

Small Intestinal Xanthomatosis Mimicking Neuroendocrine Tumor

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CASE REPORT

A 73-year-old woman with a history of CREST syndrome, gastroesophageal reflux disease, and renal stones presented to the emergency department with left-sided back pain for 2 weeks. Physical examination revealed muscle spasm in the back. She takes omeprazole as needed for gastroesophageal reflux disease. Laboratory tests, including complete blood count, complete metabolic panel, and lipid panel, were normal. Computed tomography of the abdomen and pelvis with contrast showed a 3.3-cm calcified mass in the mesentery suspicious for neuroendocrine tumor (Figure 1); her serum chromogranin A level was 701 ng/mL (reference <311 ng/mL), supporting the diagnosis of neuroendocrine tumor. She underwent exploratory laparotomy with resection of 50 cm of small bowel. The pathology showed diffuse infiltration of foamy lipid-laden histiocytic cells in all layers of the small intestine consistent with diffuse small intestinal xanthomatosis (Figures 2 and 3). The mesenteric mass contained xanthoma cells with nodular sclerotic changes and calcifications. She recovered without incident.

Xanthomatosis is a benign disease defined as the local accumulation of cholesterol-rich macrophages, usually seen in the skin.¹ Xanthomatosis in other sites may not be related to hyperlipidemia.² Gastrointestinal xanthoma commonly involves the stomach. Intestinal xanthomatosis is rare but can cause intestinal obstruction. Chromogranin A is a neurosecretory protein in endocrine and neuroendocrine cells. It is a nonspecific marker and is usually increased in neuroendocrine tumors: A recent case report of an ectopic pancreatic mass also had an increase in serum chromogranin A, suggesting other possible origins.^{3,4} Other possible causes include some medications, such as proton pump inhibitors, which might be a consideration in this case.⁵ This case demonstrates that small intestinal xanthomatosis should be considered in patients with elevated chromogranin A.

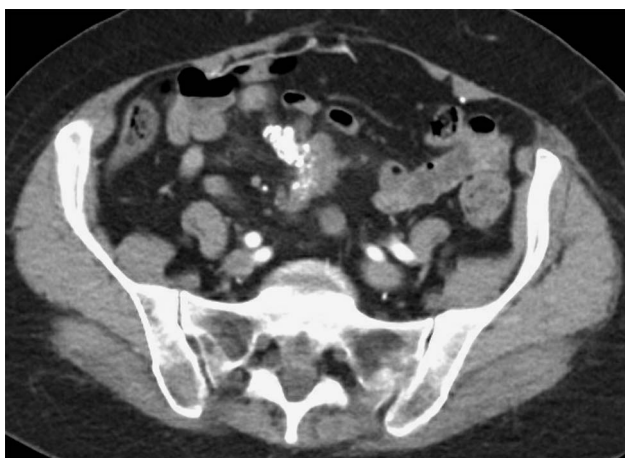


Figure 1. Computed tomography of the abdomen and pelvis with contrast showed a 3.3-cm calcified mass in the mesentery suspicious for neuroendocrine tumor.

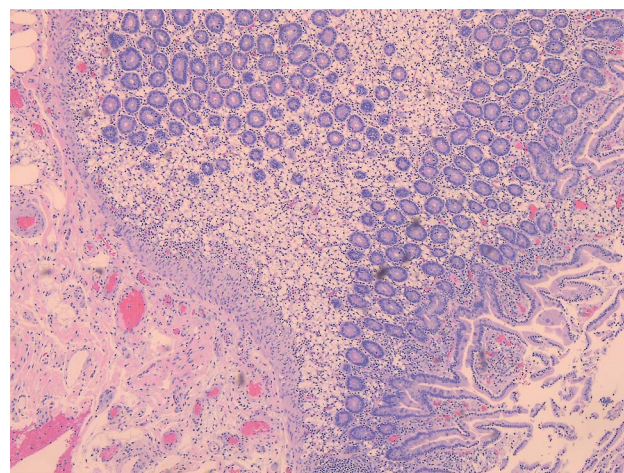


Figure 2. Hematoxylin and eosin stain shows small bowel mucosa and submucosa diffusely infiltrated by foamy lipid-laden histiocytic cells.

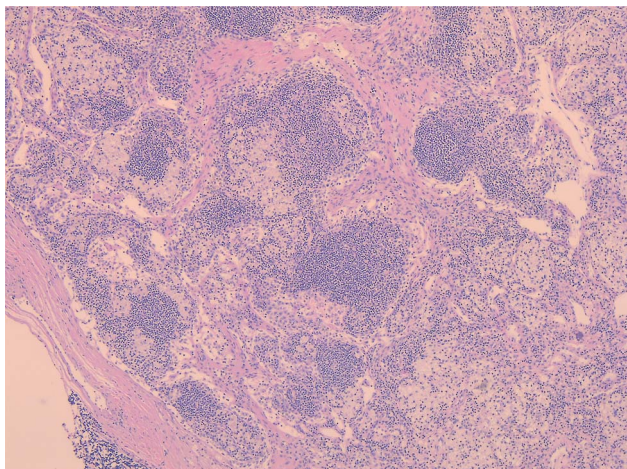


Figure 3. Hematoxylin and eosin stain shows infiltration of lipid-laden histiocytic cells in the lymph node of the mesentery.

DISCLOSURES

Author contributions: B. Songtanin wrote the manuscript and is the article guarantor. D. Arif provided pathology images. T. Mouw performed the surgery.

Financial disclosure: None to report.

Informed consent was obtained for this case report.

Received October 22, 2023; Accepted January 5, 2024

REFERENCES

1. Barrera-Herrera LE, Arias F, Rodríguez-Urrego PA, Palau-Lázaro MA. Small bowel obstruction due to intestinal xanthomatosis. *Case Rep Pathol.* 2015;2015:231830.
2. Nielsen SL, Ingeholm P, Holck S, Talbot I. Xanthomatosis of the gastrointestinal tract with focus on small bowel involvement. *J Clin Pathol.* 2007; 60(10):1164–6. (In English).
3. Dhruv S, Polavarapu A, Asuzu I, Andrawes S, Mukherjee I. Jejunal ectopic pancreas: A rare cause of small intestinal mass. *Cureus.* 2021;13(6):e15409. (In English).
4. Gut P, Czarnywojtek A, Fischbach J, et al. Chromogranin A: Unspecific neuroendocrine marker. Clinical utility and potential diagnostic pitfalls. *Arch Med Sci.* 2016;12(1):1–9. (In English).
5. Mosli HH, Dennis A, Kocho W, Asher LJ, Van Uum SHM. Effect of short-term proton pump inhibitor treatment and its discontinuation on chromogranin A in healthy subjects. *J Clin Endocrinol Metab.* 2012;97(9): E1731–5.

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