

Severe Juvenile Hypertrophy of the Breast with Hypercalcaemia; Mastectomy v's Reduction Surgery

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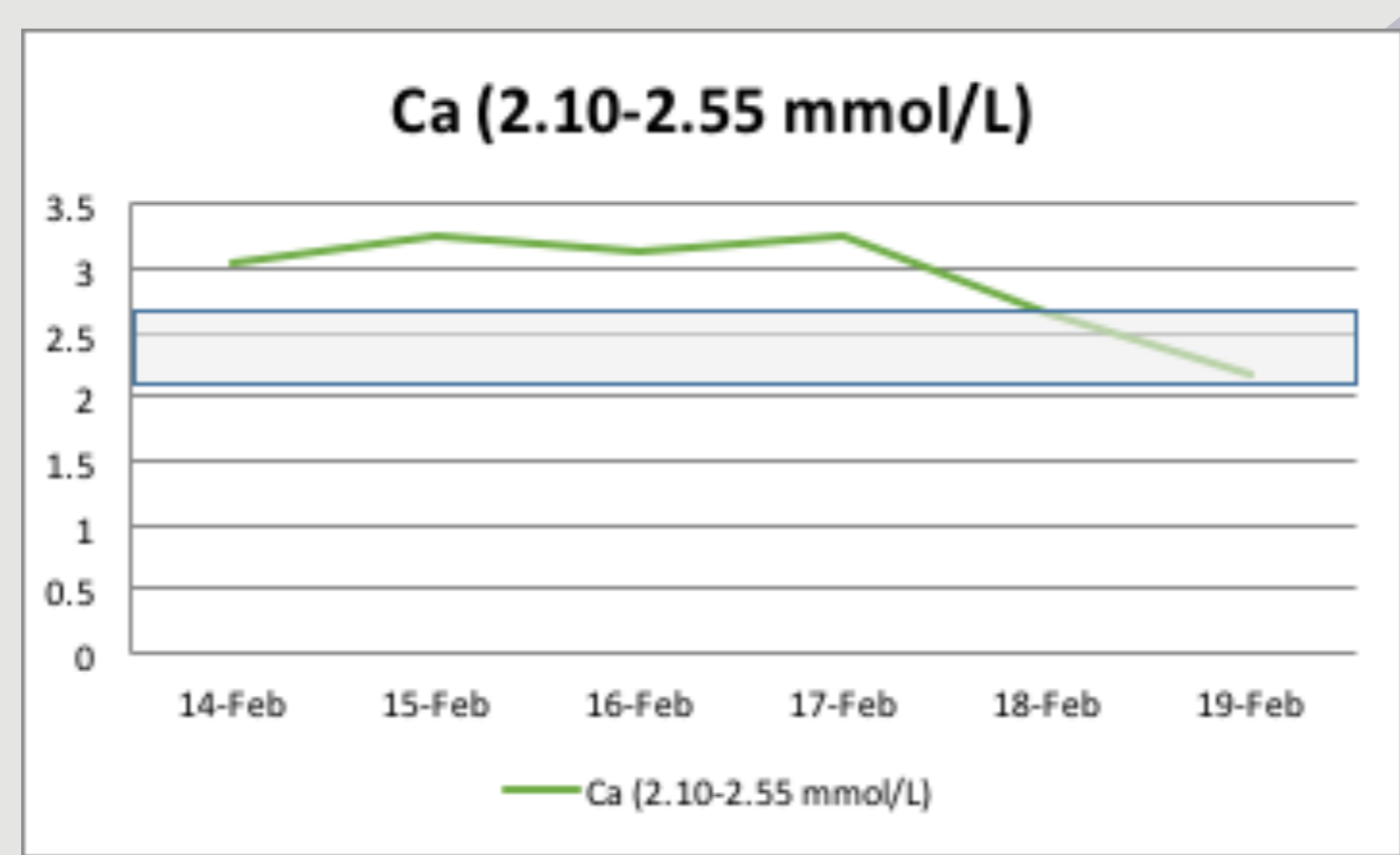
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Mastectomy v's Reduction Surgery

- Total bilateral mastectomy as a surgical intervention has been reported as the preferred course of action for severe juvenile hypertrophy. The reported rate of recurrence following reduction surgery is significant.^{1,2}
- In considering the ethical dilemma in this case the patient's severe autism and learning difficulties were considered, as was the likelihood of breastfeeding in the future.
- Both parents were adamant that no breast tissue be left behind for fear of recurrence.

Hypercalcaemia

- The hypercalcaemia resolved following a period of hyperhydration (graph 1). Possible causes of hypercalcaemia included fat tissue necrosis in view of tissue breakdown, or Parathyroid Hormone related Peptide (PTHrP).
- PTHrP was 2.0 pmol/L (< 1.8). Hypercalcaemia with raised PTHrP has been reported before in relation to breast hypertrophy.³ As well as in relation to humoral hypercalcaemia of cancer.⁴
- Parathyroid hormone was suppressed at < 1.2 pmol/L, ruling out hyperparathyroidism.



Graph 1: Resolution of Hypercalcaemia

Test	Result	Interpretation
FSH	0.6 IU/L	Pubertal
LH	3.8 IU/L	Pubertal
Prolactin	385 mIU/L	Normal
Breast USS	Hyperstimulated breast tissue	No abscess or malignant features
MRI Pituitary	Normal study	Not contributory
MRI Pelvis	Normal study	Not contributory
IGF-1	13.1 nmol/L	Normal
IGFBP3	1.5 mg/L	Normal

Table 1: Relevant Investigations

Case Presentation

A 10 yr old female with autism and learning difficulties presented with rapidly progressive asymmetrical breast development during accelerated cadence through puberty.

She exhibited increasing behavioural difficulties and anxiety thought to be associated with discomfort and significant hypercalcaemia.

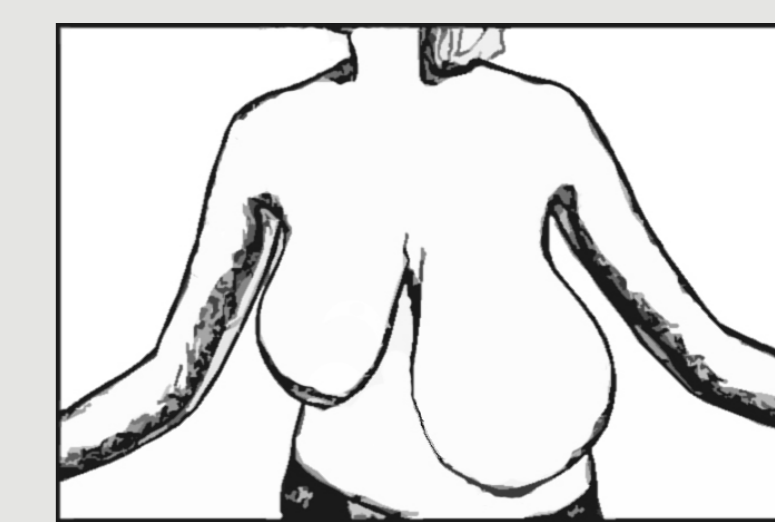
There was a strong family history of breast cancer.

April - Sept. 2015

Rapid cadence through puberty with progressive asymmetrical breast development.

Dec. 2015

Referred by GP, increasing breast size, discomfort, disturbed sleep and worsening behaviour.



Jan. 2016

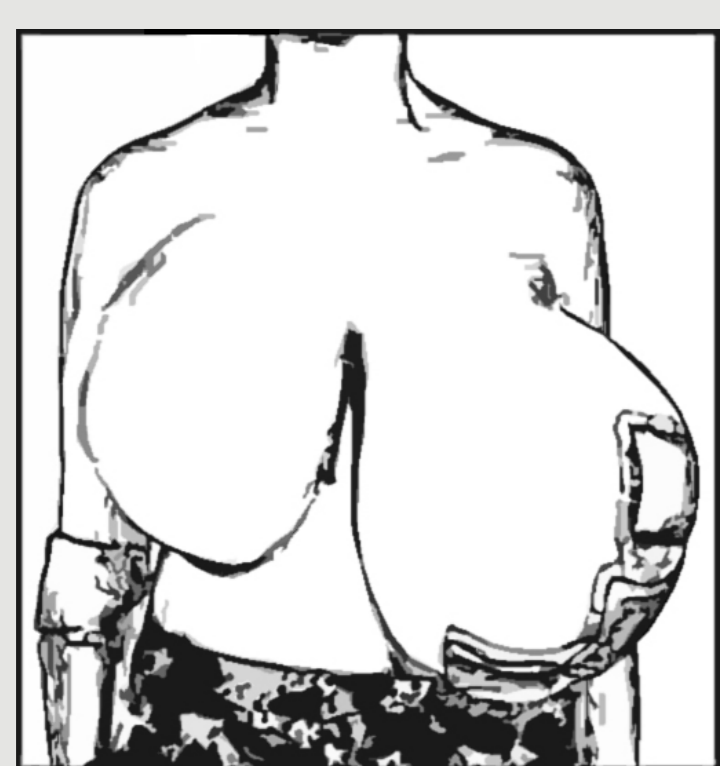
Local hospital assessment with initial investigations (table 1). Review with visiting Endocrinologist; commenced Gonapeptyl injections.

4th March 2016

She had an uncomplicated recovery and was discharged home with simple analgesia. Gonapeptyl and Tamoxifen were stopped.

1st March 2016

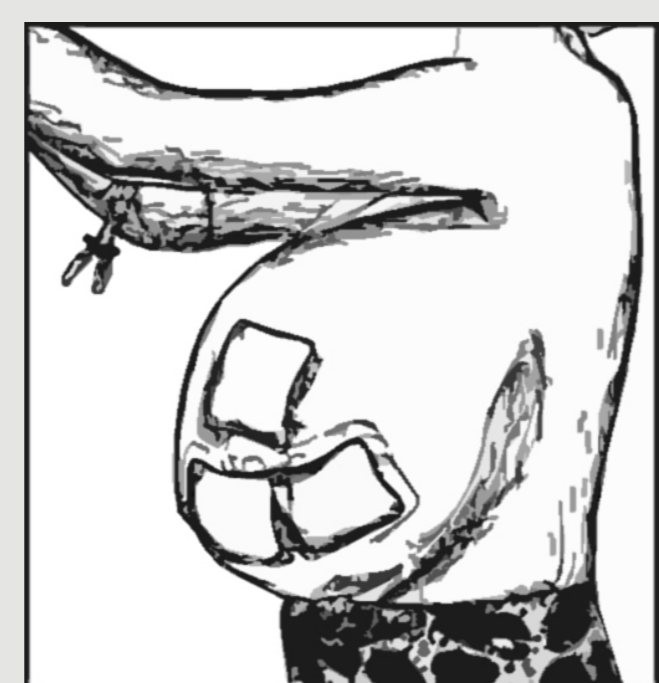
A serum sample for PTHrP was taken to investigate the aetiology of hypercalcaemia. Bilateral mastectomy was performed, the histology was consistent with massive juvenile hypertrophy with some areas of necrosis. The breast tissue also exhibited high levels of expression of oestrogen and progesterone receptors.



Right breast : 1392g
Left breast : 3025g

19th Feb.

Transfer to Regional Children's Hospital for endocrine and surgical assessment. Hypercalcaemia resolved with hyperhydration.



She developed significant hypercalcaemia which was managed with hyperhydration. Ongoing concern regarding skin breakdown and tissue necrosis. Commenced Tamoxifen to block oestrogen receptors.

12th Feb 2016

Admitted to local hospital with increasing breast enlargement. Skin changes with tissue breakdown and extreme discomfort requiring intravenous antibiotics and opiate analgesia.

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Disclosure statement: there are no conflicts of interest.

Conclusions

- Juvenile breast hypertrophy can be massively debilitating and disturbing for young people and their families.²
- Surgical intervention is indicated for juvenile breast hypertrophy to the extent represented in this case. Total mastectomy was the preferred option with an excellent outcome.
- PTHrP has become a useful diagnostic tool in the differential diagnosis of hypercalcaemia.⁴

