CASE REPORT

Granulocytic sarcoma (chloroma) presenting as colo-colic intussusception in a 16-year-old boy: an unusual presentation

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SUMMARY

A 16-year-old boy presented with intestinal obstruction with left lumbar mass. On evaluation with ultrasonography and contrast-enhanced CT of abdomen he was found to have left colo-colic intussusception. On exploratory laparotomy he was found to have invagination of necrosed distal transverse colon and splenic flexure with omentum into descending colon with enlarged mesenteric lymph nodes for which left hemicolectomy was performed. Histopathology and immunohistochemistry was suggestive of submucosal infiltration by immature myeloid precursors with few metamyelocytes and occasional eosinophils and the cells showed cytoplasmic granular positivity with myeloperoxidase stain suggestive of granulocytic sarcoma (chloroma—extra-medullary manifestation of acute myeloid leukaemia). Our patient received three cycles of cytarabine+daunorubicin chemotherapy in the postoperative period and is doing well at 12 months of follow-up. Granulocytic sarcoma (chloroma) presenting as colo-colic intussusception is very rare and has not been reported in literature so far.

BACKGROUND

Granulocytic sarcoma (also known as chloroma or myeloblastoma or extramedullary myeloid tumour or myeloid sarcoma) is a rare solid tumour composed of primitive precursors of granulocytic series of white blood cells that include myeloblast, promyelocytes and myelocytes. It is called as chloroma, because typical forms may have a green colour caused by high levels of myeloperoxidase in immature cells. Most common areas of involvement are skin and gums, lymph nodes, mediastinum, small intestine, epidural sites, uterus and ovaries. Gastrointestinal tract involvement is unfamiliar and especially the involvement of large gut (colon) is very rare in literature. Granulocytic sarcoma can precede the onset of acute myeloid leukaemia or can also occur during active phase of leukaemia and the prognosis of granulocytic sarcoma is similar to myeloid leukaemia. Chemotherapy is the treatment of choice. The development of acute myeloid leukaemia usually occurs within 11 months of diagnosis of chloroma if left untreated. So chemotherapy should be initiated after definitive management (surgery) if possible. This is a very rare case of granulocytic sarcoma (chloroma) presenting as colo-colic intussusception, as incidence of granulocytic sarcoma in colon is unusual.

CASE PRESENTATION

A 16-year-old boy was admitted in emergency surgery ward with constipation, distention abdomen and vomiting for 7 days. The patient also had history of on and off colicky pain around the umbilicus for 1.5 months. There was no history of fever, bleeding per rectum, jaundice or any urinary symptoms; thus history being suggestive of acute intestinal obstruction. On clinical examination, abdomen was found tense, distended and tenderness was more prominent in left lumbar region. A well-defined oval mass was palpable in left lumbar region. On digital rectal examination, no faecolith and no blood staining of fingers was found.

INVESTIGATIONS

Plain X-ray abdomen showed multiple air fluid levels suggestive of obstruction. Ultrasonographic findings were suggestive of intussusception of gut in left lumbar region. Contrast-enhanced CT of abdomen was performed which confirmed the diagnosis of colo-colic intussusception in left lumbar region.

TREATMENT

Diagnosed as a case of acute intestinal obstruction due to colo-colic intussusception, he was shifted to operating room and exploratory laparotomy was performed. Intraoperative findings revealed distended large gut with invagination of transverse colon along with omentum into the descending colon (transverse colon acting as intussusceptum and descending colon as intussuscepti; [figures 1 and 2](#)). Milking of descending colon was carried out to evacuate the loop of transverse colon. A constricting growth was found at splenic flexure which acted as lead point for intussusception. Hard enlarged lymph nodes were found in mesocolon. Left hemicolectomy ([figure 3](#)) was performed by anastomosing transverse colon to descending colon as the involved portion was gangrenous. Abdomen was closed in layers with drains in situ. The excised specimen along with lymph nodes was submitted for histopathological examination.

OUTCOME AND FOLLOW-UP

Postoperative period was uneventful. The excised specimen was submitted for histopathological examination. Histopathology revealed submucosal infiltration by immature myeloid precursors with few metamyelocyte and occasional eosinophils

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Figure 1 Intussusception of splenic flexure into descending colon.

(figures 4–6). The cells showed cytoplasmic granular positivity with myeloperoxidase stain on immunohistochemistry. Morphology and immunohistochemistry suggested the diagnosis of granulocytic sarcoma (chloroma). Bone marrow examination revealed mildly megaloblastic erythropoiesis and reactive changes in granulocytes. Peripheral blood film showed normocytic normochromic anaemia with blast cells and left shift. The

Figure 2 Intussuscepted segment.

Figure 3 Hemicolectomy specimen with enlarged lymph nodes.

patient was further planned for chemotherapy (cytarabine and daunorubicin) as development of acute myeloid leukaemia is nearly universal in short term after the detection of chloroma. He received three cycles of cytarabine+daunorubicin chemotherapy and is doing well after 1 year of surgery. Patients of granulocytic sarcoma should be followed up with peripheral blood smears and bone marrow aspirate.

DISCUSSION

Chloroma, a rare tumour was first described by the British physician A Burns in 1811; is basically any extra medullary manifestation of acute myeloid leukaemia.¹ Granulocytic sarcoma or chloroma, also called extramedullary myeloblastoma or myeloid sarcoma is a rare malignant tumour and is mainly associated with myeloblastic leukaemia.² Granulocytic sarcoma is essentially a solid tumour composed of granulocyte precursor cells.³ It can occur during leukaemia relapse or remission and may be the sole presentation of the disease.⁴ It has a predisposition for

Figure 4 Submucosal tumour infiltrates and normal intestinal glands.

Figure 5 Myeloid cell population.

the central nervous system, bones, soft tissues of the head and neck and skin.⁵ In gastrointestinal tract small bowel involvement is much more common than large bowel with small bowel involvement estimated to be around 10%.^{6–7} Involvement of large bowel is evident mainly in few case reports.^{8–10}

Chloroma or granulocytic sarcoma may occur in patients with diagnosis of myelodysplastic syndrome or myeloproliferative syndrome (eg, chronic myelogenous leukaemia, polycythemia vera, essential thrombocythosis or myelofibrosis). In such conditions it is evident that these premalignant conditions have transformed into an acute leukaemia suggesting blast crisis. Myeloblastomas are extramedullary tumours of myeloblasts and other immature myeloid cells appearing either green in colour (chloroma) or non-pigmented. The green colour is related to cytoplasmic myeloperoxidase (verdoperoxidase) and porphyrins, possibly destined originally in a common stem cell for erythroblastic precursors.¹¹ Involvement of the gastrointestinal tract in different haematological malignancies usually occurs in advanced stage of the disease or as a complication of chemotherapy and radiotherapy. Most common findings on endoscopy are esophagitis, colitis, gastritis, gastric erosions, duodenitis, proctitis and polyps.¹² Gastrointestinal tract involvement by myeloid sarcoma is very rare and can present with abdominal pain, bleeding, perforation, obstruction, intussusception, liver infarction, pancreatitis, appendicitis, bile duct obstruction or portal hypertension.

Figure 6 High-power view of myeloid cells.

Very rarely, primary chloroma can occur without known pre-existing diagnosis of acute leukaemia, acute promyelocytic leukaemia or myelodysplastic syndrome, myeloproliferative syndrome, which is the same as in our case. In almost all reported cases of primary chloroma, acute leukaemia developed shortly afterwards (median time 7 months, range 1–25 months).¹³ Therefore, it should be considered as initial manifestation of acute leukaemia, rather than a localised process. The prognosis is grim.

Involvement of colon by granulocytic sarcoma is very rare and its presentation as colo-colic intussusception has not been reported in the literature so far. Patients with granulocytic sarcoma should be regularly followed with peripheral blood smears and bone marrow aspirates to look for development of blast crisis. Our patient is doing well at 1 year of follow-up.

Learning points

- ▶ Granulocytic sarcoma or chloroma, also called extramedullary myeloblastoma or myeloid sarcoma is a rare malignant tumour and is mainly associated with myeloblastic leukaemia.
- ▶ It should be considered as initial manifestation of acute leukaemia, rather than a localised process and acute leukaemia developed shortly afterwards (median time 7 months, range 1–25 months).
- ▶ Gastrointestinal tract (especially colon) involvement by myeloid sarcoma is uncommon.
- ▶ Patients with granulocytic sarcoma should be regularly followed with peripheral blood smears and bone marrow aspirates to look for development of blast crisis.
- ▶ Appropriate candidates would need chemotherapy with anthracyclines and cytarabine.

Competing interests None.

Patient consent Obtained.

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