

Xanthogranuloma of the choroid plexus

Somarajan Anandan¹ , Sajeesh S Rajendran², Joesni Joy³

From ¹Consultant Neurologist, ²Resident, Department of Neurology, St Joseph Hospital, Anchal, ³Consultant Neurologist, Department of Neurology, Welcare Hospital, Kochi, Kerala, India

Xanthogranulomas (XGR) are benign lesions that consist of cholesterol clefts, lymphocytic infiltration, giant cells with multiple nucleoli, foamy macrophages (xanthoma cells), fibrous proliferation, and hemosiderin deposits. They characteristically arise in the region of the glomus of the choroid plexus of the lateral ventricles and are usually small. They are most often asymptomatic and found incidentally during imaging. Even though XGRs are described as benign tumors in the literature, they are non-neoplastic and better described as inflammatory pseudotumors with xanthoma cells.

A 68-year-old lady presented to the neurology outpatient department with a 3-year history of positional vertigo. She reported episodes of imbalance without vertigo for the past 2 weeks. There was no history of any weakness, sensory symptoms, upper limb incoordination, or ataxia. There was no headache or vomiting. On examination, her vitals and neurological examination were normal. Magnetic resonance imaging (MRI) of the brain showed bilateral lesions in the trigone of the lateral ventricle. These lesions were hypointense in T1-weighted images, hyperintense in T2-weighted images. The right-sided lesion was hyperintense in fluid-attenuated inversion recovery (FLAIR) images. Diffusion-weighted imaging (DWI) showed hyperintensity bilaterally (Figs. 1 and 2). The apparent diffusion coefficient map did not show any corresponding hypointensity. Contrast imaging was not done. The imaging pattern was suggestive of bilateral Xanthogranuloma of the choroid plexus of the lateral ventricle.

Xanthogranuloma of choroid plexus was initially reported as Cholesteatomatous endothelioma by Blummer in 1900 [1]. These are rare lesions of the central nervous system with an incidence on autopsy being reported as 1.6–7% [2,3]. These lesions are most commonly seen bilaterally in the lateral ventricles. They have also been detected in a number of other intracranial locations, such as the third ventricle, fourth ventricle, the suprasellar cistern, petrous apex, middle ear, and paranasal sinuses [4]. The size of xanthogranuloma of the choroid plexus varies widely, ranging from tiny speck-like regions, not visible on imaging, to

large prominent choroidal masses. In most cases, they measure below 1 cm in diameter and are usually located in the trigones of the lateral ventricles. They are bilateral in two-thirds of cases. However, those occurring in the third or fourth ventricle are more likely to present with obstructive hydrocephalus [5]. They can present with acute intraventricular hemorrhage or aseptic

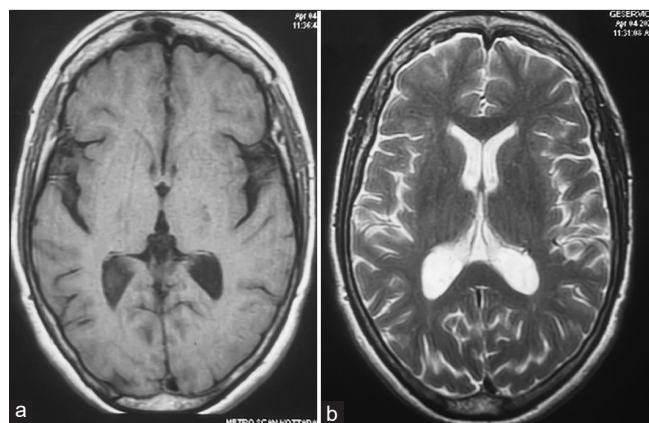


Figure 1: (a) Axial T1-weighted image showing isointense (right) and hypointense (left) lesion in the trigone of the lateral ventricle; (b) Axial T2-weighted image showing hyperintense lesion in the trigone of the lateral ventricle on both sides

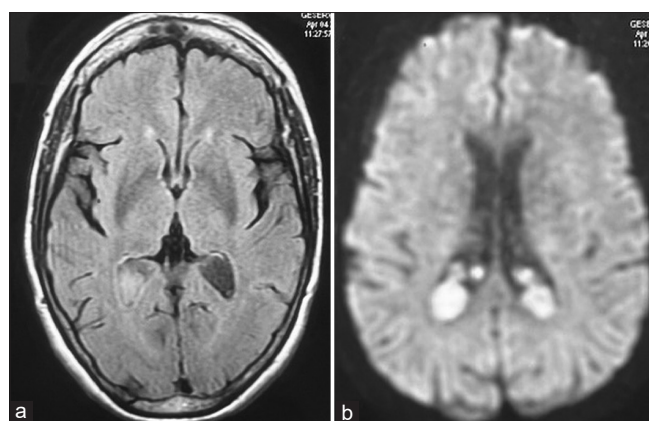



Figure 2: (a) Axial fluid-attenuated inversion recovery image showing hyperintense lesion in right lateral ventricle and hypointense lesion in left lateral ventricle; (b) Axial diffusion-weighted imaging image showing hyperintensity of the lesion bilaterally

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Correspondence to: Somarajan Anandan, Department of Neurology, St Joseph Hospital, Anchal, Kollam - 691306, Kerala, India. E mail: drsomarajan@yahoo.co.in

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chemical meningitis following the spillage of xanthomatous debris into the cerebrospinal fluid. Rarely, they can present with perilesional edema [6].

Histologically, choroidal epithelium is cuboidal, but with age, it proliferates and might show stratification. The proliferated cells may desquamate and become large, round, and foamy with intracytoplasmic lipid accumulation. Desquamated epithelial cells enter the interstitium of the plexus at the sites of disruption of basal lamina and tubular walls and release lipid into the stroma of the plexus, which evokes a granulomatous response. Choroidal epithelial cells also have a phagocytic capability that enables them to act as macrophages and accumulate lipid, glycogen, and hemosiderin [7].

The frequency of intraventricular tumors of the central nervous system is 10% and only 1% of this group contains lateral ventricle tumors, including astrocytoma and choroid plexus papilloma as the most common, and others, including meningiomas, ependymomas, central neurocytomas, XGR, and metastases.

On computerized tomography (CT) scans, XGR are usually oval-shaped and have a smooth wall similar to colloid cysts [8]. CT density of XGRs varies from hypodense to hyperdense in comparison to brain tissue. CT usually demonstrates symmetrical tumefactions of choroid plexus with peripheral calcifications and central low density, but 'not negative' Hounsfield units, located in both ventricular trigones. Signal characteristics on MRI are variable depending on the mixture of lipid, fluid, and blood products. In general, they mimic cystic lesions, although they do not fully attenuate on FLAIR. A helpful feature is that they usually have quite a high signal on DWI. This high signal is seen

as a result of both true restricted diffusion and T2 shine-through effect [9].

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