Xanthogranuloma of the Sellar Region

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23.1 Epidemiology and Clinical Presentation

- This lesion is also called a "cholesterol granuloma." (*See also* the next Chap. 24.)
- They have been reported to comprise a subset of lesions (up to 34 %) initially thought to be adamantinomatous craniopharyngiomas or Rathke's cleft cysts [1].
- They are likely to originate from Rathke's cleft cysts or pituitary adenomas with internal hemorrhage, inflammation, or both [2].
- They tend to occur in younger adult patients (mean, 27 years).
- In some patients, the lesion may be part of the Erdheim-Chester disease [3].
- They belong to a spectrum of disease that includes xanthogranulomas, xanthomatous hypophysitis, and xanthogranulomatous hypophysitis [4, 5]. (*See* "Inflammatory Hypophysitis" in Chap. 56.)
- Patients have a relatively high incidence of hypopituitarism and diabetes insipidus.

23.2 Imaging Features

- MRI shows a cystic sellar and suprasellar lesion with a thickened capsule [6].
- T2-weighted MRI shows a hypointense or mixed-intensity mass with an inhomogeneous contrast enhancement pattern [6, 7].
- Cholesterol clefts typically show high signal intensity on T1 images and low signal intensity on T2 images [2].
- Xanthogranulomas are smaller in diameter than craniopharyngiomas, and they tend to remain primarily intrasellar [1].
- Xanthogranulomas may extend to or invade the cavernous sinuses, an inflammatory process similar to the Tolosa-Hunt syndrome.

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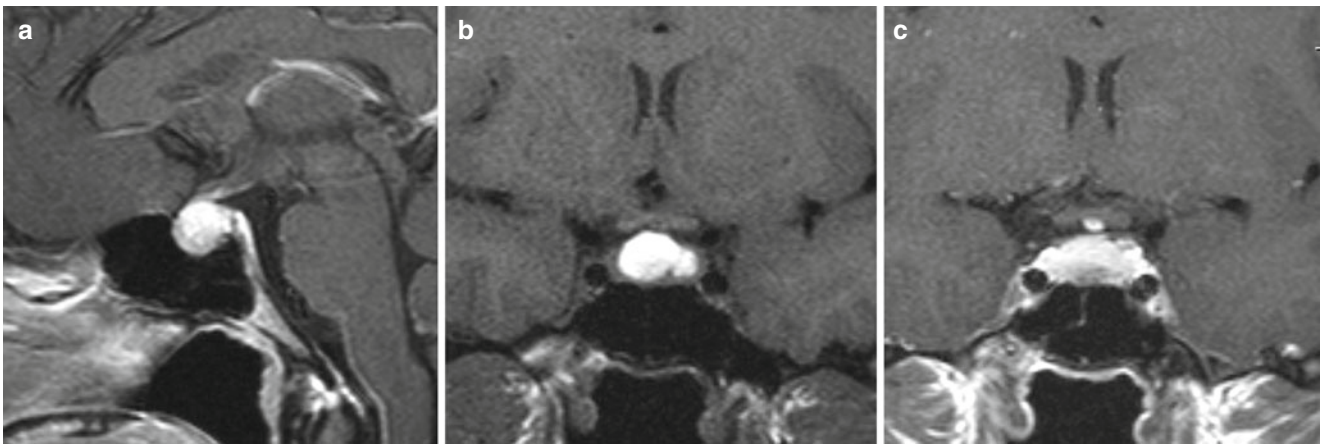

Fig. 23.1 (**a**) Sagittal T1-weighted post-gadolinium image. (**b**) Coronal T1-weighted pre-gadolinium image. (**c**) Coronal T1-weighted post-gadolinium image. There is a lobulated cystic lesion in the sella containing T1 hyperintense material. The pituitary stalk is displaced anteriorly

23.3 Histopathology

- Extensive inflammation with no readily identifiable epithelium is seen in the majority of cases [1, 8].
- Frequent histopathological findings include cholesterol clefts, macrophages, chronic inflammatory infiltrates, necrotic debris, foamy histiocytes, and hemosiderin deposits [4].
- Because these lesions may not have an epithelial lining, immunohistochemistry for cytokeratins is negative [8].
- Although they have been reported to comprise a distinct entity, it remains unknown whether they are derived from RCCs, craniopharyngiomas, and/or other lesions.
- Features of xanthogranuloma have been reported to be more consistent with RCCs than with craniopharyngiomas; they have demonstrated a high association with squamous metaplasia of these lesions [8], as seen in Fig. 22.22d.

23.4 Clinical Management

- Surgical management is the preferred primary treatment for symptomatic xanthogranulomas. Complete resection may be warranted if initial attempts at fenestration and drainage are unsuccessful.

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