

Case Report

Adult Langerhans Cell Histiocytosis in a Solitary Colon Polyp

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Abstract

Langerhans cell histiocytosis (LCH) is a rare disorder of unknown pathogenesis. It is characterized by abnormal proliferation and dissemination of Langerhans cells derived from the bone marrow. The disease most commonly affects the bones and the skin. Involvement of the gastrointestinal (GI) tract is extremely rare. We report the case of a 49 year old woman who presented to the clinic with complaints of abdominal bloating and change in bowel habits. She was scheduled for a diagnostic colonoscopy. On colonoscopy, three diminutive polyps were seen at the splenic flexure that were removed with cold biopsy polypectomy. On histology, one of the polyps showed an ovoid collection of atypical cells within the submucosa. The cells were intermediate in size with eosinophilic cytoplasm and ovoid nuclei. The nuclei were variably convoluted with few reniform shapes and some grooves. No cytoplasmic inclusions were identified. The atypical cells extended into the lamina propria. Immunohistochemical staining showed the cells to be strongly positive for CD1a and S100 which confirmed the diagnosis of LCH. On follow up as outpatient, extensive clinical workup showed mild eosinophilia with no evidence of disseminated LCH. Review of published literature showed only a handful of cases of colonic involvement with LCH in the adult population. Most patients have focal disease detected on screening colonoscopy. Systemic involvement is uncommon.

CASE PRESENTATION

A 49 year old woman presented with change in bowel habits and abdominal bloating. She denied overt GI bleeding, weight loss, abdominal pain, nausea or early satiety. Family history was significant for colon cancer in her mother and a maternal aunt. Physical examination was unremarkable. The patient was scheduled for a diagnostic colonoscopy.

On colonoscopy, three diminutive polyps were seen at the splenic flexure. The polyps were removed by cold biopsy polypectomy (Figure 1). A single sessile polyp, measuring 5-10 mm in size was found in the rectum. It was removed by a cold snare polypectomy. Otherwise, the colon appeared to be normal.

Histologically, one of the splenic flexure polyps showed an ovoid collection of atypical cells within the submucosa. These cells were intermediate in size with eosinophilic cytoplasm and ovoid nuclei. Cell borders were indistinct. The nuclei were variably convoluted with a few reniform shapes and some grooves. No cytoplasmic inclusions were identified. The atypical cells extended into the lamina propria. Intermixed with the

atypical cells were large numbers of eosinophils (Figure 2). Immunohistochemical staining showed the cells to be strongly positive for CD1a and S100 and negative for CD117, tryptase and pancytokeratin (Figure 3). These findings supported the diagnosis of Langerhans cell histiocytosis (LCH). The other two

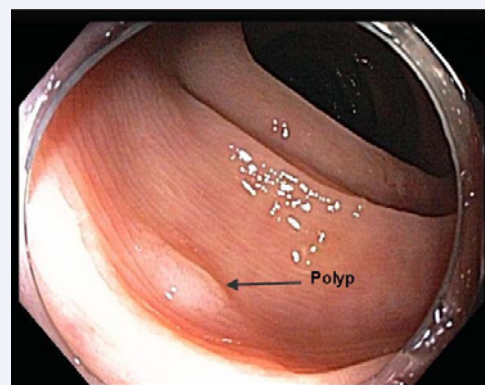


Figure 1 Endoscopic view of Langerhans cell histiocytosis colon polyp.

splenic flexure polyps were hyperplastic polyps. The rectal polyp was a sessile serrated adenoma.

Following the procedure, the patient was evaluated for signs and symptoms of systemic involvement by LCH. She remained asymptomatic. There was no cough or shortness of breath; she did not have any skin lesions. Complete blood count showed 10.3% peripheral blood eosinophils (normal 1%-4%). Liver function tests (including alkaline phosphatase and Gamma-glutamyl transpeptidase) and urinalysis were within normal limits. A chest radiograph showed no acute disease. A computed tomography scan of the chest revealed no pulmonary lesions or pleural effusion. A single 1.3 cm benign-appearing hilar lymph node was documented. There was no mediastinal or axillary lymphadenopathy. Due to the elevated eosinophil count, the patient was referred to Hematology. Work up was non-diagnostic. The patient was advised to have a surveillance colonoscopy in three years.

Langerhans cell histiocytosis is a rare histiocytic disease of unknown etiology and pathogenesis. Gastrointestinal (GI) tract LCH is exceedingly rare and is most often found in male children with high-risk multisystem disease. In adults, studies of GI tract LCH have been limited to a small number of case

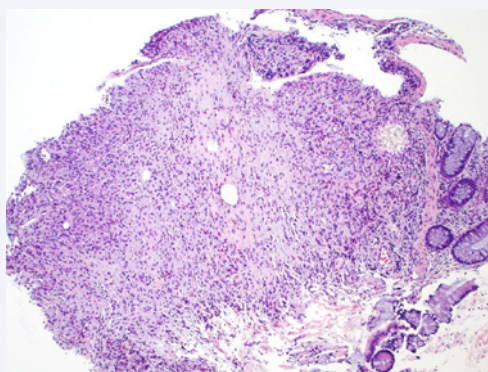


Figure 2a Low-power image of small polyp composed of Langerhans cell histiocytosis. The lesion is characterized by a submucosal proliferation of atypical cells with eosinophilic cytoplasm. (Hematoxylin and eosin, magnification 100x).

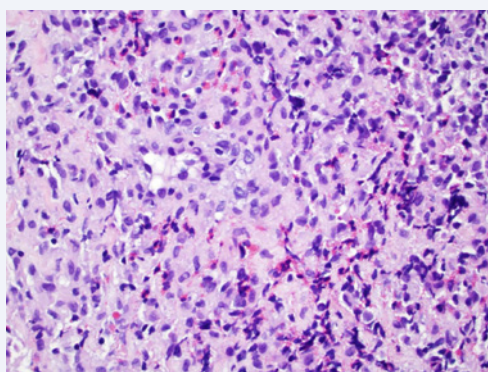


Figure 2b The Langerhans cells have indistinct cytoplasmic borders, eosinophilic cytoplasm and nuclei which are irregular and occasionally reniform and/or contain nuclear grooves. Eosinophils, some of which are degranulating, are intermixed with the Langerhans cells. (Hematoxylin and eosin, magnification 400x).

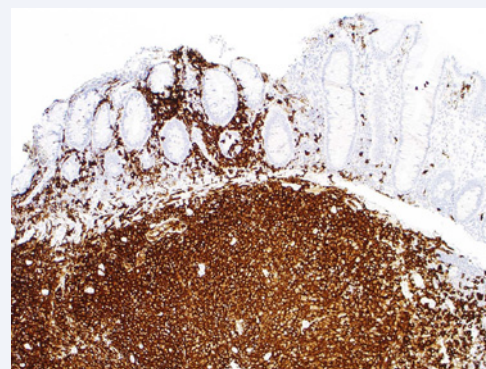


Figure 3a An immunohistochemical stain for CD1a is diffusely and strongly positive in the Langerhans cells. In this level, the Langerhans cells also extend into the lamina propria. (CD1a immunohistochemical stain, magnification 100x).

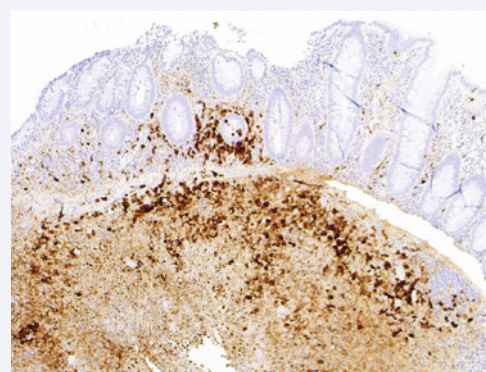


Figure 3b Many of the Langerhans cells are also positive for S100 by immunohistochemistry. (S100 immunohistochemical stain, magnification 100x).

reports. It most commonly affects the bones and the skin. Disease presentations vary from single system lesions in the stomach or liver to multisystem involvement and severe organ dysfunction. A literature search showed only a handful of cases of colonic involvement by LCH in the adult population.

Sharma et al. in 2006, documented a 49 year old male with a single Langerhans cell polyp, measuring 2mm in the ascending colon on routine screening colonoscopy [1]. The patient was asymptomatic and follow-up computed tomography scan of the head and chest X-ray were reported as normal.

In 2009, Kibria et al. reported an isolated 6 mm sessile colonic polyp in the descending colon which showed LCH [2]. The patient was clinically asymptomatic. Chest and skull radiographs, bone scan, abdominal ultrasound, high-resolution computed tomography of the chest and bone marrow aspiration and biopsy were unremarkable.

Singhi et al. , reports the largest series of adult GI tract LCH. Seven of ten patients presented with a solitary colonic polyp. Anatomic distribution was characterized by involvement of the cecum (n=2), ascending colon (n=1), transverse colon (n=1) sigmoid colon (n=2), and anus (n=1), five of the patients with a solitary polyp were asymptomatic and the remaining five

presented with non-specific GI symptoms. Only one patient progressed to extracolonic disease with involvement of the skin [3].

Shankar et al. in 2012 reported a 53 year old woman who was found to have a solitary 4mm sessile polyp on screening colonoscopy which proved to be LCH. The diagnosis was confirmed by immunochemical staining for CD1a antigen. Computerized tomography of the chest, abdomen and pelvis showed no evidence of disseminated LCH. Repeat colonoscopy one year later showed no disease recurrence [4]. Chronic ulcerative mucosa and perianal involvement similar to Crohn's disease has also been reported in LCH [5].

In summary colonic involvement with LCH in adults is extremely rare. Most patients are asymptomatic. LCH is found incidentally in small polyps. Systemic involvement or disease progression is rare.

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