

## Liposarcoma masquerading as IgG4-related disease

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### INTRODUCTION:

- IgG4-related disease (IGRD) is characterized by lymphoplasmacytic tissue infiltration by IgG4-positive plasma cells and small lymphocytes and is known to involve multiple organs, sometimes simultaneously.<sup>1</sup>
- Retroperitoneal involvement is common with IGRD, often presenting as mass-like lesions and fibrosing disease such as retroperitoneal fibrosis<sup>2</sup> and sclerosing mesenteritis<sup>3</sup>.
- While there is no established association, very few cases have been reported in literature about strong IgG4 positivity and sarcomas.<sup>3,4</sup> Investigation usually warrants biopsy, as imaging studies and serologies cannot differentiate between IGRDs and sarcomas. We present a case of an aggressive liposarcoma mimicking IGRD.

### CASE DESCRIPTION:

- A 40-year-old female presented with right upper quadrant (RUQ) pain for 3 months. Patient was hemodynamically stable with physical exam demonstrating fullness and tenderness in RUQ.
- CT abdomen revealed a 12.9x11.8x8.7cm heterogenous macro-lobulated mass located below the inferior hepatic border in the retroperitoneal space and hepatorenal recess (Image A). CT-guided biopsy of the mass revealed increased IgG4-positive cells identified by immunohistochemical studies, however, FISH MDM2 amplification studies were inconclusive. Patient's serum IgG4 levels were elevated, and she was started on prednisone therapy.
- At 2-month follow-up, the patient's serum IgG4 levels had decreased but upon physical examination, the mass had increased in size and was firmer. Decision was made to repeat CT-guided biopsy, wherein MDM2 amplification studies revealed that the mass was consistent with well-differentiated liposarcoma (Image B).
- She underwent liposarcoma resection, lymph node dissection, and right nephrectomy (adherent to the mass). Histopathologic analysis revealed a final diagnosis of stage I well-differentiated liposarcoma. Patient now follows for surveillance of her liposarcoma.

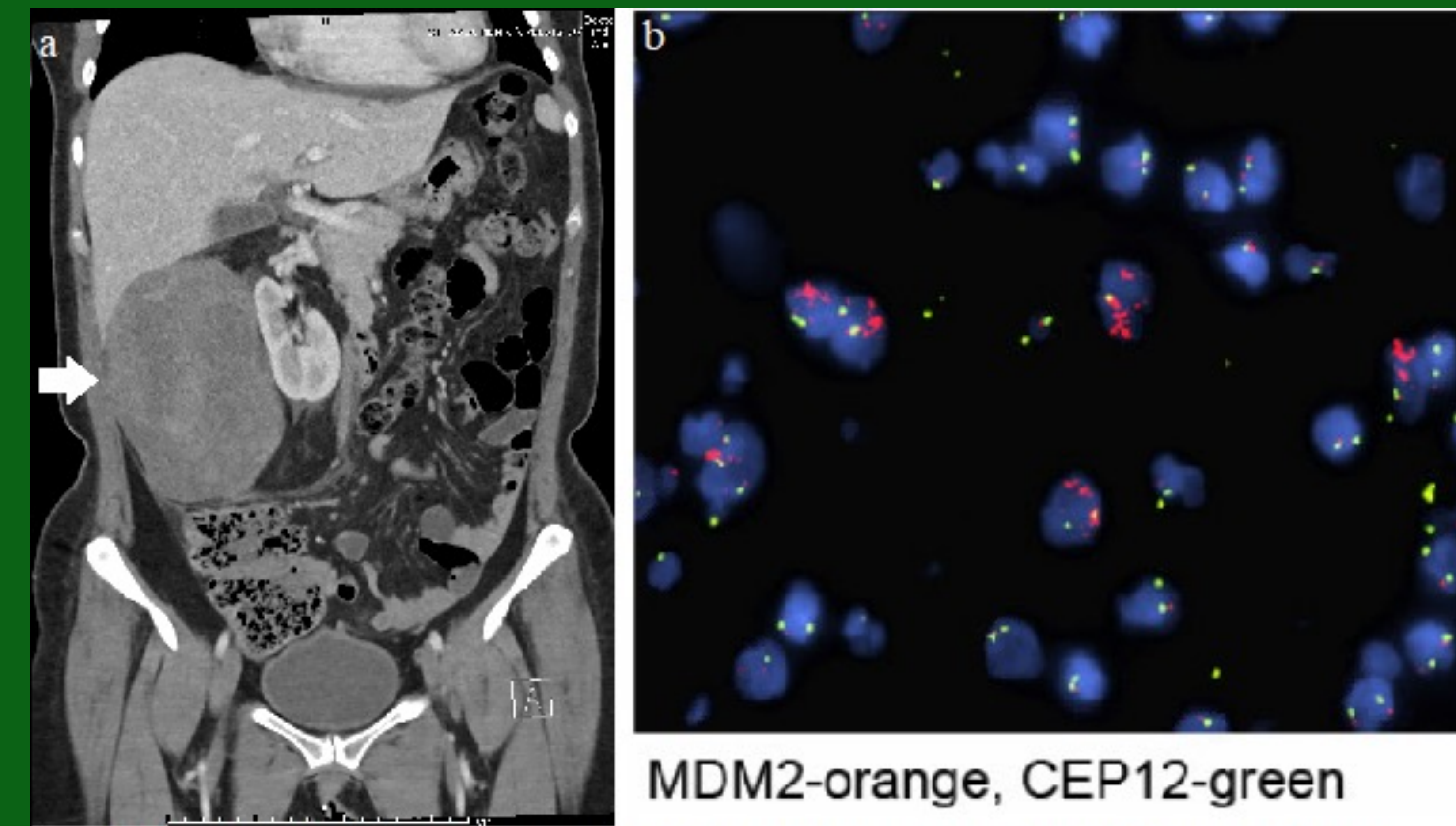
### DISCUSSION:

- IGRD and liposarcoma can have common initial presentations and at times overlapping serological markers such as IgG4 levels, which necessitates histopathologic analysis for confirmation of the disease.
- In addition, raised IgG4 levels are not specific and can be elevated in infections and autoimmune conditions<sup>5</sup>.
- Our case emphasizes the importance of considering liposarcomas in retroperitoneal masses with elevated IgG4 levels, as they can mimic IGRD. A repeat biopsy should be considered if the initial diagnosis is doubtful, or patient responds poorly to corticosteroids.

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IgG4-related disease and Liposarcoma can have similar presentation and overlapping serological markers. **Biopsy can differentiate.**



A) CT scan showing a large macro-lobulated heterogenous mass  
B) MDM2 staining of the mass consistent with liposarcoma