

"A Gut Feeling"

Identifying the cause after an AA Amyloidosis diagnosis

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AA Amyloidosis is caused by states of chronic inflammation. These can be chronic inflammatory conditions, infections or malignancy. Rarely Unicentric Castleman disease can be a cause of AA Amyloidosis.

Case Report

Presentation

A 46-year-old female was referred to renal clinic with hypoalbuminaemia (24g/L), proteinuria (3.1g/24hrs) and preserved kidney function (Creatinine 70umol/L, eGFR >90ml/min/1.73m²). Her background only included obesity.

Investigations

- Ultrasound and serological testing were unremarkable, including serum protein electrophoresis and free light chains.
- The kidney biopsy demonstrated segmental expansion of glomeruli with homogenous eosinophil acellular material, positive for congo red stain and green-yellow birefringence under polarised light. Immunohistochemistry was positive for amyloid A protein.
- A bone marrow aspirate and trephine demonstrated amyloid deposits in the vessels outside of the marrow, with clonal population. Her serum amyloid A level was 39mg/L (Figure 1A).

Progress

- Extensive screening for a secondary cause for AA amyloid was negative, and it was thought to be secondary to obesity. She was trialled on oral colchicine with no effect.
- Two months later, the patient noted a palpable lump in her upper abdomen. A PET scan was performed due to concerns for a malignant process, and this demonstrated an intensely FDG avid abdominal mesenteric lymph node (Figure 2A-B). Excisional biopsy demonstrated histology consistent with Unicentric Castleman disease, plasma cell variant.
- She underwent a complete resection of the lymph node later that year.
- Her CRP, proteinuria and Serum Amyloid A levels have improved since surgical resection (Figure 1A-C).

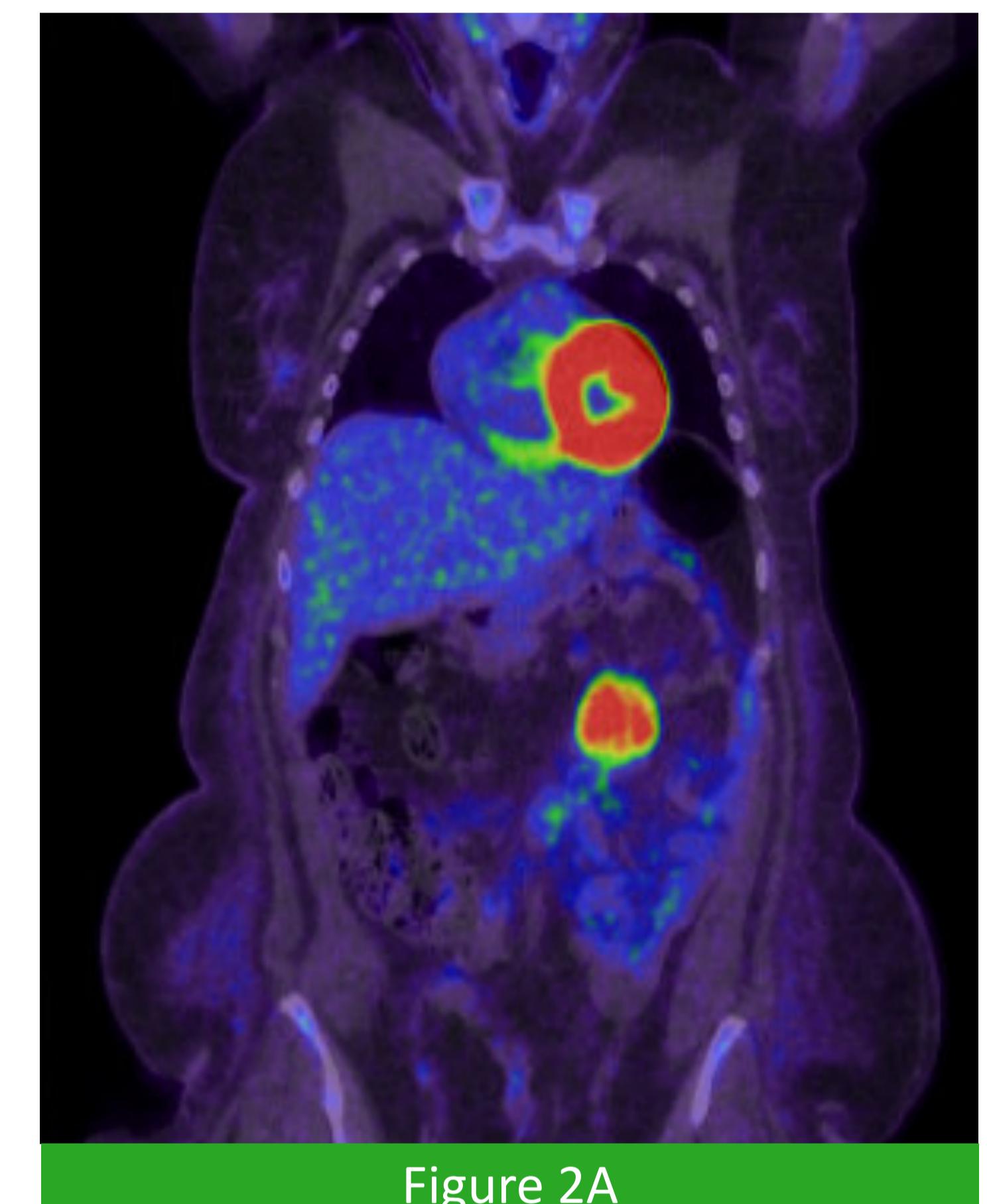


Figure 2A

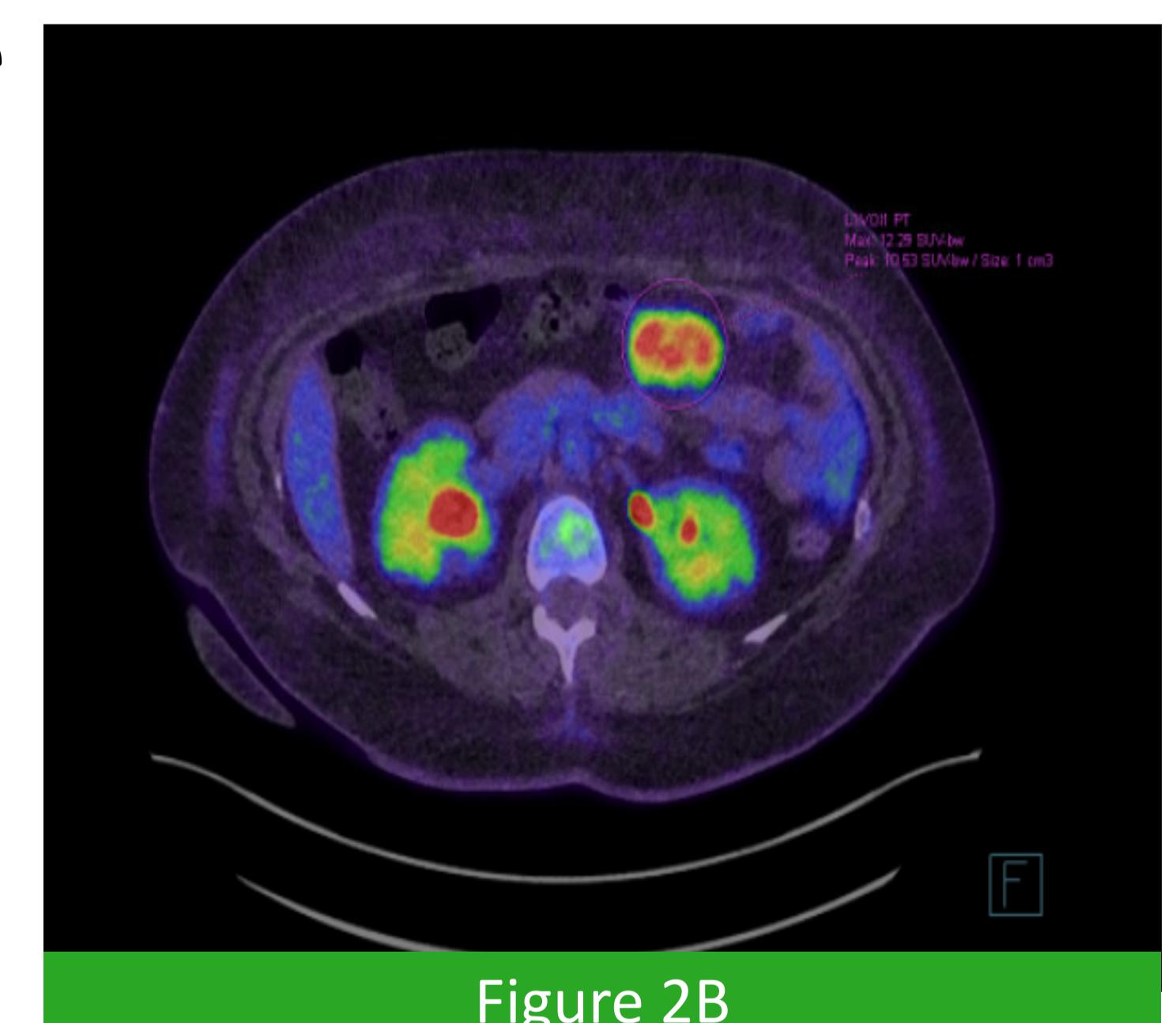
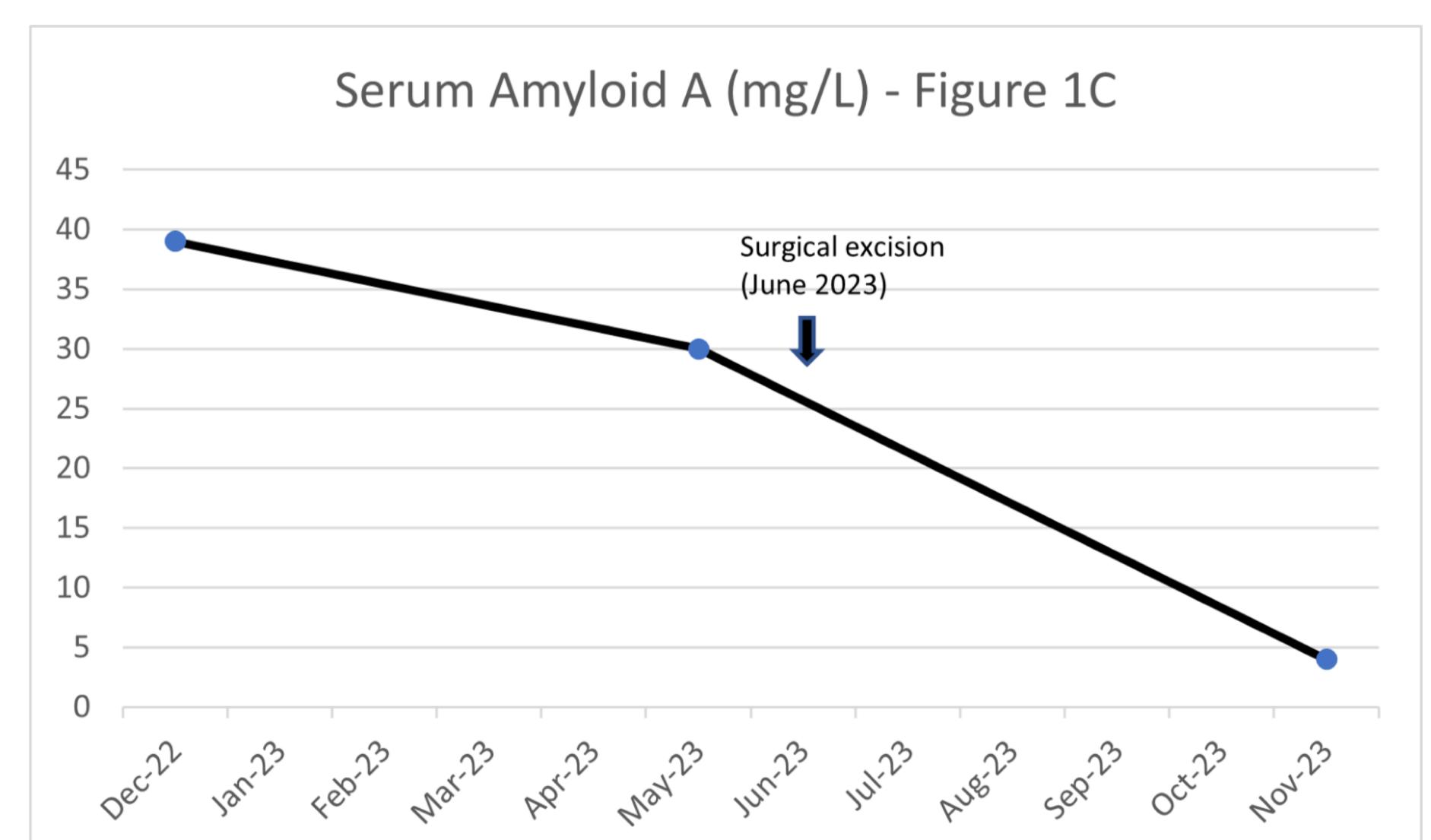
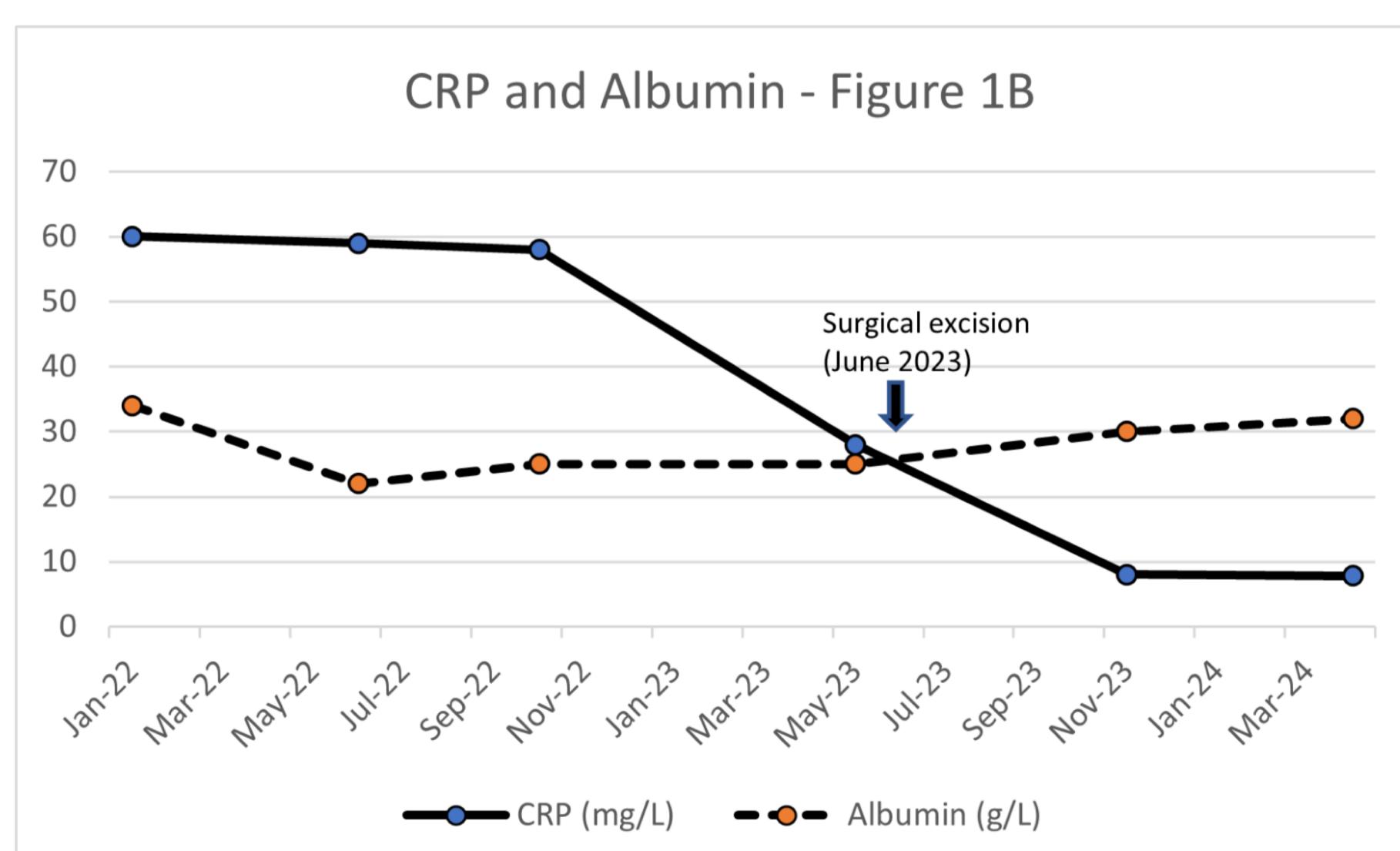
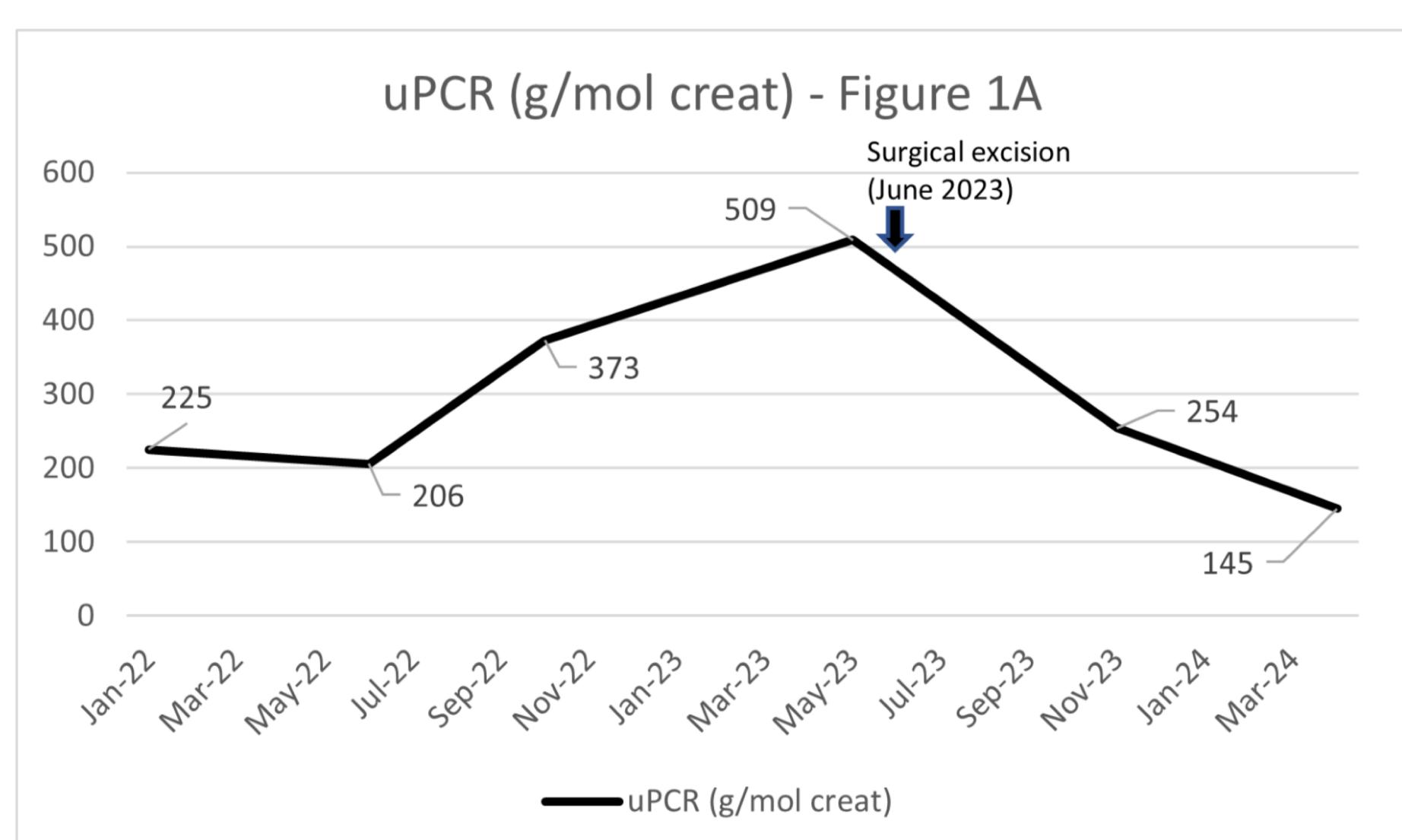


Figure 2B



Conclusion

In patients with AA Amyloidosis due to Unicentric Castleman disease, systemic symptoms may resolve with resection of the lymph node. This case highlights the importance of identifying the cause of AA Amyloidosis to direct management.