Symptomatic xanthogranuloma of choroid plexus with unilateral hydrocephalus

Case report

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Xanthogranulomas involving the choroid plexus of the lateral ventricles are generally asymptomatic lesions. The case is reported of a 50-year-old man in whom a xanthogranuloma of the choroid plexus had occluded the left trigone, causing unilateral hydrocephalus of the left temporal horn and neurological symptoms. A review of the literature shows that xanthogranulomas of the glomus of the lateral ventricles differ from the xanthomatous cystic lesions of the third ventricle, which are probably akin to colloid cysts.

Key Words • xanthogranuloma • intraventricular tumor • hydrocephalus

XANTHOGRA NULOMAS, or xanthomas, are uncommon lesions of the glomus of the lateral ventricle choroid plexus. Most are found incidentally at autopsy, their frequency varying from 1.6% to 7%. These lesions tend to be more common in older than in younger subjects and their size usually does not exceed 2 cm.22 Large xanthogranulomas causing clinical symptoms are rare in man; in the horse, such lesions are well known in the veterinary literature.27 In the patient reported here, xanthogranuloma of the left glomus caused unilateral hydrocephalus and neurological symptoms.

Case Report

This 50-year-old man was admitted to the Department of Neurosurgery of the Protestant Hospital of Oldenburg with complaints of left frontoparietal headaches for the previous 5 months. Forgetfulness and amnestic aphasia had manifested 6 weeks prior to admission and motor disturbances followed 2 weeks later. Neurological examination at admission showed amnestic aphasia, a pronation sign, and a tendency to fall to the right. A beginning papilledema was seen on the left. Computerized tomography (CT) showed bilateral lesions in the trigone of both lateral ventricles. The larger, left-sided lesion was enhanced with contrast material. Plain T1-weighted magnetic resonance (MR) images disclosed bilateral lesions with increased signal intensity in the glomus of the lateral ventricles; these lesions showed decreased signal intensity on T2-weighted images and massive enhancement on T1-weighted images after injection of gadolinium. The larger, left-sided lesion had caused complete obstruction of the trigone of the left lateral ventricle. The sequestered left temporal horn had developed a massive hydrocephalic dilatation (Fig. 1a and b).

Operation. A left parietotemporal craniotomy was performed and a partially calcified tumor, at least the size of a cherry, was removed from the dilated trigone. The clinical symptoms disappeared within 10 days postoperatively. A follow-up CT scan showed a significant reduction of the left-sided hydrocephalus (Fig. 1c).

Pathological Examination. A biopsy specimen, 2 × 1.8 × 1 cm, was obtained for histological examination. A densely collagenized tissue contained multiple dissecting tissue clefs; choroid plexus villi adhered to its surface. Many calcified concretions were dispersed throughout the tissue, having a psammoma-like concentric structure. Multinucleated giant cells and sparse inflammatory infiltrates were found near those calcifications. Some cholesterol crystal clefs and nests of lipid-laden foamy cells were also found (Fig. 2). None of the tissue examined showed any structures reminiscent of a meningioma, an ependymoma, or any other type of neoplastic lesion.
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FIG. 1. a: Plain T₂-weighted magnetic resonance (MR) image showing bilateral lesions with decreased signal intensity in the trigone of both lateral ventricles and the massive dilatation of the left temporal horn. b: T₁-weighted MR image after gadolinium infusion. The bilateral lesions show massive enhancement. c: Post-operative computerized tomography scan showing the unilateral hydrocephalus significantly reduced.

FIG. 2. Photomicrographs of the surgical specimen. a: Multinucleated giant cells near mineralization embedded in a dense fibrous stroma. H & E, × 90. b: View showing cholesterol-crystal clefts with a nest of foamy cells. H & E, × 145.
Discussion

On reviewing the literature, we found 19 patients (including ours) in whom intraventricular xanthogranulomas had presented as clinically symptomatic lesions. We excluded reports on plexus granulomas in Hand-Schüller-Christian disease,1,2,3 which evidently differ from the sporadic lesions described here. Of the 19 patients, seven (including ours) had lesions in the lateral ventricles and 12 in the third ventricle. These two groups differ in many ways and we will discuss them separately.

Histology of Incidental Xanthogranulomas of the Glomus

Non-symptomatic xanthogranulomas of the glomus occur either unilaterally or bilaterally. Based on histological criteria, one may distinguish between xanthomas and xanthogranulomas.2,22 The former are usually quite small and are composed of xanthoma cells only, in the absence of cholesterol crystals, foreign body giant cells, or hemorrhage. Xanthogranulomas tend to be larger; they may also contain xanthoma cells, but their prevalent features are cholesterol-crystal clefts, often enclosed by foreign-body giant cells and embedded in a fibrous stroma and residua of hemorrhage. Psammoma bodies are also found. Lesions of this kind typically form in the glomus; they are never seen in the choroid plexus of the third or the fourth ventricle.

Pathogenesis of Xanthogranulomas

There are many theories on the pathogenesis of these lesions, including cellular degeneration,17 tissue reaction to hemorrhage,2 or general disturbance of lipid metabolism. The high levels of free cholesterol and cholesterol esters in xanthogranulomas indicate that these lesions result from active lipid storage caused by a metabolic derangement of the xanthoma cells.6

Symptomatic Xanthogranulomas of Lateral Ventricles

The seven patients (including ours) who had symptomatic xanthogranulomas in the lateral ventricles1,2,12,14,16,20 all had the characteristic histological features of xanthogranulomas. Only one lesion was cystic,20 and all were in the trigone. The lesions, therefore, corresponded to the incidental xanthogranulomas of the glomus in terms of location and structure. The sex ratio approximated 1:1; the youngest patient was 6 years old and the oldest 68 years of age. Most granulomas measured approximately 2 to 3 cm; the largest reported was 4 × 3 × 2 cm.14

The most common neurological symptoms included headache, papilledema, and visual disturbances, but there were also reports of nystagmus,14 epilepsy and mental retardation,20 paragueusia and parosmia,1 and other complaints. Neurological symptoms disappeared after surgery in three patients1,12,14,16 xanthogranuloma was a cause of death in one.3 Bilateral xanthogranulomas with slight ventricle dilatation were described by Terao, et al.20 and Handagooon, et al.3 In the former patient, removal of the tumor had no effect on the neurological symptoms and there was no postoperative change in the ventricle size;20 however, in the latter patient, a correlation between clinical symptoms and the described lesions is not beyond dispute.2 Slight dilatation of the ventricles combined with a contralateral lesion was found in one patient;2 none of the others had ventricle dilatation or contralateral lesions. Our patient had the first clinically documented symptomatic xanthogranuloma with significant unilateral hydrocephalus and a radiologically documented contralateral lesion.

Xanthomatous Third Ventricular Lesions

The 12 patients in whom such lesions were found in the third ventricle are usually lumped together with cases of xanthogranuloma of the trigone. Nonetheless, their features differ in many respects. Some were designated as xanthogranulomas,4,5,10,13,15,19 others as xanthogranulomatous colloid cysts16 or combined colloid cysts and xanthogranulomas.1,18 The patients' ages ranged between 18 and 67 years, with an equal sex distribution; headache and papilledema were the most common symptoms. These lesions were more often lethal than were those in the trigone.

All of the lesions in the third ventricle were cystic, in addition to having xanthomatous changes. Most showed distinct histological patterns which differed from the typical features of xanthogranulomas. Microscopically, the tumors are often described as being cystic and lined by a single- or a multilayered ciliated, cuboidal epithelium. They had also xanthomatous components such as cholesterol crystal clefts, foreign-body giant cells, foamy cells, and inflammatory infiltrates of varying degree. By comparison, classic xanthogranulomas of the lateral ventricles are noncystic lesions without an epithelial component, and no autopsy study has yet recorded them as an incidental finding in the third ventricle. There is good reason, therefore, to distinguish bilateral xanthogranulomas of the glomus from the xanthomatous cystic lesions of the third ventricle. Indeed, the latter may represent xanthomatous change in the well-known third ventricular colloid cysts.

Radiological Studies

Xanthogranulomas may be documented clinically by CT or MR imaging, as in our patient. Bilateral masses located in the trigone of the lateral ventricles with areas of central lucency and marked marginal enhancement were thought to be highly suggestive.7 In contrast, Zimmerman and Bilaniuk23 described a xanthogranuloma as a small area of intraplexal fat without enhancement.

Conclusions

Two important aspects concerning the diagnosis of xanthogranulomas should be stressed. First, the lesion

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should clearly correspond to the clinical and radiological findings of the patient to exclude a coincidental nonsymptomatic xanthogranuloma. Second, xanthogranulomas of the lateral ventricles must be distinguished from cystic lesions with xanthomatous features in the third ventricle, which are more akin to colloid cysts.

References


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