

Extra nodal Rosai Dorfman Disease Presenting as a Colonic Mass

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Introduction

- Rosai-Dorfman disease, also known as sinus histiocytosis with massive lymphadenopathy, is a rare benign non-Langerhans cell histiocytic disease of unknown origin.
- Systemic symptoms of Rosai-Dorfman disease include fever, painless lymphadenopathy, elevated erythrocyte sedimentation rate, leukocytosis, hypergammaglobulinemia, and neutrophilia.
- Rosai-Dorfman disease presents with gastrointestinal involvement in <1% of cases.

Case Presentation

- A 21-year-old woman initially presented to her outpatient surgeons office with a history of multiple lipomas on the inner thigh.
- Results of an excision and biopsy revealed fat necrosis with an associated histolytic reaction consistent with Rosai-Dorfman's disease.
- These lesions were excised and she did not experience recurrence of symptoms.
- Nine years later, she was incidentally found to have mildly elevated liver enzymes and was referred to the outpatient Gastroenterology clinic.
- Initial imaging studies included a CT of the chest abdomen and pelvis, which revealed mild hepatosplenomegaly, diffuse lymphadenopathy in the chest, indeterminate lesions in the colon, a 3.9 cm mass in the cecum, and a new 2.9 cm soft tissue density mass along the right rectal.
- A colonoscopy revealed a non-circumferential, non-obstructing submucosal ulcerated mass in the rectum measuring 3 cm x 3 cm.
- Also visualized during the procedure was a submucosal partially obstructing large cecal mass measuring 5 cm x 4 cm.

Imaging



Figure 1. Contrast enhanced computed tomography of the abdomen and pelvis revealing a 2.9 cm soft tissue density mass along right rectal wall and a 3.9 cm soft tissue density mass in cecum

Colonoscopy

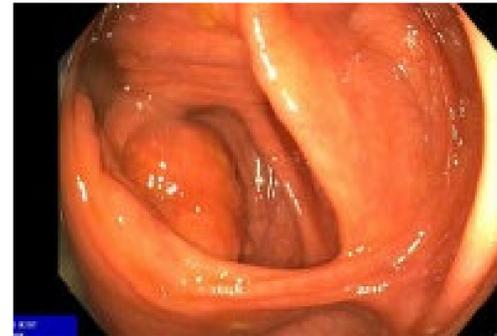


Figure 2. Direct visualization of a 5cm x 4cm partially obstructing mass in the right cecum



Figure 3. Direct visualization of a 3cm x 3cm submucosal and ulcerated non-obstructing mass in the right rectal wall

Histopathology

- Using cold forceps, five irregular pieces of tan, yellow-tinged, shiny, partially friable soft tissue was obtained from the rectal mass and seven similar specimens were obtained from the cecal mass.
- H and E staining revealed fragments of colonic mucosa with hyperplastic glands with reactive changes and focal ulcerations. The lamina propria had atypical histiocytic, some with conspicuous nuclei and some with emperipolesis.
- Immunohistochemistry staining of the specimens showed histiocytic in the lamina propria that were positive for S100, CD163, CD68 (partial), and negative for CD1 and Pan CK.
- These findings were consistent with Rosai-Dorfman's Disease. She is currently following up with oncology to pursue further treatment.

Discussion

- This case presents Rosai-Dorfman disease manifesting with extra nodal disease affecting the gastrointestinal tract, which has rarely been described in the literature.
- Rosai-Dorfman typically manifests as Lymphadenopathy, however this patient has demonstrated two different manifestations of extra-nodal Rosai-Dorfman's disease.
- It also emphasizes the importance of having a heightened suspicion for the potential progression of a usually benign case and the need for surveillance monitoring in patients with evidence of extra-nodal involvement of Rosai-Dorfman disease.

References

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