

Adamantinomatous craniopharyngioma

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Section: Neuroradiology

Area of Interest: Neuroradiology brain

Procedure: Diagnostic procedure

Imaging Technique: MR

Imaging Technique: CT

Special Focus: Neoplasia Case Type: Clinical Cases

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Patient: 9 years, male

Clinical History:

Nine-year-old boy with frequent headaches and loss of visual acuity in the past weeks.

Imaging Findings:

On unenhanced CT a hypodense inhomogeneous sellar mass that presents punctate and clumpy peripheral calcifications is typical (Fig. 1 and 2).

On MRI, there is a solid-cystic sellar mass with suprasellar extension occupying the sella, the suprasellar region, the sphenoid sinus and clivus, with intense and heterogeneous enhancement of the solid area and the walls of the cystic zones (Fig. 3, 4, and 5). Its approximate dimensions are 3x3, 6x5 cm (TxAPxCC). There is associated mass effect on adjacent structures.

Discussion:

Craniopharyngiomas are WHO classification grade I tumours that are derived from squamous epithelial remnants of Rathke's cleft. Most involve both sellar and suprasellar regions, but less commonly, they can be located solely within the sella or the third ventricle [3].

The triad of suprasellar location, calcifications and cysts is highly suggestive of craniopharyngioma.

Two distinct clinicopathologic variants are recognised. The adamantinomatous type (our case) is the more common of the two variants and can be seen in both adults and children, but is more common in the latter. This tumour type more frequently is calcified, exhibits necrosis, and contains cholesterol within cysts. The papillary form almost always occurs in adults but still represents only about one-third of adult craniopharyngiomas. It is more often solid, less frequently calcified, and has been reported to have a better surgical outcome than adamantinomatous tumours in the adult [1, 2].

So adamantinomatous craniopharyngiomas (AC) are the most common form and typically have a lobulated contour as a result of usually multiple cystic lesions (arrow, bottom image, Fig. 4). About 90% of craniopharyngiomas are partially cystic. Rarely, the lesion is purely cystic. The cyst contents consist of straw-coloured or oily brownish fluid with variable amounts of cholesterol crystals and triglycerides, which often produces a hyperintense appearance on

unenanced T1WI (yellow arrow, Fig. 3). Cysts with higher water content are more hypointense (red arrow, Fig. 3). Solid components are present, but often form a relatively minor part of the mass, and enhance vividly on both CT and MRI (arrows, Fig. 5, top and bottom image).

Calcifications are seen in 90% of cases, typically stippled and often peripheral in location (NECT images). On MR spectroscopy: cyst contents may show a broad lipid spectrum, with an otherwise flat baseline [3].

On another front, the papillary type has a more uniform CT and MR appearance with less frequent calcification and cyst formation [4, 2].

Treatment: Adamantinomatous lesions may have a surrounding region of gliosis at the interface of the lesion with the brain tissue [4], which can cause the tumour to tightly adhere to brain tissue, often making complete resection impossible [3, 5].

Differential Diagnosis List: Adamantinomatous craniopharyngioma, Pituitary macroadenoma, Intracranial teratoma, Meningioma

Final Diagnosis: Adamantinomatous craniopharyngioma

References:

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Figure 1

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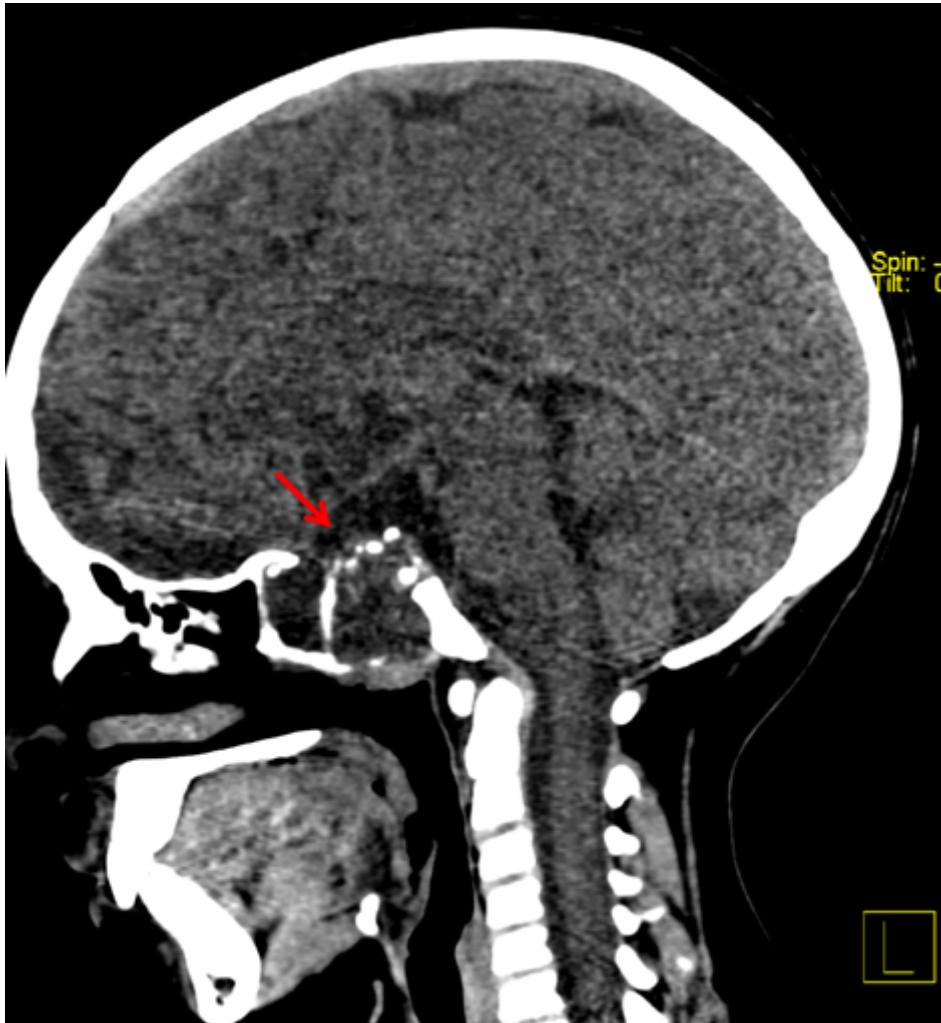


Description: Hypodense sellar mass with suprasellar extension. Clumpy calcifications are visualised.

Origin: Department of Radiology. Hospital Virgen de la Arrixaca, Murcia (Spain).

Figure 2

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Description: Hypodense sellar mass with suprasellar extension (red arrow). Clumpy calcifications are visualised. **Origin:** Department of Radiology. Hospital Virgen de la Arrixaca, Murcia (Spain).

Figure 3

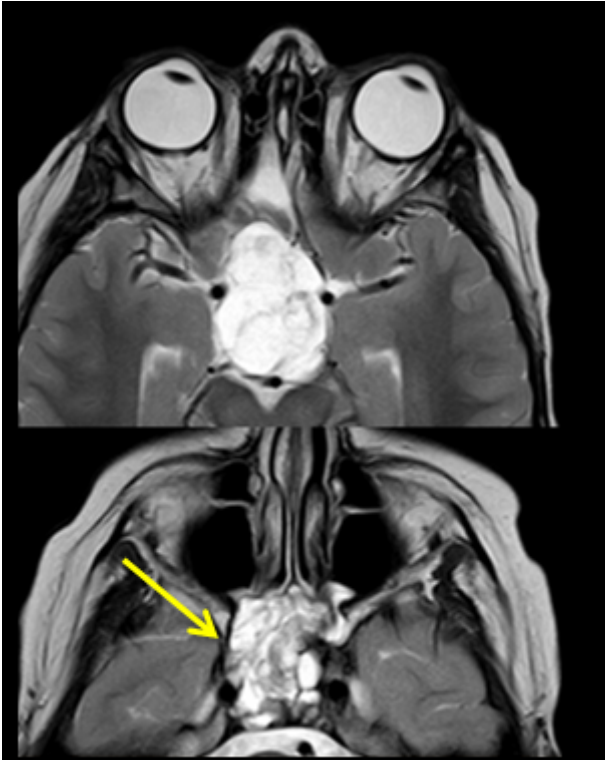
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Description: The cysts content consist of oily brownish fluid with cholesterol crystals, which often produces a hyperintense appearance on T1WI (yellow arrow). Cysts with higher water content are more hypointense (red arrow). **Origin:** Department of Radiology. Hospital Virgen de la Arrixaca, Murcia (Spain).

Figure 4

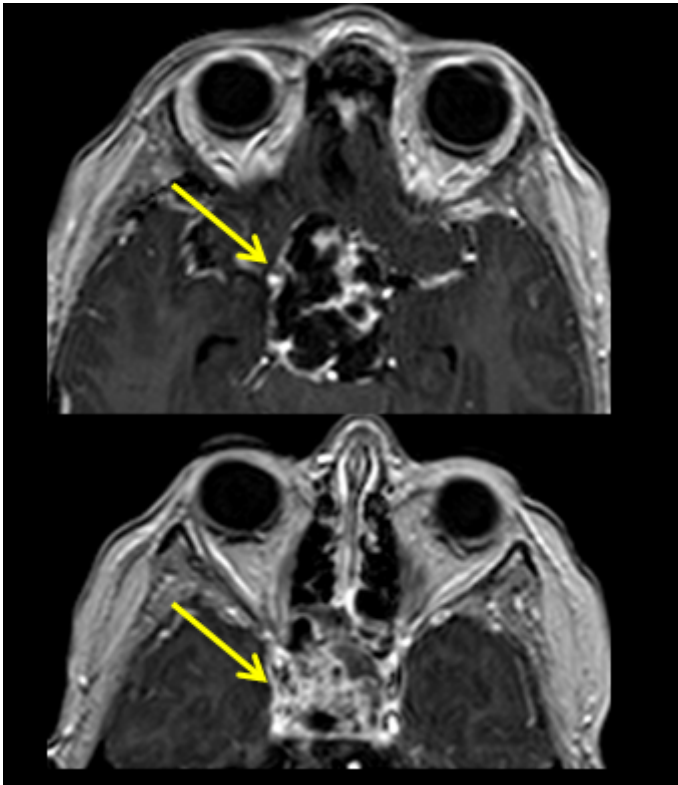
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Description: Adamantinomatous craniopharyngiomas typically have a lobulated contour as a result of usually multiple cystic lesions (arrow). **Origin:** Department of Radiology. Hospital Virgen de la Arrixaca, Murcia (Spain).

Figure 5

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Description: The mass enhances vividly on MRI (arrows). **Origin:** Department of Radiology. Hospital Virgen de la Arrixaca, Murcia (Spain).