensity might be attributed to the extent of the diffuse deposition of hemosiderin derived from hemorrhage residue.

The differential diagnosis of masses of the lateral ventricle includes meningioma, papilloma, ependymoma, arteriovenous malformation, metastatic neoplasm, and xanthogranuloma. Most of the tumors except for calcified meningioma can be easily differentiated from xanthogranuloma owing to the signal intensity on T1- and T2-weighted images. While a heavily calcified meningioma may appear hypointense on T2-weighted images, it may also appear hypointense on T1-weighted images. Therefore, with the use of MR imaging, it might be possible to differentiate a rare xanthogranuloma from more common tumors of the lateral ventricle.

In addition, in our case, several foci of hyperintensity on T2-weighted images were detected in the tegmentum of the pons as well as within the middle cerebellar peduncles. A possible relationship may exist between choroid plexus xanthogranuloma and the presence of several foci of high signal on T2-weighted images within the inferior tracts, since these lesions may be attributed to pathologically accumulated lipids, whose deposition might cause the pathologic changes of xanthomatous lesions and secondary demyelination of neural tissues. Focal lesions in the white matter that displayed high signal on T2-weighted images, considered to reflect demyelination of the neural tissues, were shown in the cases of cerebrotendinous xanthomatosis described by Hokezu et al (9), and Swanson and Cromwell (10). Although neither of these reports alluded to xanthogranulomatous lesions of the lateral ventricle choroid plexus, we can see a possible tumefacient form of xanthogranuloma of the choroid plexus of the lateral ventricle in the figures accompanying case 8 in the article by Hokezu et al and in the case presented by Swanson and Cromwell. On the other hand, the cases of xanthogranuloma of the choroid plexus we found in the literature did not show the focal lesions in the inferior tracts. In our case, these lesions were observed only with MR imaging, which might be attributed to the superior sensitivity of the imaging method to the pathologic changes of neuronal tissues. There is also the possibility that these two lesions may be unrelated.

Knowledge of the MR characteristics of xanthogranulomas is useful for differentiating tumors of the lateral ventricle. We suggest that xanthogranuloma should be considered when masses showing hypointensity on T2-weighted images and isointensity or hyperintensity on T1-weighted images with homogeneous contrast enhancement are observed in the lateral ventricle choroid plexus. The lesion may be accompanied by focal areas of hyperintensity on T2-weighted images in the posterior fossa.

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