Child's Nervous System
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CASE REPORT

Symptomatic bilateral choroid plexus xanthogranuloma in a pediatric patient

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Abstract

Introduction Central nervous system xanthogranulomas are uncommon clinical entities, and symptomatic bilateral choroid plexus xanthogranulomas are rare.

Case presentation We present the case of a 15-year-old male patient with bilateral choroid plexus xanthogranulomas with symptoms of increased intracranial pressure. Gross total resection of the tumor in the left lateral ventricle was performed. The patient improved, and asymptomatic right-sided tumor was monitored at follow-up.

Conclusion The main treatment objective in these tumors is gross total resection. Asymptomatic tumors can be followed without intervention. However, surgery should be performed for symptomatic tumors that cause hydrocephalus or symptoms of increased intracranial pressure.

Keywords Choroid plexus · Xanthogranuloma · Intraventricular tumor · Pediatric

Introduction

Xanthogranulomas are benign idiopathic histiocytic neoplasms characterized by cholesterol clefts, hemosiderin deposits, macrophages (xanthoma cells), lymphocytic infiltration, and fibrous proliferation [1–4]. They are usually seen as skin lesions in children up to 2 years of age. Central nervous system xanthogranulomas are rare clinical entities and usually asymptomatic. The incidence of central nervous system xanthogranulomas is 1.6–7% in autopsy series [2, 4, 5]. The third ventricle, lateral ventricles, suprasellar and parasellar regions, cerebral and cerebellar parenchyma, the fourth ventricle, and dura are the locations of central nervous system xanthogranulomas [3]. This report describes a rare case of a 15-year-old boy with symptomatic bilateral xanthogranulomas of the choroid plexus and presents a comprehensive literature review.

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Case report

A 15-year-old male patient was admitted to our clinic with altered consciousness, headache, and polyuria. There was no recent history of trauma, tumor, or neurological disease. However, he had a history of oral desmopressin use between 5 and 12 years of age due to diabetes insipidus. The drug was discontinued at the age of 12 for an unknown reason, and he did not continue outpatient controls. On neurological examination, the patient was lethargic, his pupils were equal and reactive to light, and he had right-sided hemiparesis. His blood tests revealed hypernatremia. He was assessed in the pediatric endocrinology clinic, where he was diagnosed with diabetes insipidus; oral desmopressin medication was administered.

Computed tomography brain scans showed bilateral hypodense lesions in the trigone of the lateral ventricles (Fig. 1a). A brain magnetic resonance imaging (MRI) scan revealed bilateral, well-circumscribed, lobulated, hypointense (on T1- and T2-weighted images), homogeneously enhancing mass lesions filling the temporal and occipital horns of the lateral ventricles (Fig. 1b–d). The mass in the left lateral ventricle was 55 × 42 × 26 mm, and the mass in the right ventricle was 35 × 20 × 19 mm. Ventricular dilatation or hydrocephalus was not detected.

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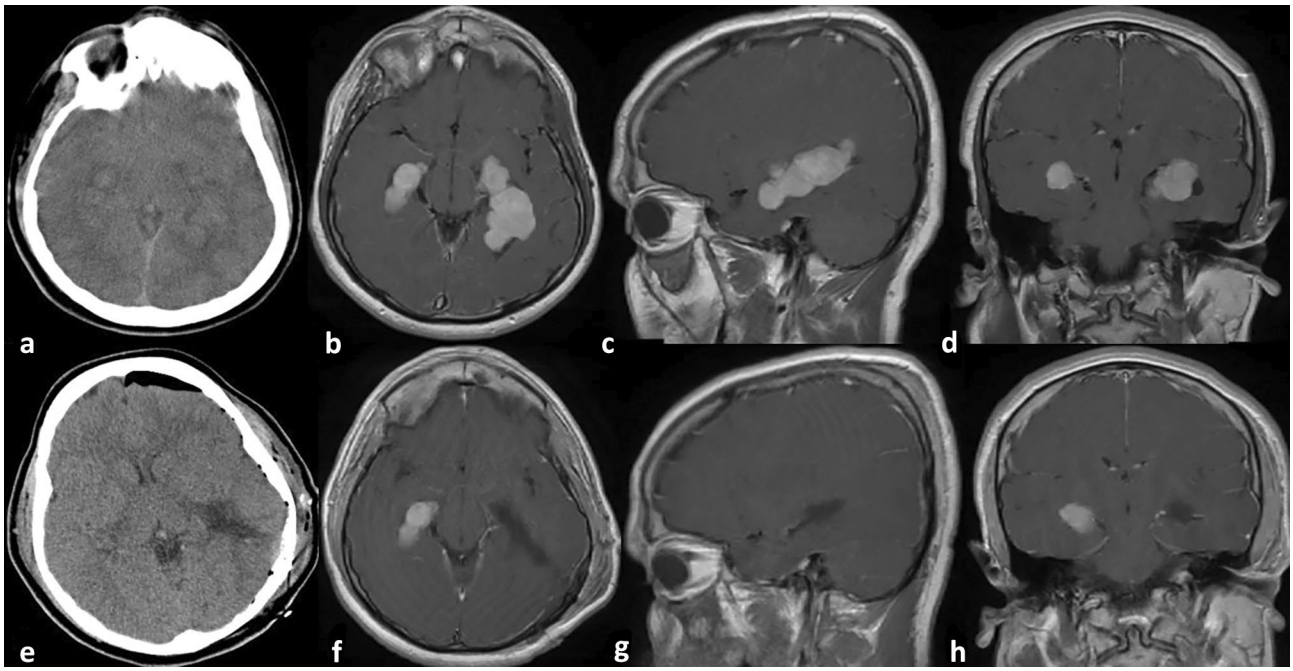

Fig. 1 **a** Preoperative axial computed tomography scan revealing bilateral hypodense lesions in the trigones of the lateral ventricles. Preoperative **b** Axial, **c** sagittal, and **d** coronal contrast-enhanced magnetic resonance images revealing bilateral, well-circumscribed, lobulated, homogeneously enhancing mass lesions filling the temporal and occipital horns of the lateral ventricles. Postoperative **e** axial computed tomography scan and **f** axial, **g** sagittal, and **h** coronal contrast-enhanced magnetic resonance images revealing gross total resection of the left lateral ventricular tumor

Surgical resection of the tumor in the left lateral ventricle was planned because of its size and the presence of right hemiparesis. Gross total resection of the tumor in the left lateral ventricle was performed through a transcortical transventricular approach with a neuronavigation system (Navient image-guided navigation system, ClaroNav Kolahi Inc., Toronto, Canada). Histopathological examination revealed a tumor composed of foamy histiocytes, lymphocytes, and cholesterol clefts under the choroid plexus epithelium (Fig. 2). Immunohistochemically, CD68 was positive in histiocytes, and keratin was negative. Histological and immunohistochemical findings were compatible with choroid plexus xanthogranuloma.

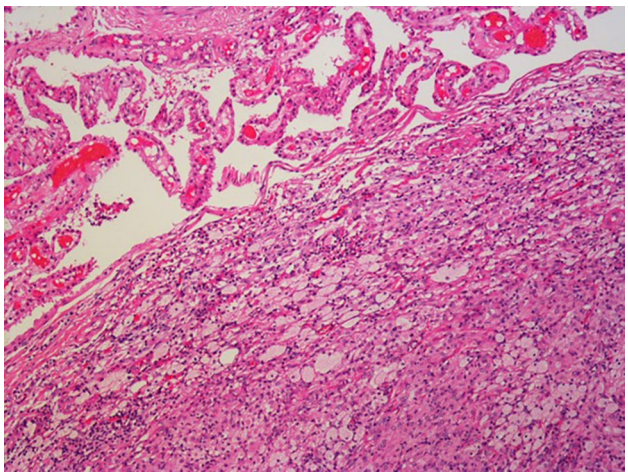

Fig. 2 Foamy histiocytes mixed with lymphocytes are present under choroid plexus epithelium (H&E, × 100 magnification)

The patient's postoperative course was uneventful, and his altered consciousness, headache, and right hemiparesis improved. Diabetes insipidus was controlled with oral desmopressin. The postoperative MRI scans revealed total tumor resection in the left lateral ventricle (Fig. 1f–h). The patient's symptoms completely resolved, and the tumor in the right ventricle continued to be stable in control MRI scans (Fig. 1f–h). For this reason, the second session for this lesion was postponed until it became symptomatic. He had had no recurrence or new complaints at 9 months of follow-up.

Discussion

The first case of choroid plexus xanthogranuloma was described by Blumer in 1900 [6]. Xanthogranulomas are rare intracranial lesions usually detected incidentally [5, 7]. Most are asymptomatic. However, lesions located in the third ventricle can cause obstructive hydrocephalus, and

Table 1 Data of the patients with bilateral choroid plexus xanthogranulomas in the literature

Authors, year	Age (years)/gender	Presenting symptoms	Radiological findings	Treatment	Outcome (follow-up)
Terao et al. [14]	6, F	Seizure	Bilateral lateral ventricular calcified masses	GTR via craniotomy in 2 sessions	No recurrence (1 month)
Brück et al. [9]	50, M	Headache, amnesic aphasia, right hemiparesis, papilledema	Bilateral lateral ventricular masses + left-sided ventricular dilatation	Left-sided GTR via craniotomy + right-sided tumor followed	Symptoms improved, HCP resolved after operation (NA)
Gaskill et al. [10]	6, M	Headache, seizure, lethargy, fasial palsy, left hemiparesis, atrophic optic disks	Bilateral lateral ventricular masses + right-sided ventricular dilatation	GTR via craniotomy in 2 sessions	Symptoms improved except vision loss, no recurrence (9 years)
Mendez-Martinez et al. [12]	6, F	Visual disorder, pale optic disks	Bilateral lateral ventricular masses + optic chiasm compression	Biopsy via craniotomy + radiosurgery	Vision loss remained the same, stable tumor size (7 months)
Jain and Mehta [11]	8, M	Headache, nausea	Bilateral lateral ventricular masses + right-sided ventricular dilatation	Right-sided GTR via craniotomy + left-sided tumor followed	Symptoms improved, HCP resolved after operation (NA)
Moreau et al. [5]	50, F	Incidentally at postmortem examination	NA	NA	NA
Tovar-Spinoza and Choi [15]	4, M	Headache	Bilateral lateral ventricular masses	Magnetic resonance-guided laser interstitial thermal therapy	No recurrence (26 months)
Emon et al. [1]	47, M	Headache	Bilateral lateral ventricular masses	GTR via craniotomy in 2 sessions	No recurrence (6 months)
Shuangshoti [13]	12, M	Seizure, diabetes insipidus, nausea, atrophic optic disks	Bilateral lateral ventricular masses	GTR via craniotomy in 1 session	Died immediately after operation
AlQazlan et al. [8]	9, M	Headache, nausea, lethargy, diabetes insipidus	Bilateral lateral ventricular masses + bilateral ventricular dilatation	GTR via craniotomy in 2 sessions*	No recurrence (12 years)
Patel et al. [7]	1 m, M	Incidentally (neonatal meningitis)	Bilateral lateral ventricular masses + bilateral ventricular dilatation	NA	NA
Present case	15, M	Headache, lethargy, right hemiparesis, diabetes insipidus	Bilateral lateral ventricular masses	Left-sided GTR via craniotomy + right-sided tumor followed	No recurrence (9 months)

*Diffuse dural recurrence in the right cerebral hemisphere occurred 6 years after the operations and GTR via craniotomy was performed

F female, *M* male, *GTR* gross total resection, *NA* not available, *HCP* hydrocephalus

large intracranial lesions can cause symptoms of increased intracranial pressure, as in this case [2, 3, 8–14].

Xanthogranulomas, also known as cholesterol granulomas, are often yellow, soft, encapsulated lesions. In our case, the lesion borders were distinctly firm and could be separated from the surrounding tissues. Xanthogranulomas consist of inflammatory cells such as cholesterol clefts, foam cells, histiocytes, hemosiderin deposits, and fibrous proliferation. These characteristics can produce a pseudotumor appearance. Intralesional bleeding, fibrosis, and focal calcifications are secondary changes that can also occur [5]. The preoperative diagnosis of xanthogranulomas can be very challenging and is based on clinical information and radiographic findings. Preoperatively diagnosing these lesions can be challenging due to the lack of typical distinguishing radiological signs. These lesions are hypo or isodense on computed tomography scans. Brain MRI is the gold standard for choroid plexus xanthogranulomas. These lesions are hypo or isointense on T1-weighted images, hypointense on T2-weighted images, and present heterogeneous contrast enhancement with gadolinium [1, 5, 11]. Therefore, differential diagnosis with other lateral ventricle masses such as meningioma, papilloma, ependymoma, and arteriovenous malformations is difficult [5].

The main treatment modality for choroid plexus xanthogranulomas is surgical resection. In patients with hydrocephalus or increased intracranial pressure, surgical resection should be performed immediately. The aim of this surgical treatment should be gross total resection of the tumor in appropriate cases [1, 8–11, 14]. Tovar-Spinoza et al. presented the successful application of magnetic resonance-guided laser interstitial thermal therapy for choroid plexus xanthogranuloma in a 4-year-old child [15]. However, there is no widely accepted adjuvant treatment modality for these lesions.

There are 11 cases of bilateral choroid plexus xanthogranulomas reported in the literature (Table 1). Two were found incidentally, and nine were symptomatic patients. According to our literature review, our case is the seventh pediatric case of symptomatic bilateral choroid plexus xanthogranuloma and the third case with diabetes insipidus (Table 1).

Conclusion

Bilateral choroid plexus xanthogranulomas are uncommon, rarely symptomatic tumors. Clinicians should be aware of these lesions in the differential diagnosis of intraventricular tumors. Asymptomatic tumors can be followed without any intervention. However, surgical intervention should be performed for symptomatic tumors that cause hydrocephalus or increased intracranial pressure, and gross total resection should be the aim.

Declarations

Conflict of interest The authors declare that they have no conflict of interest.

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