XANTHOMATOUS PSEUDOTUMOR AS A RARE CAUSE OF SMALL BOWEL OBSTRUCTION AND MIMIC OF MALIGNANCY: CASE REPORT AND REVIEW OF THE LITERATURE

Jessica G. Rogers, M.D., Dorina Gui, M.D., Ph.D., Kristin A. Olson, M.D.
Department of Pathology and Laboratory Medicine, University of California, Davis Medical Center, Sacramento, CA

INTRODUCTION

Small bowel obstruction is a relatively frequent event after abdominal surgery, with an incidence of almost ten percent.[1] While the most common cause of postoperative bowel obstruction is adhesions, other classic causes include neoplasm, intussusception, volvulus, foreign bodies, and pseudo-obstruction. We present a highly unusual case of small bowel obstruction due to xanthomatous pseudotumor.

METHODS

A review of the literature was performed with a PubMed search for the phrases "xanthomatous pseudotumor" and "segmental xanthomatosis," as well as the key words "xanthomatosis" and "bowel."

RESULTS

Fewer than five cases of true xanthomatous pseudotumors of the gastrointestinal tract were identified in our search. One case occurred in a 9-year-old boy who had abdominal involvement due to disseminated Burkitt lymphoma. He developed a small bowel obstruction after four months of chemotherapy. Another case occurred in a 22-year-old man who had intestinal obstruction 16 years after radiation therapy for Ewing sarcoma of the right hip. Both individuals required surgery. Another case arose in the stomach of an individual who had undergone chemotherapy for gastric lymphoma. The remaining cases tended to be idiopathic with no postulated etiology.

CASE

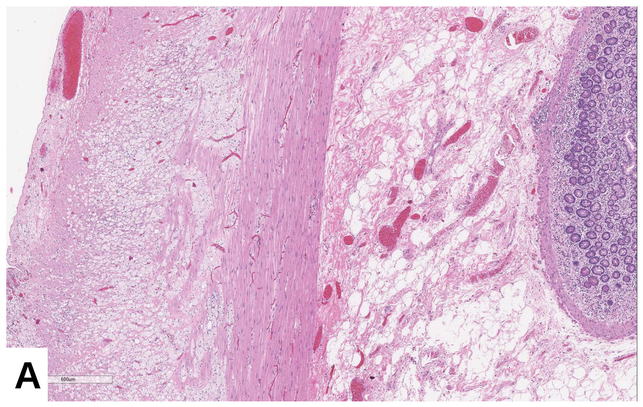
We present a case of a 48-year-old man with obstructive symptoms that developed eight years after he had undergone colectomy and chemoradiation for colon cancer. Abdominal CT scan demonstrated a "transition point" in the small bowel thought to be consistent with mechanical obstruction. The surgical team was very concerned about the possibility of a malignant stricture. The patient was taken to the operating room, where his involved small bowel segment was resected.

Grossly, the small bowel was dilated proximal to a segment with mural thickening and luminal narrowing; on cut section, the thickened bowel wall had a tan-white appearance.

Microscopically, clusters and sheets of foamy histiocytes infiltrated the small bowel serosa and muscularis propria (Figure 1A). The overlying mucosa and submucosa were unremarkable.

A CD163 immunostain confirmed the morphologic impression of histiocytic infiltration of the bowel wall (Figure 1B).

The constellation of findings was considered diagnostic of xanthomatous pseudotumor of the small bowel.



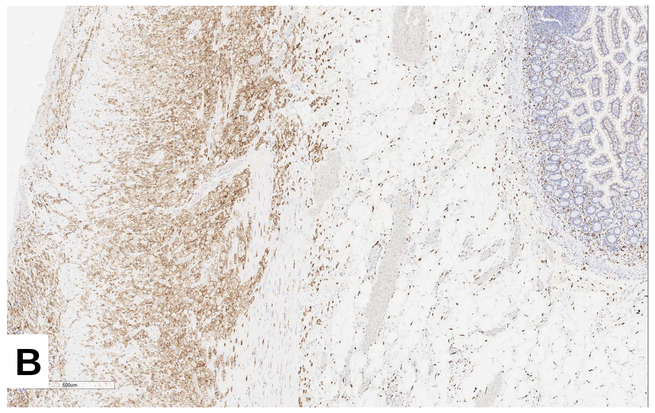
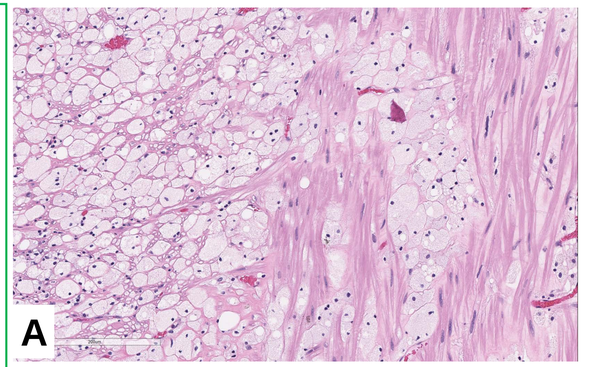


Figure 1: A. Full-thickness bowel wall with histiocytic infiltration (40x, H&E stain). B. CD163 immunostain highlights the histiocytes scattered throughout the bowel wall (40X).

DISCUSSION

While xanthomatous pseudotumor can occasionally be seen involving the kidney or gallbladder, it rarely involves the gastrointestinal tract. The medical literature on xanthomatous pseudotumor of the bowel is quite scant, primarily consisting of case reports[2-5] or small case series. Some authors have proposed that this xanthomatous change represents treatment-related sequelae of a remote metachronous tumor, which appears to apply in our patient as well. Histologically, xanthomatous pseudotumor is characterized by numerous foamy macrophages (histiocytes) with small, centrally located nuclei and plentiful, finely vacuolated, pale to clear cytoplasm. The macrophages tend to be present singly and in clusters, and may assume an "infiltrative" appearance. The differential diagnosis of this entity includes the important diagnostic pitfall of signet ring cell carcinoma. Primary signet ring cell carcinoma of the bowel is an unusual but known entity; fortunately, it is frequently associated with dysplastic surface epithelium and other characteristic histologic features, such as malignant gland formation. The signet ring cells themselves tend to be large and contain hyperchromatic, crescentic nuclei compressed by a mucin vacuole. Differentiation of xanthomatous pseudotumor from metastatic signet ring cell carcinoma can be more challenging, as metastatic signet ring cell carcinoma is often focal and may also be confined to the deeper layers of the bowel wall (Figure 2B). If the morphologic findings are not sufficient to allow for differentiation between the two entities, an immunostain panel may be helpful (e.g., CD68 or CD163 to identify macrophages, and a pancytokeratin stain such as AE1/AE3 to identify signet ring cells).



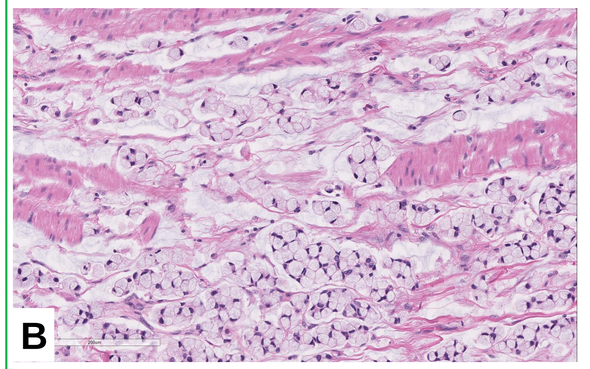


Figure 2: A. Histiocytes in our case of xanthomatous pseudotumor (200x, H&E stain). B. A different case of signet ring cell carcinoma involving the bowel wall (200x, H&E stain).

DISCUSSION (CONT.)

This case is therefore important in that it exemplifies a rare benign condition that can be mistaken by both surgeon and pathologist for a more common malignant condition.

REFERENCES

1. Parikh JA, Ko CY, Maggard MA, Zingmond DS. What is the rate of small bowel obstruction after colectomy? Am Surg. 2008 Oct;74(10):1001-5.
2. Wong KC, Tsui WM, Chang SJ. Xanthogranulomatous inflammation of terminal ileum: report of a case with small bowel involvement. Hong Kong Med J 2015 Feb;21(1):69–72.
3. Victor Delacruz MD, Hidenori Takahashi MD, Seigo Nishida MD, Andreas Tzakis MD, PhD, Phillip Ruiz MD, PhD. Segmental xanthomatosis of the small intestine. A case report and review of the literature. Human Pathology (2009) 40, 139–142.
4. Ashfaq R, Timmons CF. Xanthomatous pseudotumor of the small intestine following treatment for Burkitt's lymphoma. Arch Pathol Lab Med. 1992 Mar;116(3):299-301.
5. Fernandez-Alonso J et al. Xanthomatosis pseudotumor of the stomach after chemotherapy for gastric lymphoma in a patient with acquired immunodeficiency syndrome. Histopathology 1994; 25(6)589-591.

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