

# Xanthomatous hypophysitis : a rare case in a paediatric patient

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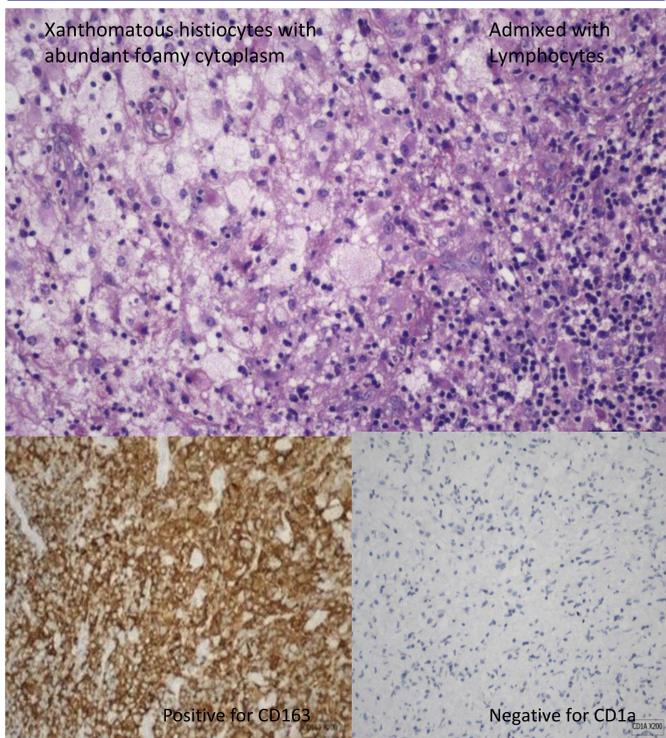
## BACKGROUND

Hypophysitis is an inflammatory condition of the pituitary that can mimic a neoplastic lesion<sup>1-5</sup> It is rare with an annual incidence estimated at 1 in 7-9 million<sup>1-2</sup> Treatment options are surgery, glucocorticoid, radiotherapy & immunotherapy<sup>1-5</sup>

Histopathology subtypes include lymphocytic, granulomatous, xanthomatous, plasmacytic or a mixed picture<sup>1-5</sup>. Among these, XH is the rarest and mostly reported in adult female patients.<sup>1-2</sup>

The aetiology of XH is unknown. Theories include an initial precipitating event (infectious, autoimmune or localised endothelial dysfunction) that leads to extravasation of macrophages into inflamed interstitial tissue. Macrophages subsequently begin to phagocytose injured cell membranes containing phospholipids and cholesterol and become lipid-laden xanthoma cells.<sup>2</sup>

Unlike lymphocytic hypophysitis, XH is rarely reported to be associated with other autoimmune disease and response to glucocorticoid is unclear.<sup>2-5</sup>



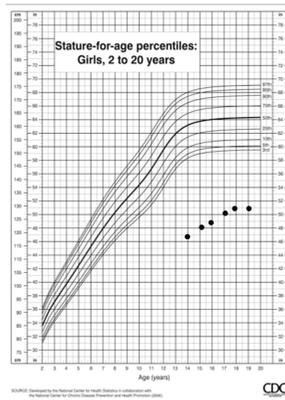
HPE: Fibrovascular tissue densely infiltrated by xanthomatous and histiocytic cells admixed with lymphocytes, plasma cells, eosinophils and neutrophils. No Langerhans cells, epithelioid granuloma or malignant cells seen. No infective organisms (fungal bodies, acid fast bacilli) identified on special stains.

### Immunohistochemistry staining:

Histiocytes are immunoreactive to CD163 and CD 68. Negative for PLAP, CD 117 (markers for germinoma) Negative for S100 and CD1a (markers for Langerhans cell histiocytosis)

## CASE REPORT

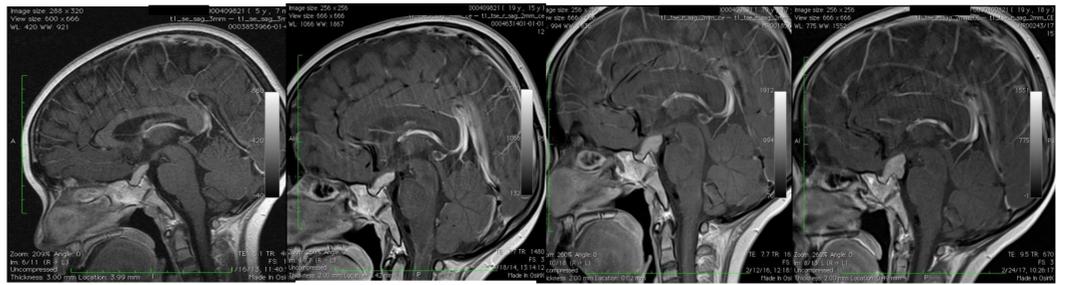
21 year-old woman, referred at the age of 14 years for delayed puberty and stunted growth for 2 years.



Height 120cm (>2 SD below her mid-parental height), weight 22kg. Final height achieved 130cm, current weight 30kg. Neurology, visual fields and other systems normal. Bone age 11 years at chronological age 14 years

Multiple pituitary deficiencies on presentation : TSH, growth hormone, gonadotrophins. Development of diabetes insipidus five years later . Started on thyroxine, pubertal induction and oral desmopressin. Growth hormone was not started. Adrenal function – continued surveillance remained satisfactory (similar with other cases of XH whereby adrenal deficiency is less common). Secondary workout (autoimmune, infectious, neoplasm including germinoma) was negative.

MRI 2013, 2014, 2016, 2017

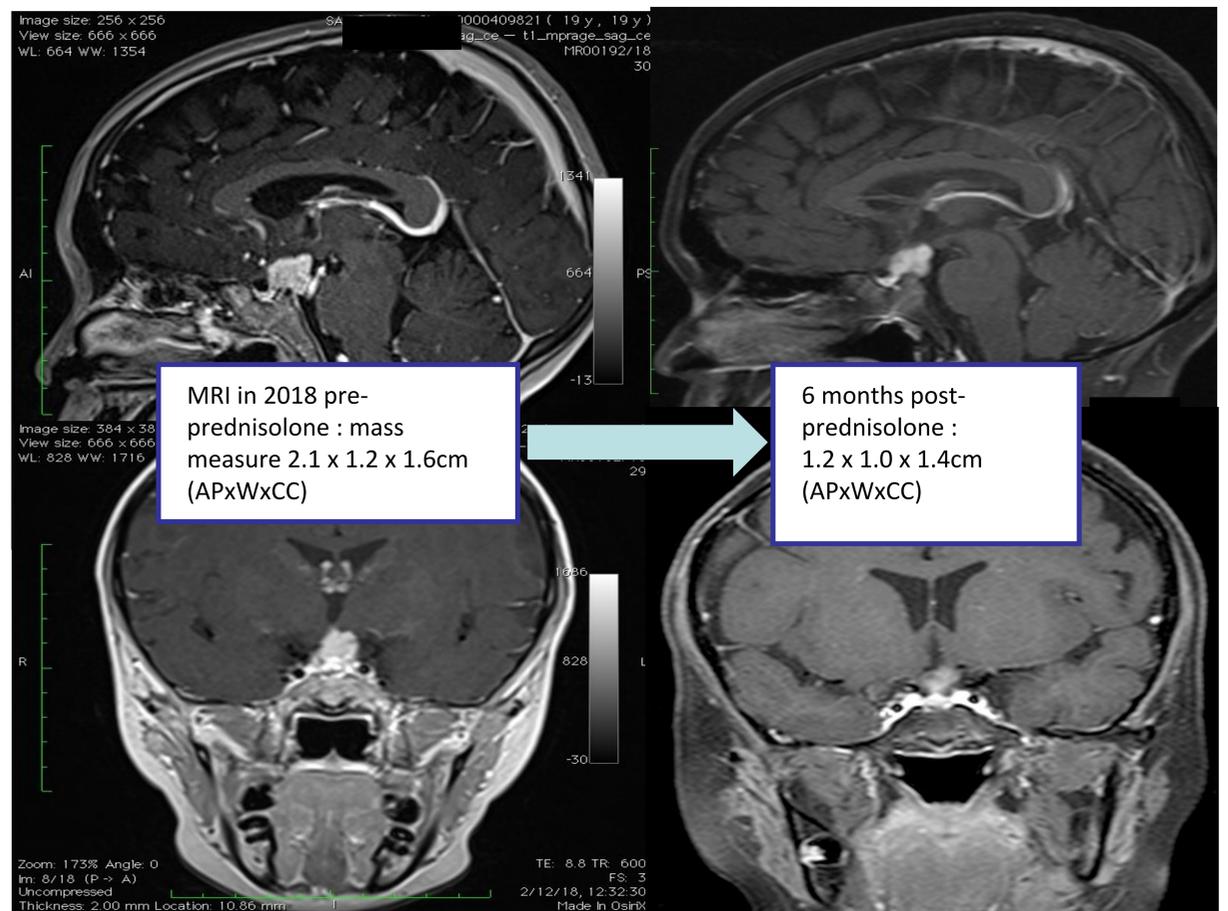


MRI in January 2013 : Homogenous lobulated mass 0.6x1.3x1.7cm (APxWxCC) arising from the pituitary stalk with extension to the sellar, suprasellar regions abutting the optic chiasm.

MRI 2014-2015 no change in the mass size

MRI 2016 increment in size measuring 1.8 x 1.4 x 1cm (APxWxCC) → underwent a partial resection and biopsy in 2016. Histopathology of the biopsy was suggestive of XH.

Post-resection MRI showed a continued increment of mass size. After a multidisciplinary discussion, oral prednisolone 30mg (1mg/kg) once daily was administered for 2 months and tapered off weekly. Repeated MRI six months after completion of prednisolone showed a smaller mass.



## CONCLUSIONS

We report a girl with XH post-surgical resection with a residual progressive lesion that showed a response to glucocorticoid with no acute side effects.

While glucocorticoid could be a treatment option for selected cases of XH, there is insufficient evidence to recommend its routine use as well as the best regimen and timing.

In addition, understanding of the pathogenesis, progression and prognosis of XH remains limited.

## References

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