



# FROM IMMUNOPROLIFERATIVE SMALL INTESTINAL DISEASE TO LYMPHOMA: A RARE PEDIATRIC CASE OF DUODENOJEJUNAL MALT LYMPHOMA TRANSFORMATION



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## CASE SUMMARY

A 12-year-old girl presented with chronic gastrointestinal symptoms and was diagnosed with IPSID. She received prolonged treatment with metronidazole and amoxicillin-clavulanate; however, symptoms persisted. Repeat endoscopy and biopsy after 5 months demonstrated duodenal–jejunal MALT lymphoma. She subsequently received six cycles of rituximab-based R-CHOP chemotherapy, with complete radiological resolution of lesions on post-treatment PET-CT. She remains in remission six months after completion of therapy.

## BACKGROUND

- MALT lymphoma accounts for **5–8% of adult NHL**, but is exceptionally rare in children, with only a handful of pediatric cases reported. Gastrointestinal MALT lymphomas remain uncommon.
- Most MALTomas occur in the stomach, with **less than 30% occurring in the small intestine**, even in adults.
- IPSID to synchronous duodenal and jejunal MALT lymphoma progression is even rarer**, making this case notable for its unusual site, age, and possible infectious etiology.
- Persistent symptoms in IPSID **warrant repeat endoscopy and biopsy** to detect possible progression to intestinal lymphoma.

## IPSID & SERUM IgA

**IPSID** is characterized by small-intestinal involvement with **abnormal IgA/ $\alpha$ -heavy-chain production** and characteristic histopathological changes.

Serum IgA at presentation

**12.37 g/L**



Normal: under 2.82 g/L

## DIAGNOSIS & TREATMENT

IPSID MALT lymphoma Remission

### Presentation and *H. pylori* eradication

Pain, dyspepsia and fever for six months. Two weeks of PPI triple therapy gave no relief.

### First endoscopy: **IPSID, Galian stage A**

Nodular duodenum; plasma-cell-rich lamina propria infiltrate, CD138, MUM1, CD79a positive.

### Five months of antibiotics: **no response**

CT: duodenal and jejunal wall thickening with enlarged abdominal lymph nodes.

### Repeat endoscopy: **MALT lymphoma**

Nodular lesions D1 to D4. CD79a+, CD20 focal+, Bcl-2 weak+, CD10 and TdT negative, Ki-67 low.

### PET-CT: stage IE based on NCCN guidelines

FDG-avid bowel wall (SUVmax 5.0), no nodal disease.

### Six cycles of R-CHOP with IT methotrexate

Chosen over surgery and radiotherapy; only mild nausea and fatigue. **Deauville 4** after cycle 3, **Deauville 1** at end of treatment.

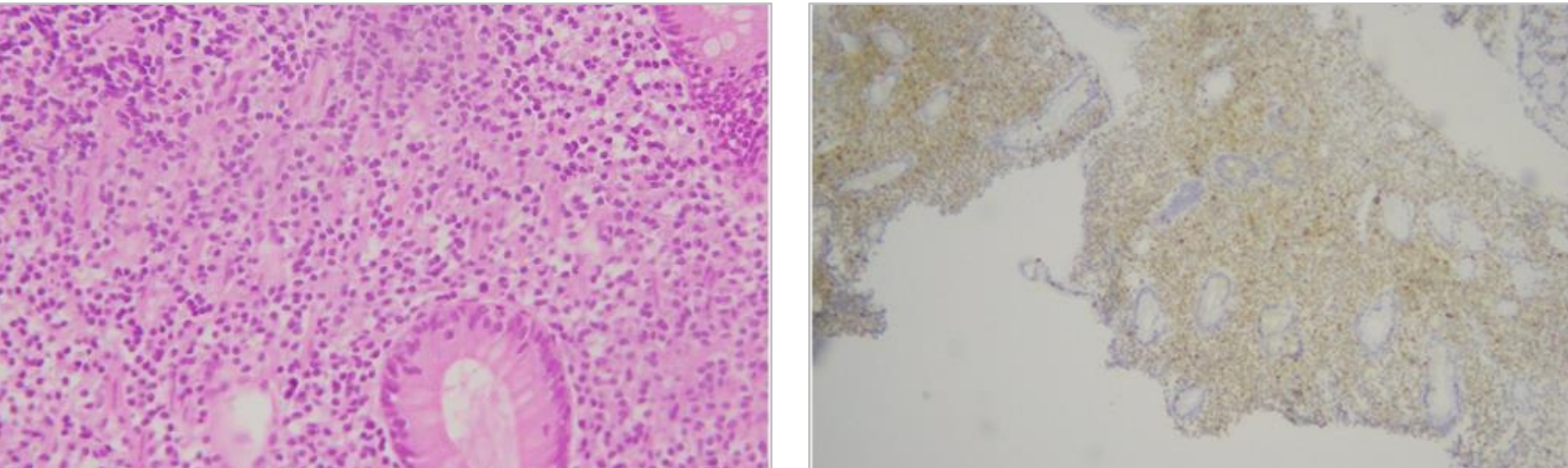
### In remission 14 months from diagnosis

Symptom-free with normal growth, six months off therapy.

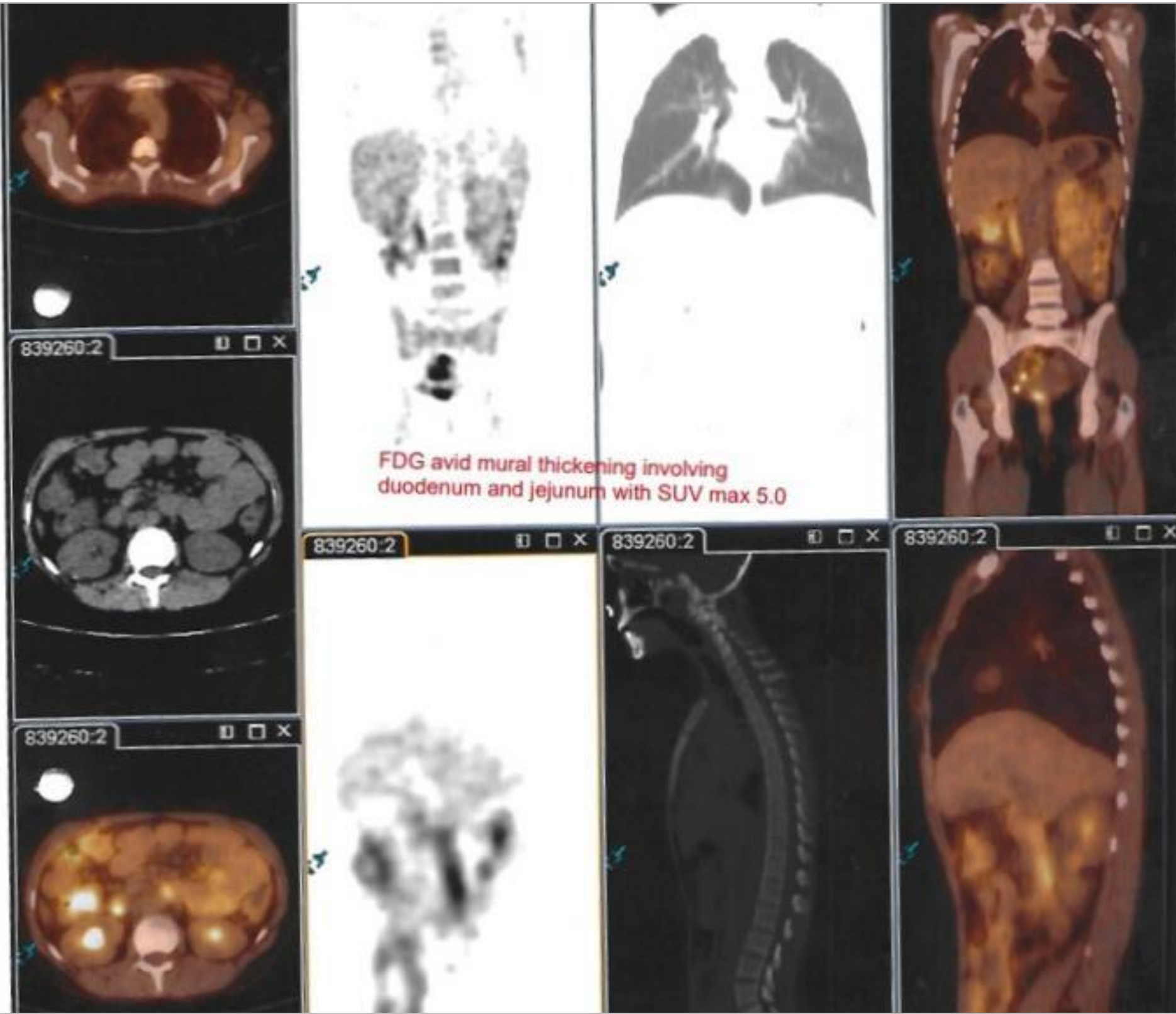
## CLINICAL IMAGES



**Repeat endoscopy:** nodular duodenum



**Duodenal biopsy:** H&E and Bcl-2 weak focal positive



**Staging PET-CT:** bowel wall uptake

## CONCLUSION

- Persistent symptoms despite *H. pylori* eradication should prompt **close surveillance and repeat endoscopy with biopsy** in pediatric IPSID to assess for possible evolution to MALT lymphoma.
- In high infectious-burden settings, **IPSID and MALT lymphoma should remain in the differential diagnosis** of persistent gastrointestinal symptoms in children.

## REFERENCE

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Molecular studies unavailable.



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