An uncommon cause of small bowel obstruction: isolated primary granulocytic sarcoma

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Case report

A 55-year-old woman was admitted with a 10-day history of colicky, left lower-quadrant abdominal pain and intermittent vomiting. She also reported a 2-month history of alternating diarrhea and constipation, with a 20-pound weight loss. She denied malena or hematemesis. She had a 40-pack year history of smoking.

On admission, she was normotensive, tachycardic and appeared dehydrated. Generalized abdominal tenderness was noted on palpation, with hyperactive bowel sounds and no guarding or rigidity. There was no palpable lymphadenopathy. The remainder physical examination was unremarkable.

Laboratory examination including a complete blood count, Erythrocyte sedimentation rate, liver and renal profile were normal. Stool was positive for occult blood with negative cultures. Multiple air fluid levels were noted on an upright abdominal radiograph. A computerized tomogram (CT) of the abdomen (Figure 1) demonstrated irregular thickened loops of small bowel with a mesenteric mass, suggestive of a primary mesenteric or small bowel tumor. The diagnosis of mechanical small bowel obstruction prompted an explorative laparotomy, which revealed multiple nodular masses in mesentery and small bowel causing luminal narrowing. A wedge resection of the small bowel was undertaken to relieve obstruction.

Histological examination demonstrated, infiltrates of primitive cells involving full thickness of the small bowel and extending into the adjacent mesentery (Figure 2A). There were numerous scattered eosinophils, many of which were myelocytes and metamyelocytes (Figure 2B). Large areas of necrosis were noted in the deeper tissue. A differential diagnosis of lymphoma and granulocytic sarcoma (GS) was considered. Further, immunohistochemical staining was strongly positive for CD43, myeloperoxidase (MPO) (Figure 3A), CD34, CD117 (Figure 3B) and CD45; which were characteristic of GS. A cytogenetic study demonstrated that 92.5% of nuclei had a MYH11/CFB fusion, indicating an inv (16) or a t (8:21). A bone marrow biopsy excluded acute myelogenous leukemia (AML), with no metastasis noted on a staging work-up. The patient had an uneventful recovery. Intensive chemotherapy for AML with high-dose cytosine arabinoside and idarubicin (7 + 3 regime), was instituted on follow-up in the oncology clinic, which the patient has been tolerating well.

Discussion

GS is an extramedullary tumor of immature myeloid series cells. It generally occurs in association with AML, as an initial presentation or a relapse.¹ An isolated primary GS of small intestine in a non-leukemic patient is uncommon.

Primary GS have been described in virtually every anatomic location, with a particular predilection for skin, soft tissue, bone, periosteum and lymph nodes.
Gastrointestinal involvement is infrequent. Of the tumors, >50% are often misdiagnosed as non-Hodgkin’s lymphoma. Acute abdominal pain, from partial or complete bowel obstruction, is the most common clinical presentation. Grossly, the lesions present as polypoid or exophytic masses, regions of wall thickening and/or ulcerations, with a high proclivity for mesenteric and peritoneal spread.

Histologic features consist of diffuse, infiltrating medium to large cells, with occasional prominent nucleoli and minimal to moderate eosinophilic cytoplasm. The presence of admixed maturing eosinophils typically supports the diagnosis of GS. Positive stains for MPO, lysozyme (Ly) (89–90%), naphthol AS-D chloracetate esterase (75–80%), CD34 and CD117,1 are characteristic of GS. The tumor cells frequently stain with T cell markers such as CD43 and variably stain with B cell markers such as CD79a. This may lead to an erroneous diagnosis of lymphoma, if a GS is not considered.2

GS has been associated with various cytogenetic abnormalities, particularly t(8;21) (54%) and less frequently, inv (16) (p13; q22) (25%). The prognostic significance of these chromosomal rearrangements in primary GS remains uncertain. The molecular counterpart of inv (16) is the chimeric gene

Figure 1. CT abdomen with contrast (coronal section) showing irregular thickened small bowel loops (mid-distal) with adjacent mesenteric mass (arrow).

Figure 2. (A) Jejunal and adjacent mesenteric biopsy specimen showing a dense infiltrate of myeloid precursor cells involving full thickness of the bowel and extending into the adjacent mesentery (40X H&E stain). (B) Biopsy specimen showing sheets of myeloid precursor cells (blue) with nuclear cleavages, indentations, small nucleoli and moderate cytoplasm. Numerous scattered eosinophils (arrows) are noted (200X H&E stain).

Figure 3. (A) Immunohistochemical stain for MPO reveals positive granular cytoplasmic staining of most of the myeloid cells (brownish red staining) (400X). (B) Immunohistochemical stain for CD117 (c-kit) showing staining of the cell membrane of the tumor cells (violet staining) (400X).
CBFβ/MYH11 that involves the core binding factor β and the smooth muscle myosin heavy chain genes.\textsuperscript{4}

The optimal therapy for primary GS remains undetermined. If left untreated, most cases (88\%) progress to AML in a mean duration of 11 months.\textsuperscript{1} Traditional chemotherapy for AML (high-dose cytosine arabinoside and anthracycline) can prolong non-leukemic periods to 2 years, and is advocated in all patients. Supplementary surgical removal and/or local radiation therapy may result in longer remissions.\textsuperscript{1} Targeting c-kit (CD117) expressing tumors with tyrosine kinase inhibitors is a novel therapeutic option.\textsuperscript{2} Prognosis is variable and somewhat similar to AML.

In conclusion, primary GS of small intestine is an uncommon entity with a unique histopathology mimicking other solid neoplasms, making it a diagnostic challenge. Prompt recognition and early institution of therapy is crucial, and may delay its progression to a systemic disease.

Conflict of interest: None declared.

References