

Atypical choroid plexus papilloma

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Section: Neuroradiology

Area of Interest: Neuroradiology brain Education

Procedure: Education

Imaging Technique: MR

Special Focus: Pathology Case Type: Clinical Cases

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Patient: 13 years, female

Clinical History:

A teenage patient presented with a history of 2 years of symptoms characterised by a frontal headache accompanied by night awakenings. However, paresthesias began on the left side of the head during the past week. Then, she started with an acute central facial palsy, positive Romberg and nystagmus.

Imaging Findings:

Brain MRI demonstrates a fourth ventricular mass. The lesion mass is isointense on T1WI, hyperintense on T2WI and FLAIR, no diffusion restriction, foci of haemorrhage seen on SWAN, spread into the foramen of Luschka and demonstrates avid enhancement on postcontrast sequences.

There is ventricular enlargement consistent with communicating hydrocephalus.

Discussion:

Choroidal plexus tumors originate in the epithelium of the choroid plexus, are located intraventricularly, and are classified by The World Health Organization according to their pathological findings, grade I to refer to papilloma, grade II atypical papilloma and grade III when there is carcinoma. [1, 2]

Choroidal plexus papillomas (CPP) are uncommon, they occur predominantly in the paediatric population, representing from 2% to 5% of intracranial tumours and 0.4-0.6% of all intracranial neoplasms in the same population, and they are usually unilateral, however, occasionally they can be found bilaterally. [1, 2]

On imaging, CPP are found most commonly in the fourth and lateral ventricle, on T1 they are typically isointense, on T2 iso to hyperintense and characterised by vividly enhancing mass sometimes with frond-like morphology. The mainly affected location are the lateral ventricles in young children, most of them under two years of age. The fourth ventricle is the least frequent in children, but it is most common in adults. [3] Intraventricular localisation in the third ventricle and cerebellopontine angle are less frequent. [1, 2]

Less common locations are in the foramen of Luschka extending to extraventricular areas, and in some cases, it can be predominantly extraventricular. [4]

In its clinical presentation, hydrocephalus is the most common clinical manifestation; it is produced by obstruction or overproduction of cerebrospinal fluid, mainly when there is a compromise of the third and fourth ventricle. [1, 2]

The differential diagnoses on magnetic resonance imaging are meningioma, haemangioblastoma, metastasis, and neurinoma. The meningioma is very difficult to differentiate from CCP, but the meningioma has a broad dural base, iso-signal intensity on T2-weighted images, and homogeneous enhancement. [2]

They can be classified histologically, well-differentiated and atypical as the case of our patient; they are characterised by high cellularity, greater cytoplasm nucleus ratio, and epithelial cell conglomeration. Although they are still considered as benign, this category helps to recognise its aggressiveness, and thus allows to establish management strategies. [1]

The choice of management described in the literature is surgical removal, in conjunction with radiotherapy according to some authors. [2, 4, 5]

Regarding prognosis, the event-free survival rate is 92% in the choroid plexus papilloma and around 83% in the case of atypical papillomas. [4]

Differential Diagnosis List: Atypical choroid plexus papilloma, Meningioma, Ependymoma, Haemangioblastoma

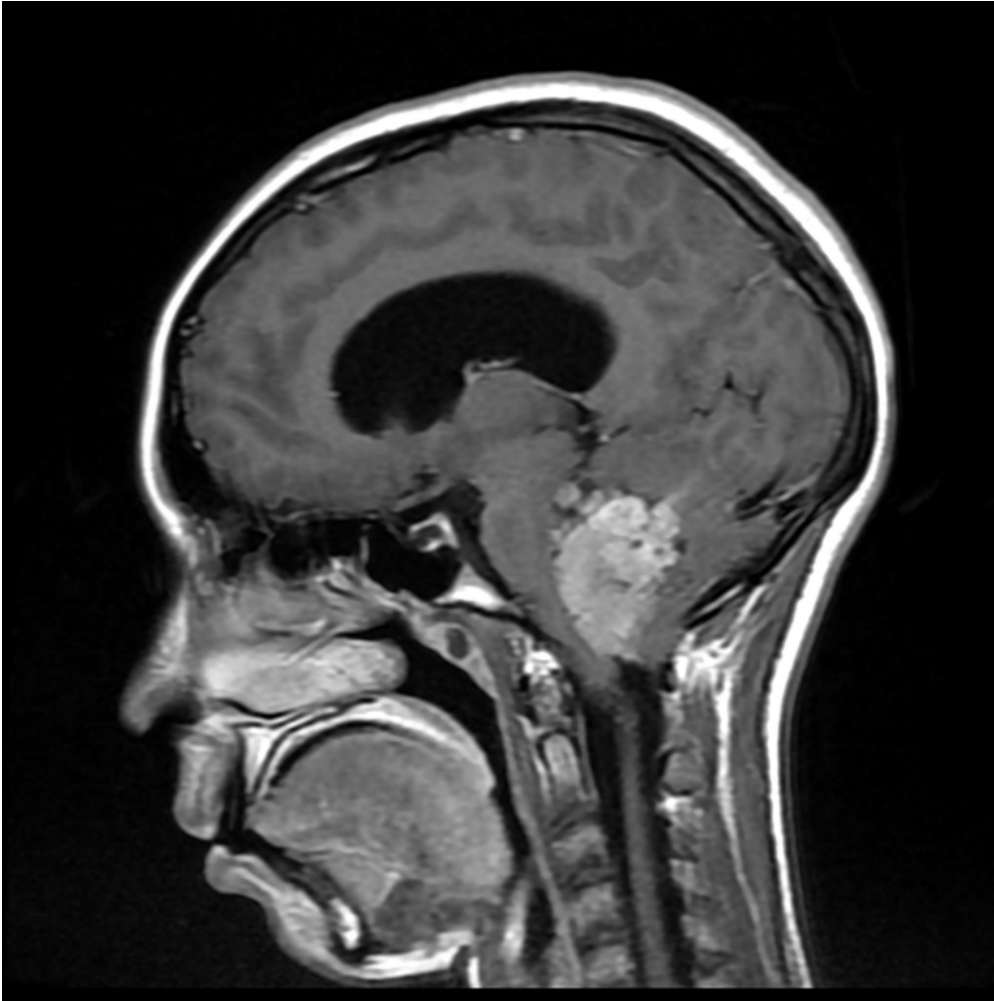
Final Diagnosis: Atypical choroid plexus papilloma

References:

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- Bostrom,A.B.,J.VonLehe,M.Kandenwein,J.Schramm,J.Simon,M (2011) Surgical treatment of choroid plexus tumors. Acta neurochir 153:p.371-376 (PMID: [20936311](#))
- Kumabe,T.F.,M.Jokura,H.Tominaga,T (2008) Surgical treatment for choroid plexus tumors in the fourthventricle:brainstem infiltration hinders total extirpation. Neurosurgery 31:p.165-172. (PMID: [17912563](#))

Figure 1

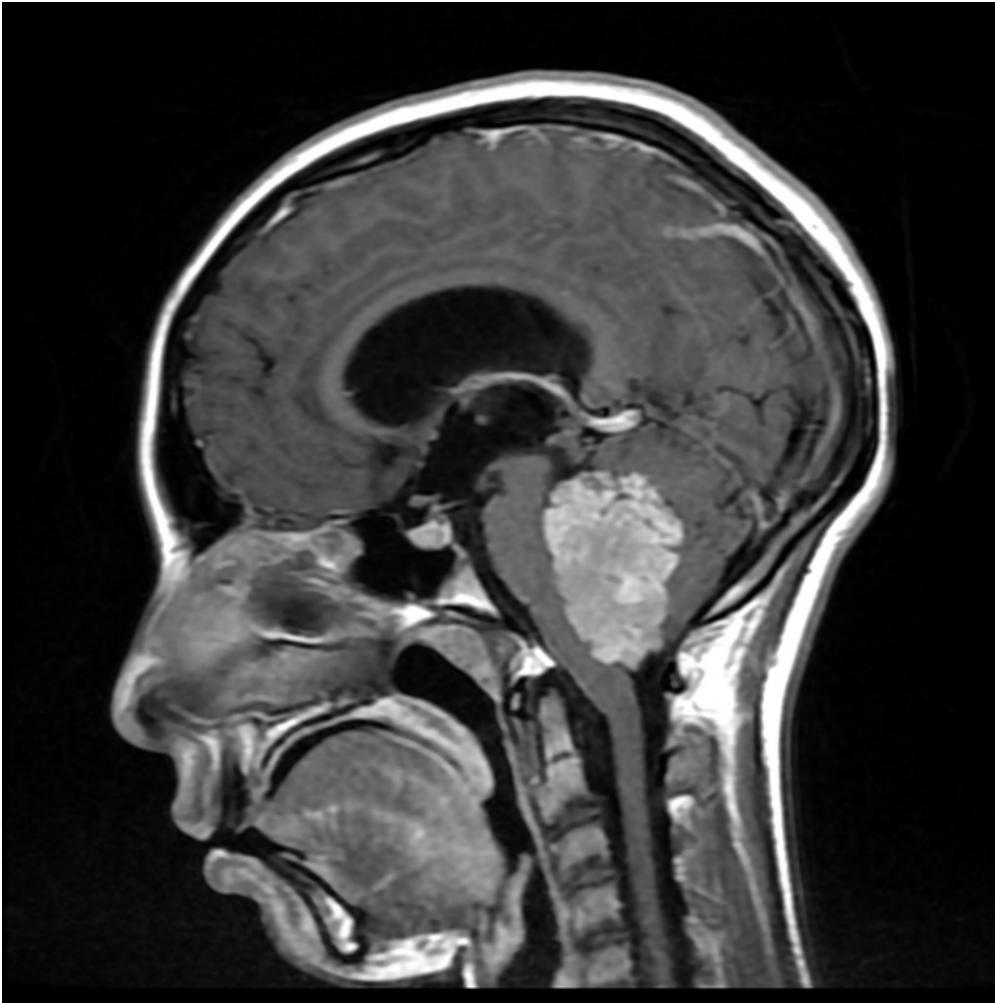
a



Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.

Origin: Hospital Militar Central

b



Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.

Origin: Hospital Militar Central

c



Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.
Origin: Hospital Militar Central

d



Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.

Origin: Hospital Militar Central

Figure 2

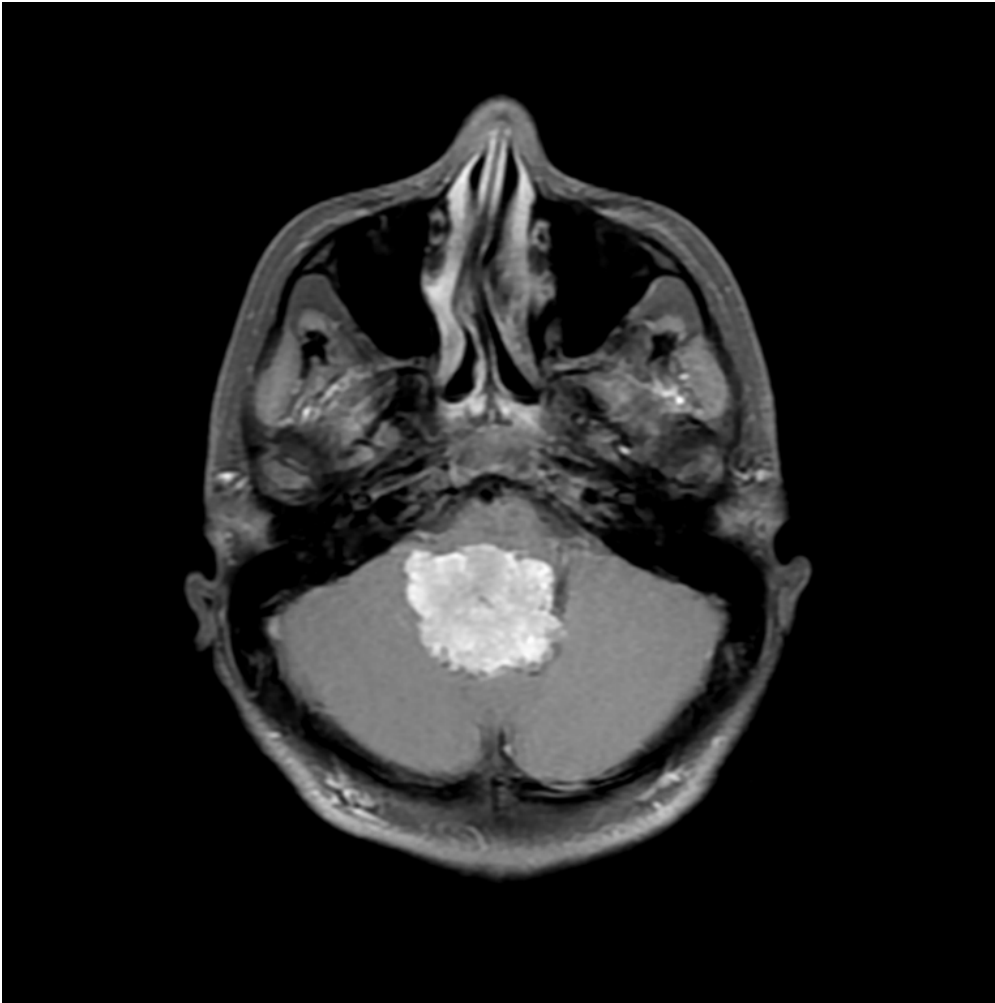
a



Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.

Origin: Hospital Militar Central

b



Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.

Origin: Hospital Militar Central

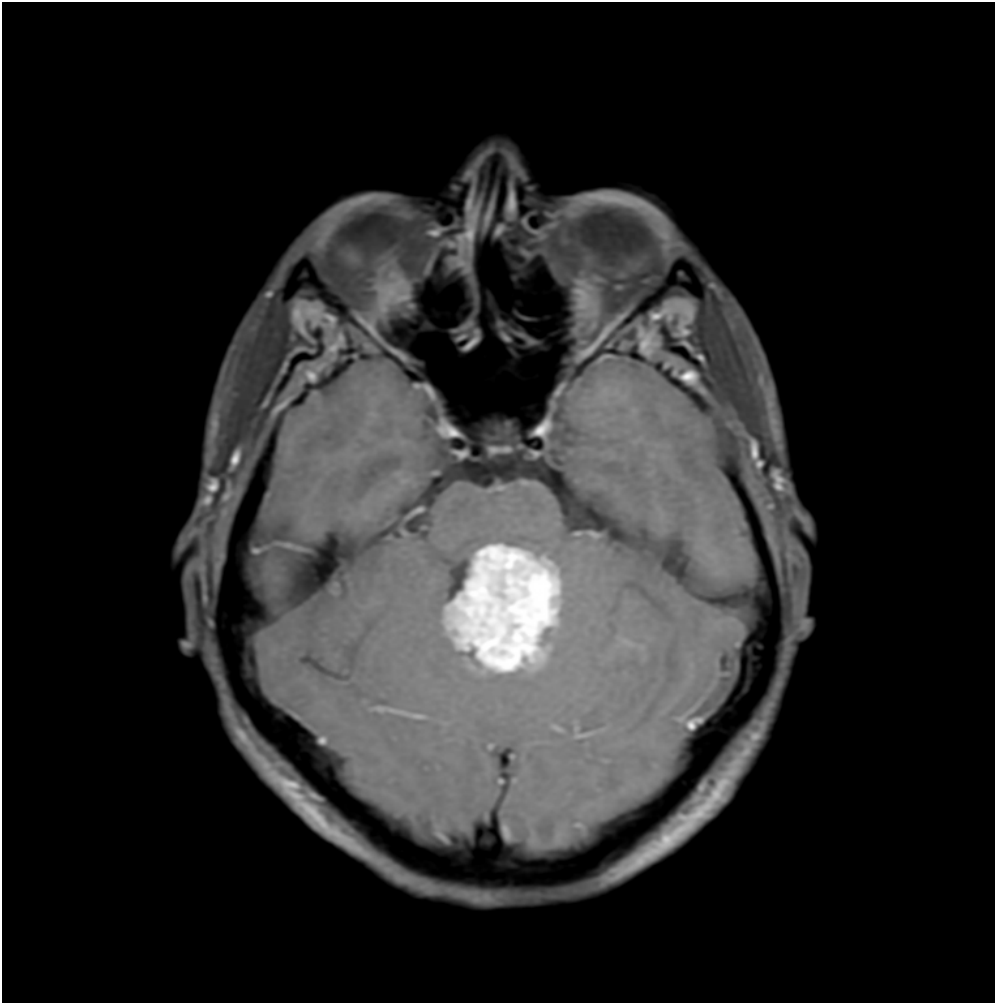
c



Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.

Origin: Hospital Militar Central

d

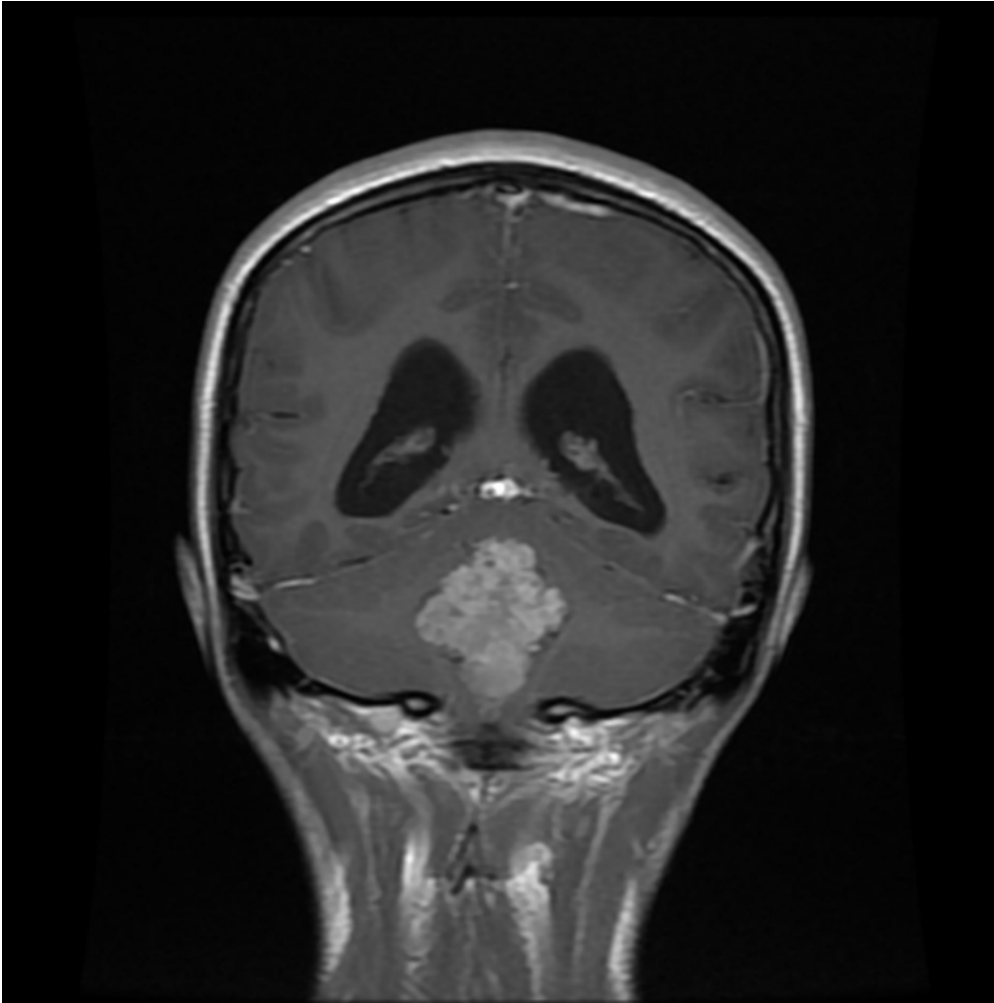


Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.

Origin: Hospital Militar Central

Figure 3

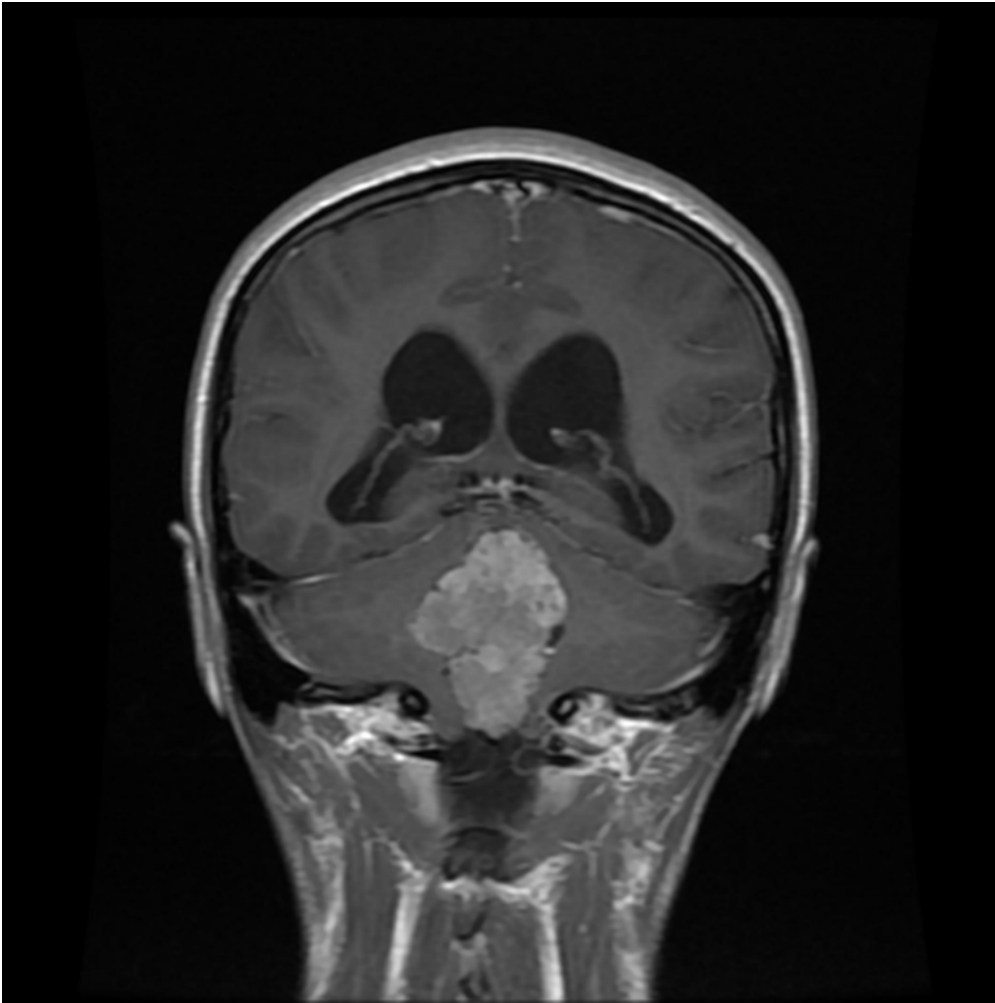
a



Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.

Origin: Hospital Militar Central

b

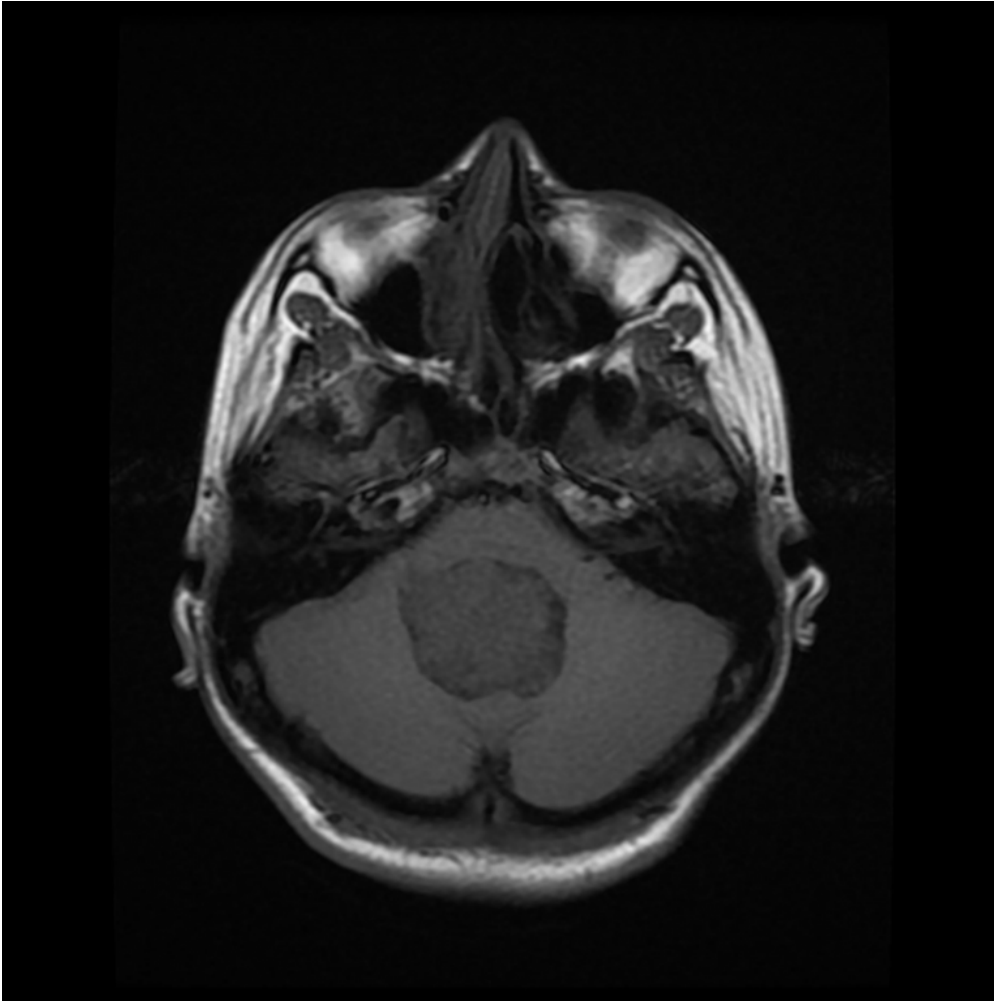


Description: Fourth ventricular mass demonstrates avid enhancement on post-contrast sequences.

Origin: Hospital Militar Central

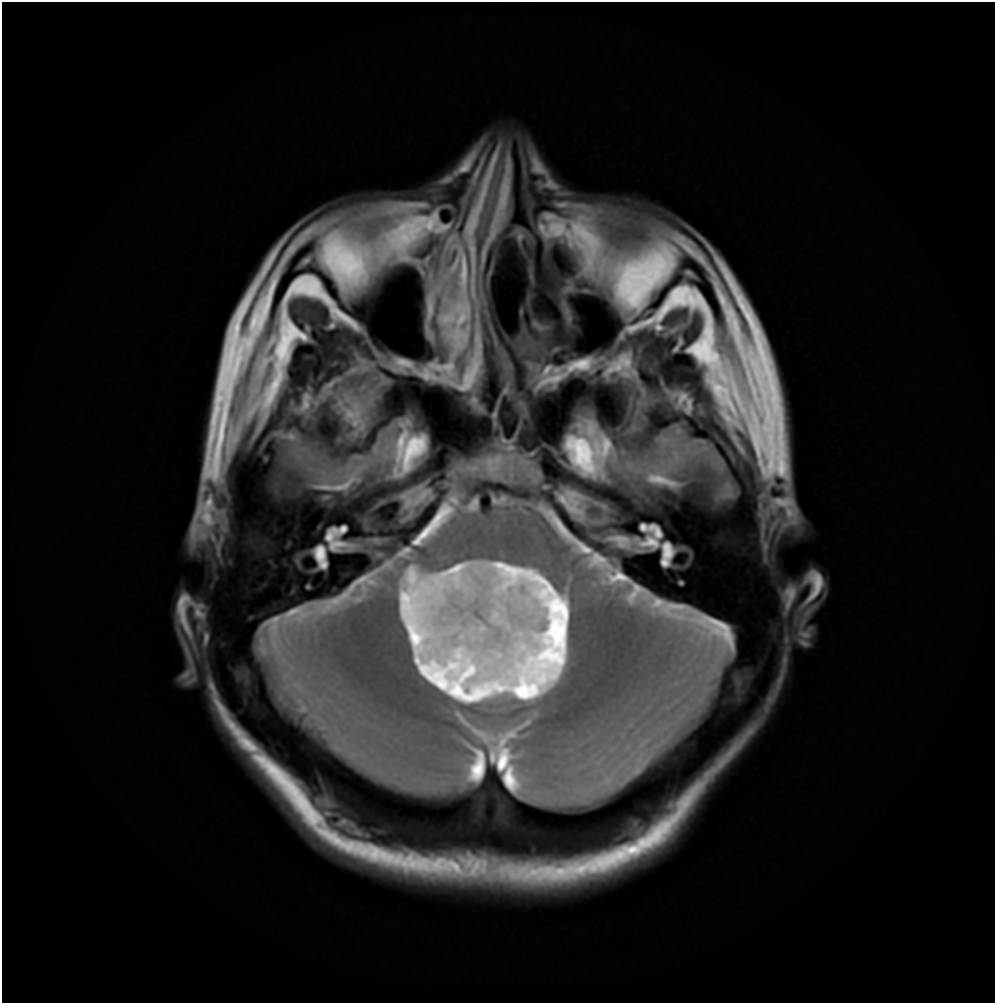
Figure 4

a



Description: Brain MRI demonstrates a fourth ventricular mass. The mass is isointense to gray matter on T1. **Origin:** Hospital Militar Central

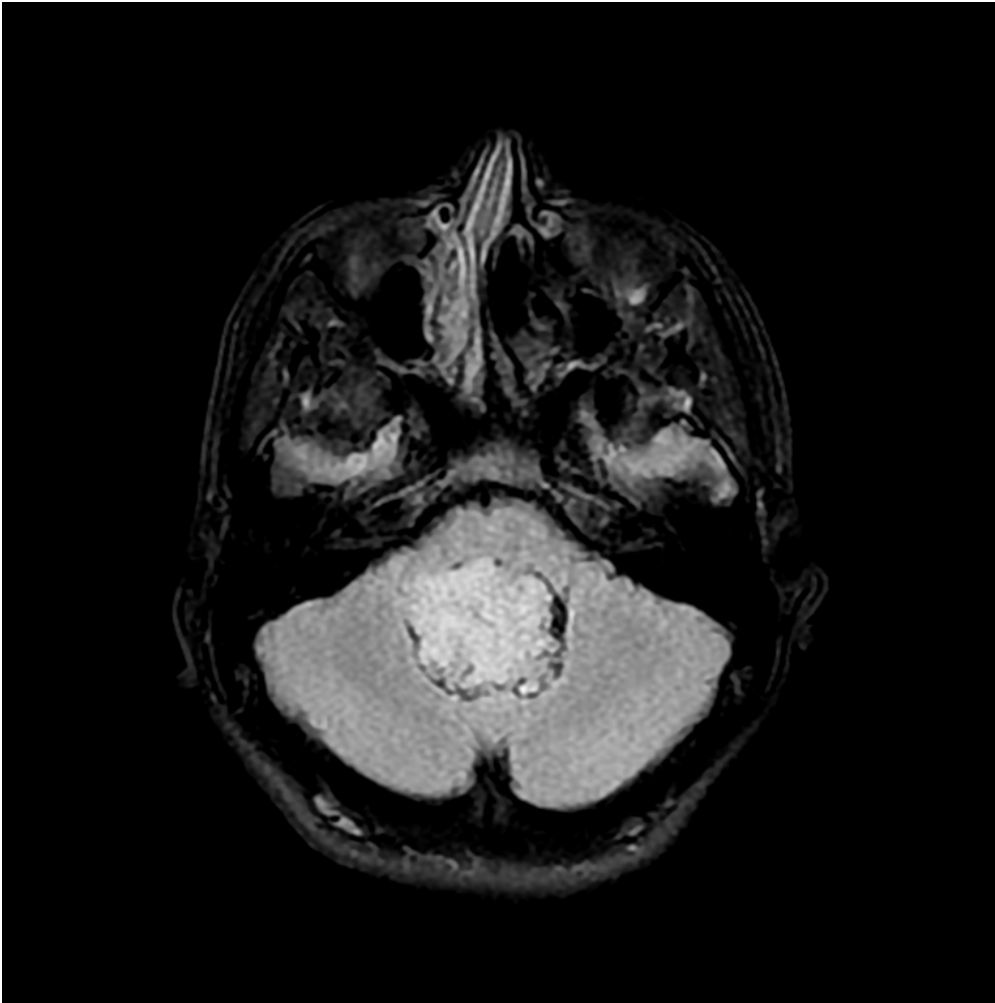
b



Description: Brain MRI demonstrates a fourth ventricular mass. The mass is hyperintense on T2.

Origin: Hospital Militar Central

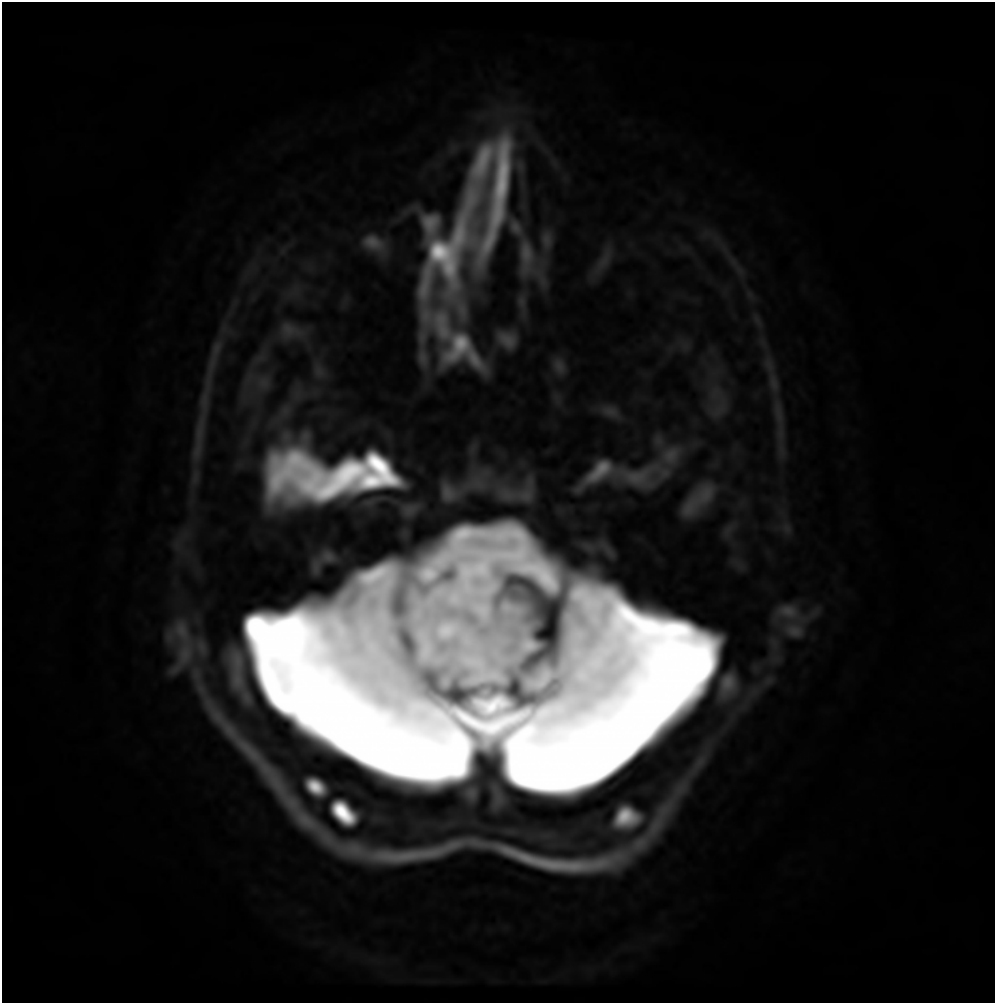
c



Description: Brain MRI demonstrates a fourth ventricular mass. The mass hyperintense on FLAIR.

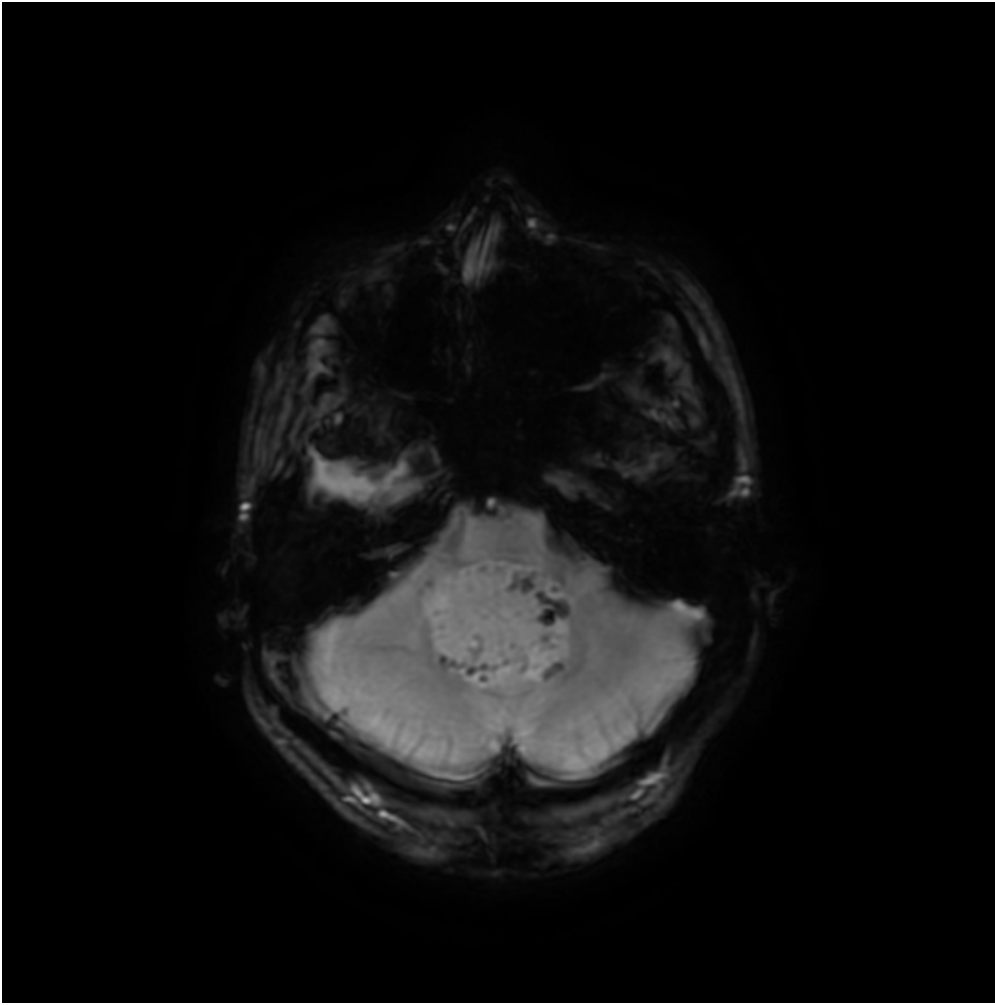
Origin: Hospital Militar Central

d



Description: Brain MRI demonstrates a fourth ventricular mass. The mass does not show diffusion restriction. **Origin:** Hospital Militar Central

e



Description: Brain MRI demonstrates a fourth ventricular mass. The mass has foci of haemorrhage seen on SWAN. **Origin:** Hospital Militar Central