Endoscopic Endonasal Management of Sellar Xanthogranuloma: A Three-Case Experience and Review of the Literature

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Introduction

Xanthogranuloma (XG) can be identified in many anatomical sites, however, its occurrence in the sellar region is very uncommon among intracranial lesions. They are a rare begin entity described only in case reports, case series and two systematic reviews. We aim to present a series of three cases of sellar-region xanthogranulomas diagnosed and treated in Brazil, as well as to perform a literature review.

Methods

This study comprises a case series of sellar xanthogranulomas and a literature review conducted through the PubMed database, covering the last 10 years. The review included only articles published in English and used the following search terms: “pituitary xanthogranuloma” OR “sellar xanthogranuloma” OR “suprasellar xanthogranuloma” OR “parasellar xanthogranuloma”.

Results

We report a series of three cases—one male and two females, aged between 17 and 37 years—treated at two care centers in Brazil. Two patients were symptomatic at diagnosis, both presenting hypopituitarism, requiring levothyroxine and glucocorticoids, one of them even had insipidus diabetes, while the third case was identified incidentally and later developed hyperprolactinemia. All lesions involved the sellar and suprasellar regions, with cystic or mixed solid-cystic components, and optic chiasm compression observed in two cases. MRI findings revealed hyperintense signals on T1- and STIR-weighted images, with areas of hypo- and isointensity on T2, and no contrast enhancement. All patients underwent endoscopic endonasal surgery with gross total resection and no recurrence to date. Preoperative endocrinological disturbances persisted postoperatively in all cases, except for one patient who progressed to hypopituitarism requiring lifelong corticosteroid and levothyroxine replacement therapy. The average follow-up period for the cases was 3 years (the longest was 5 years and the shortest was 1 year).

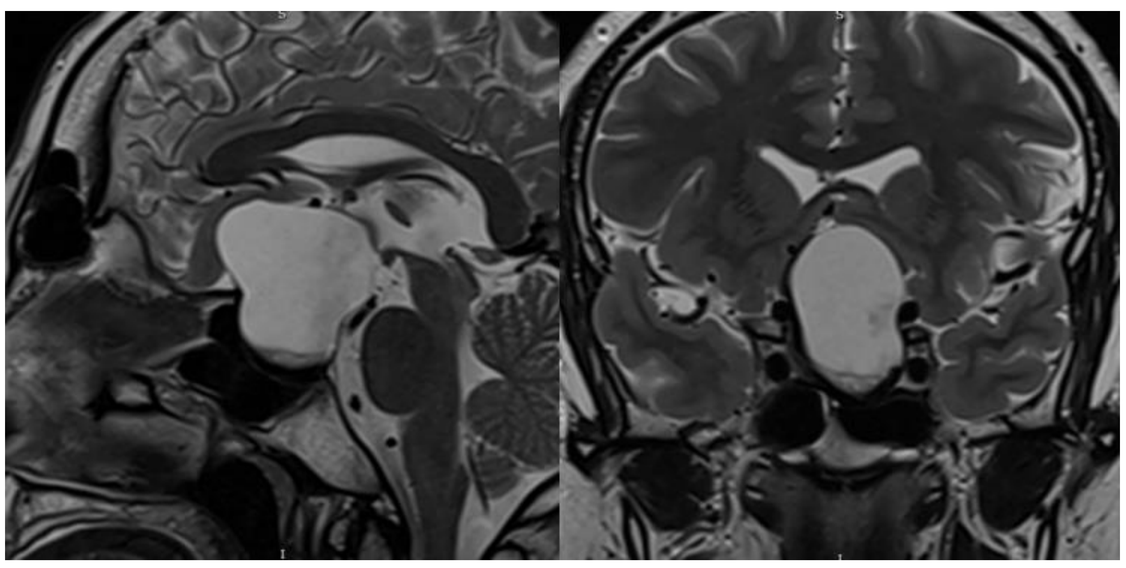

Figure 1. First case - Magnetic resonance imaging of patient 1 showing in sagittal (A) and coronal (B) views a large, predominantly cystic lesion hyperintense on T2, with a small solid hypointense component on T2, exhibiting mass effect and compression of adjacent structure, including the optic chiasm and the anterior communicating complex.

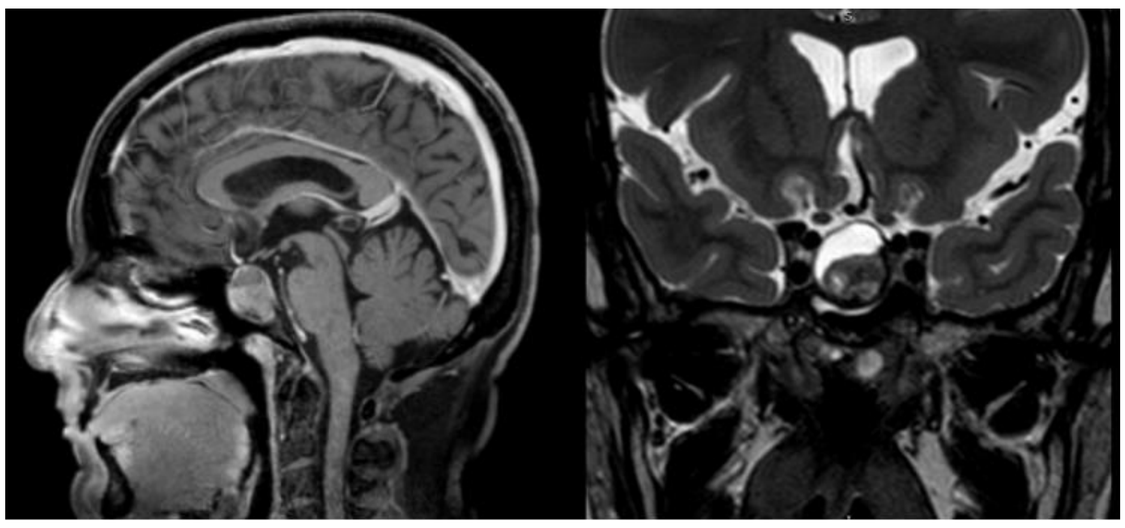

Figure 2. Second case - Magnetic resonance imaging of patient 2 showing in sagittal (A) and coronal (B) views a moderately sized solid-cystic lesion with fluid level, mild mass effect, and heterogeneous contrast enhancement.

Discussion

Sellar xanthogranuloma (SXG) is a rare, benign, chronic inflammatory lesion of the sellar and suprasellar region, characterized histopathological by cholesterol clefts, foamy macrophages (xanthoma cells), multinucleated giant cells, hemosiderin deposits, fibrous tissue, and chronic inflammatory infiltrates, histopathological characteristic presented in all three cases reported. However, lacking the epithelial components usually present in craniopharyngiomas or Rathke's cleft cysts. Etiology involves a secondary inflammatory response, often from prior cystic lesions like Rathke's cysts, and less commonly from craniopharyngiomas, followed by events such as cyst rupture, leakage or hemorrhage, inciting granulomatous reaction and xanthomatous transformation.

Clinically, SXG presents with persistent headaches, visual disturbances from optic chiasm compression, and anterior/posterior pituitary dysfunction such as hypopituitarism or diabetes insipidus, matching the clinical and endocrinological presentation of our 3 cases. Imaging reveals a cystic or mixed cystic-solid sellar mass - cystic component hyperintense on the T1-weighted image (WI) and T2WI and a solid component hyperintense on T1WI and hypointense on T2WI, also presenting sometimes fluid-fluid levels.

Definitive diagnosis requires histopathology, distinguishing SXG from other sellar lesions by the absence of epithelial lining and others radiologic xanthogranulomatous features seen in all our three cases, such as chronic granulomatous inflammation, foreign-body giant cells, cholesterol clefts, necrosis and fibrosis. Surgical resection, typically via endonasal transsphenoidal approach, is the mainstay of treatment and generally curative, although pre-existing pituitary dysfunction often persists postoperatively. The rarity and phenotypic heterogeneity of SXG remain a clinical challenge.

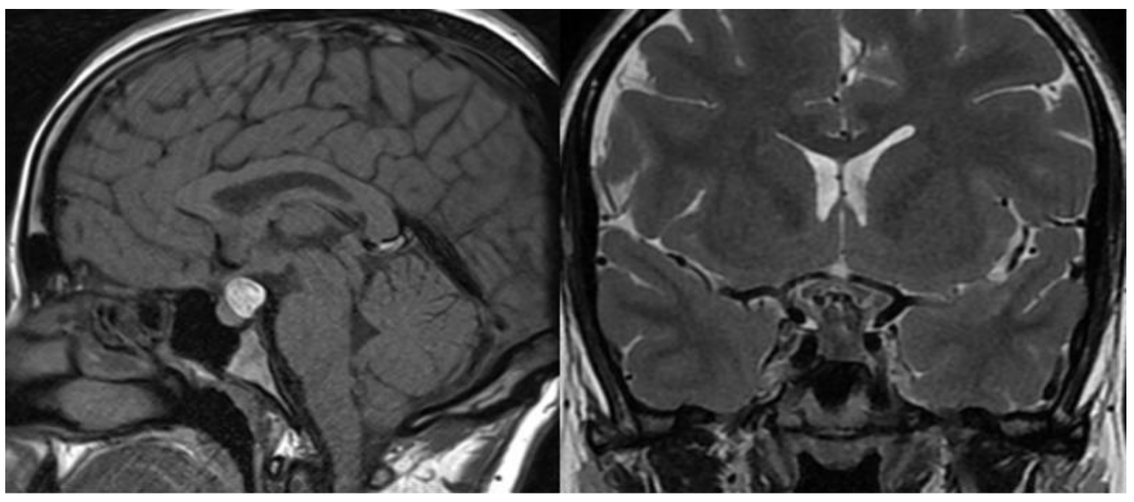

Figure 3. Third case - Preoperative brain MRI showing a lesion in the posterior sella region and suprasellar area, hyperintense on T1, without contrast enhancement, and areas of hypointensity and isointensity on T2.

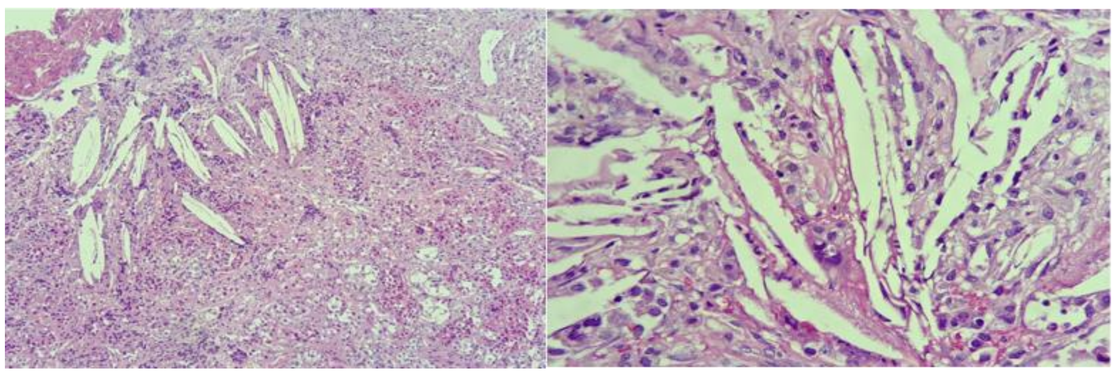

Figure 4. Histopathological analysis revealing cholesterol clefts (above, in the detail) and a xanthogranulomatous inflammatory process.

Conclusions

SXG remains a rare but important differential diagnosis for sellar masses, often mimicking other lesions on imaging. Definitive diagnosis relies on histopathological examination following surgical resection, which is typically curative. However, pre-existing pituitary dysfunction may persist, highlighting the need for careful pre- and postoperative endocrine management. Increased awareness of SXG's varied presentation is crucial for timely and accurate diagnosis.

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References