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Xanthogranulomatous Hypophysitis: A rare but mistaken pituitary lesion

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Introduction

- Xanthogranulomatous hypophysitis (XGH) is a very rare form of pituitary hypophysitis that may present both clinically and radiologically as a neoplastic lesion or craniopharyngioma. [1,2]
- Our case series compares the paediatric and adult presentations of XGH and the differential diagnoses considered.

Case series

Patient 1:

- A 15 year old female presented with refractory headache, lethargy, short stature, delayed growth [weight [-3.36] SDS, height [-1.73] SDS, BMI 14kg/m^2], primary amenorrhoea and pubertal arrest over 18 months.
- Visual examination showed bitemporal quadrantanopia.

Patient 2:

- A 21 year old female presented with lethargy, frontal headaches and secondary amenorrhoea, three years after delivery.
- Postpartum she had initial galactorrhoea and irregular periods which stopped after a year.
- Visual examination was normal.

Patient 3:

- A 64 year old female presented with multiple syncopal attacks over 7 years.
- A MRI scan performed for suspected vertebrobasilar insufficiency revealed a pituitary mass.
- There was a previous history of hypothyroidism for 20 years and hysterectomy at 39 years of age for irregular periods.
- Eye assessment revealed left temporal quadrantanopia.

Management

- Endocrine investigations suggested panhypopituitarism in all three patients and thus they were commenced on treatment. Patient 2 had high prolactin levels requiring Cabergoline.
- Pituitary MRI revealed a suprasellar mass compressing the optic chiasm suggestive of craniopharyngioma or rathke's cleft cyst in patient 1; non-functioning pituitary macroadenoma in patients 2&3. [Fig 1&2]
- MRI appearance was of mixed signal intensities on T1-and T2-weighted sequences.
- All 3 patients underwent an endoscopic trans-sphenoidal surgery.
- Histology revealed areas with cholesterol cleft formation associated with multinucleate giant cells and numerous macrophages.[Fig 3]

Figure 1: Patient 2's pre: Coronal (a) and sagittal (b) T1 contrast enhanced MRI images demonstrating the pituitary lesion (white arrow) with suprasellar extension and compression of the optic chiasm.

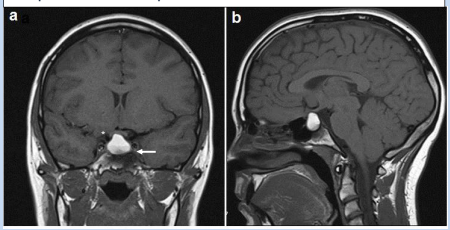


Figure 2: Patient 2's MRI scan of pituitary post-surgery

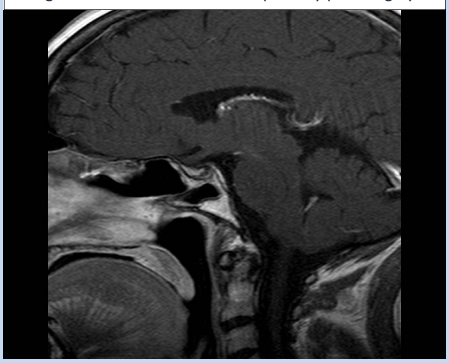
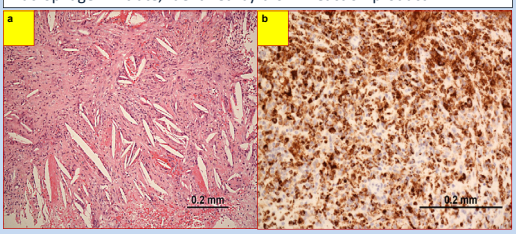


Figure 3. Patient 2: Histological analysis of pituitary lesion. (a) Area of abundant cholesterol cleft formation, haematoxylin and eosin. (b) Immunohistochemistry to CD68 demonstrating a heavy macrophage infiltrate, identified by brown reaction product.



Conclusions

- XGH in children primarily presents with growth arrest and pubertal arrest in adolescents. In adults the presentation may vary.
- XGH being a treatable condition and might also be suggesting other areas of xanthogranulamotus change must be considered as part of the differential diagnosis when a pituitary mass is demonstrating mixed signal intensities.

Reference

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2. Yokoyama S, Sano T, Tajitsu K, Kusumoto K. Xanthogranulomatous hypophysitis mimicking a pituitary neoplasm. Endocr Pathol. 2004 15:351–357.