Chordoid glioma of the third ventricle

Clinical History:

A 20-year-old woman with occasional sharp bilateral headache for 2 years, effectively controlled with analgesics. Lately, headache intensity has gradually increased and symptoms of nausea and vomiting started, neither of which could be controlled with treatment. Neurological examination demonstrated visual field defects, superior nasal quadrantanopia in the right eye and superior temporal quadrantanopia in the left eye.

Imaging Findings:

The unenhanced brain cranial tomography (CT) performed detected a polylobulated, hyperdense and discretely heterogeneous suprasellar solid mass (Figure 1). This finding was associated with left lateral ventricle dilation secondary to the occlusion of the ipsilateral foramina of Monro. The magnetic resonance imaging (MRI) carried out, clarified that the centre of the 30x27x20 mm lesion was located on the third ventricle anterior recess. T-1 weighted images (Figure 2A) show a slightly heterogeneous isointense mass and in T-2 weighted images (Figure 3) it is hypointense with peripheral hyperintense areas. After paramagnetic contrast intravenous injection, the lesion demonstrates intense heterogenous uptake (Figure 2B-D). Hypothalamus and optic chiasm are compressed and displaced inferiorly and laterally because of the tumour. Optic tracts showed diffuse increased uptake in relation with oedema (Figure 3B).

Subfrontal interhemispheric approach through lamina-terminalis was performed identifying a bloody brownish mass adhered to the walls of the third ventricle that was completely removed (Figure 4).

Discussion:

Chordoid glioma features were first described in 1995 by Wanschitz et al, reporting a suprasellar meningioma that contained glial fibrillary acidic protein (GFAP) positive cells [1]. Three years later, this meningioma variant was codified by Brat in a 8 cases and reported as a new tumour entity typically located in the third ventricle with well-defined histological and immunophenotypic characteristics [2]. It is considered a slow-growing, non-invasive, rare tumour with glial and chordoid features of low malignant potential (2007 classification: grade II malignant glioma) with its origin in the third ventricle [3].

The origins of the chordoid glioma according to its clinical, radiological and histological features could be the ependymal surface of the vasculosum organ of the lamina terminalis and the hypothalamic nuclei [4]. Although most cases were found at the typical location [5], three of them were described at atypical places: one in an adult [6]
and the other two in children [7, 8].

This entity affects adults predominantly, median age of diagnosis is 46 years old [9], showing a clear predilection for women (female/male ratio 2:1), although there are a few cases reported in children [7, 8, 10] and elderly people [2, 11, 12, 13].

Patients present with hydrocephalia (25%) [5, 9, 14] or a wide variety of non-specific symptoms secondary to its location and bordering structures affection. The most common clinical manifestations include headache, visual disturbances, behavioural and endocrinological alterations (Diabetes insipidus, amenorrhea and hypothyroidism)[5, 9].

From a radiological point of view, this kind of tumour is described as a hyperdense well-defined ovoid mass mainly located on the third ventricle in unenhanced CT, that because of its signal could be mistaken for a lymphoma, meningioma or even an aneurysm [15, 16]. MRI imagining characteristics of chordoid glioma are described as isointense on T-1 and hyperintense on T-2, but also isointense or heterogeneous on T-2; after the gadolinium administration the uptake is homogeneous [14, 15, 17]. Most of the tumours are solid but up to 25% can be manifested with little cystic central areas [9, 15]. Calcifications, perilesional and optic tracts oedema are frequently described [14, 15, 16].

Usually the first choice of treatment consists of a surgical subfrontal interhemispheric approach through lamina-terminalis, showing a very low recurrence rate. After subtotal resection surgery, early and late recurrence rate increase, so for these cases it seems appropriate to add adjuvant radiation treatment, particularly gamma-knife radiosurgery [5, 18].

**Differential Diagnosis List:** Chordoid glioma of the third ventricle, Chordoid glioma of the third ventricle, Craniopharyngioma

**Final Diagnosis:** Chordoid glioma of the third ventricle

**References:**


**Description:** Sagital T1-weighted SE image shows an ovoid mass circumscribed to the anterior region of the third ventricle (intraventricular) displacing but not infiltrating surrounding tissues. **Origin:** Romeu D, Department of Radiology, Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.
Description: Postcontrast sagittal T1-weighted SE evidence a heterogeneous lesion uptake displacing posterior infundibulum (yellow arrow) Origin: Romeu D, Department of Radiology, Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.
Description: Postcontrast coronal T1-weighted SE evidence a heterogeneous lesion uptake. The superoinferior orientation of the mass blockades the left foramina of Monro. Origin: Romeu D, Department of Radiology, Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.
Description: Axial T1-weighted SE evidence a heterogeneous lesion uptake on the third ventricle.
Origin: Romeu D, Department of Radiology, Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.
**Description:** Unenhanced CT showing a hyperdense mass apparently suprasellar secondary occluding left foramine of Monro. **Origin:** Romeu D, Department of Radiology, Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.
Description: Axial T2 weighted SE image demonstrate a heterogenous solid mass located on the third ventricle accompanied by perilesional and optical tracts edema (yellow arrows). Origin: Romeu D, Department of Radiology, Complejo Hospitalario Universitario A Coruña, A Coruña Spain.
Description: Axial T1 weighted SE image after contrast administration showing the complete tumor excision. Origin: Romeu D, Department of Radiology, Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.
Description: Histologically the tumor is characterized by cords of epithelioid cells with abundant eosinophilic cytoplasm. Origin: Álvarez Martínez, M. Department of Pathology Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.
**Description:** Other histological features include blue mucinous vacuolated stroma containing a lymphoplasmocytic infiltrate. **Origin:** Álvarez Martínez, M. Department of Pathology Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.
Figure 6

Description: The most characteristic immunohistochemical feature is a strong and diffuse reactivity for glial fibrillary acidic protein (GFAP). Origin: Álvarez Martínez, M. Department of Pathology Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.
**Description:** Staining for vimentin is also strong. **Origin:** Álvarez Martínez, M. Department of Pathology Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.

**Description:** Epithelial membrane antigen (EMA) staining is seen focally but it is more prominent in stromal plasma cells. **Origin:** Álvarez Martínez, M. Department of Pathology Complejo Hospitalario Universitario A Coruña, A Coruña, Spain.