Small bowel obstruction is a relatively frequent event after abdominal surgery, with an incidence of almost ten percent. 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.”

Fewer than five cases of true xanthomatous pseudotumors of the gastrointestinal tract were identified in our search. One case arose in the stomach of an individual who had undergone chemotherapy for gastric lymphoma. The remaining cases occurred in the right hip. Both individuals required surgery. Another case occurred in a 22-year-old man who had intestinal obstruction 16 years after radiation therapy for Ewing sarcoma due to disseminated Burkitt lymphoma. He developed a small bowel obstruction after four months of chemotherapy.

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.

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 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).

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