Case Report

Myxoid liposarcoma of small bowel presenting with intussusception and obstruction: A case report

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ABSTRACT

Gastrointestinal liposarcomas are extremely rare with the most common reported morphological subtype being dedifferentiated liposarcoma and well-differentiated liposarcoma. These tumors are rarely diagnosed preoperatively and diagnosis is only confirmed on histopathological analysis. Treatment of gastrointestinal liposarcomas consists of surgical excision with widely negative margins followed by post-operative irradiation and close follow-up. We report an exceedingly rare case of myxoid liposarcoma of the small bowel (ileum) presenting with an unusual presentation with intussusception and intestinal obstruction. A 42-year-old male presented to the emergency department with features of intestinal obstruction. Contrast-enhanced computed tomography abdomen revealed ileo-ileal intussusception with an endoluminal soft-tissue lesion at the leading edge. The patient was taken for surgical intervention and the involved segment of the bowel along with the lesion was resected and re-anastomosis done. Histological sections of the mass along with immunohistochemistry suggested the pathological diagnosis of myxoid liposarcoma.

Key words: Gastrointestinal liposarcoma, Intussusception, Myxoid liposarcoma

Liposarcoma is the most common malignant mesenchymal neoplasm. It is also the most common primary tumor of the thigh and retroperitoneum. Primary gastrointestinal (GI) liposarcoma is a rare entity. The majority of them arise from the esophagus, closely followed by the stomach, small intestine, and large intestine [1]. Dedifferentiated and well-differentiated morphological forms of liposarcomas have been described as the most common subtypes seen in the GI tract. Only a few case reports have described the rare occurrence of myxoid liposarcomas [2-4].

We report an exceedingly rare case of GI myxoid liposarcoma in the small bowel (ileum) and presenting with an unusual presentation of intussusception and intestinal obstruction.

CASE REPORT

A 42-year-old male presented to the emergency department with recurrent episodes (1–2 episodes/month) of severe abdominal colic, and non-projectile vomiting for the past 6 months.

On examination, the vitals were stable except for mild tachypnea and abdomen distension was noted.

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Abdominal radiographs revealed dilated small bowel loops with multiple air-fluid levels. Contrast-enhanced computed tomography (CT) abdomen was advised to rule out the cause of obstruction. CT revealed telescoping of a loop of the ileum into the distal part of the loop giving bowel within the bowel configuration suggesting intussusception. A relatively well-demarcated enhancing endoluminal soft-tissue mass measuring approximately 3 × 3 cm was seen at the leading edge (Fig. 1).

The patient was considered for surgical intervention to relieve the cause of obstruction. The involved segment of the bowel along with the lesion was resected and re-anastomosis was done. Intraoperatively, ileo-ileal intussusception was present and a well-circumscribed and encapsulated soft to firm endoluminal mass was noted at the leading edge. The post-operative period was uneventful.

The resected bowel along with the soft-tissue mass was sent for histopathological examination. Histological sections revealed diffuse proliferation of the low-grade round to oval stellate and fusiform cells with mild nuclear pleomorphism, vesicular nuclei, and vacuolated cytoplasm (Fig. 2a). Scattered lipoblast-like cells in the background of myxoid stroma and thin-walled vascular proliferation were seen (Fig. 2b). On immunohistochemistry, lipoblast-like cells were positive for S-100 (Fig. 3). On the basis of these findings, the pathological diagnosis of myxoid

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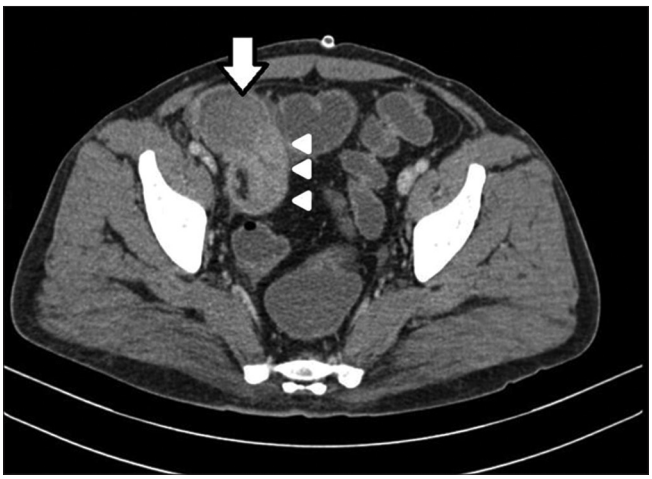

Figure 1: (Original): Contrast-enhanced CT images show a well-defined homogeneously enhancing mass (white arrow) at the leading edge of intussusceptum. Note that the typical bowel within the bowel configuration of intussception is also well-demonstrated (arrowheads)

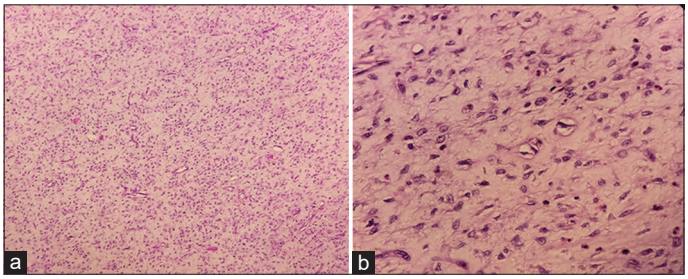

Figure 2: (Original): (a) H&E section shows diffuse proliferation of round to oval stellate and fusiform cells with thin walled vascular proliferation (×10); (b) H&E section shows stellate cells having mild nuclear pleomorphism vesicular nuclei and vacuolated cytoplasm in a myxoid stroma. Scattered lipoblasts are seen (×40)

liposarcoma was made. Adjuvant radiotherapy was initiated and close follow-up imaging was advised.

DISCUSSION

Liposarcomas are the most common soft-tissue sarcomas. According to the World Health Organization 2020 classification, liposarcomas have been classified into these morphologic subgroups: Well-differentiated liposarcoma, myxoid liposarcoma, dedifferentiated liposarcoma, pleomorphic liposarcoma, and pleomorphic myxoid liposarcoma. The well-differentiated type is the most common histological type and has a relatively better prognosis, while the pleomorphic cell subtype carries a higher risk of post-operative recurrence [5].

The lower extremities and retroperitoneum are the most common sites of involvement. GI tract involvement is extremely rare. The esophagus has been reported as the most common site of involvement. A slight male predominance is seen. The clinical presentations of GI sarcomas are non-specific. The reported clinical symptoms include abdominal pain, diarrhea, weight loss, hematemesis, melena, anemia, constipation, and a palpable mass. It may also result in intestinal obstruction, intussusception [6-8], or may mimic acute appendicitis [4].

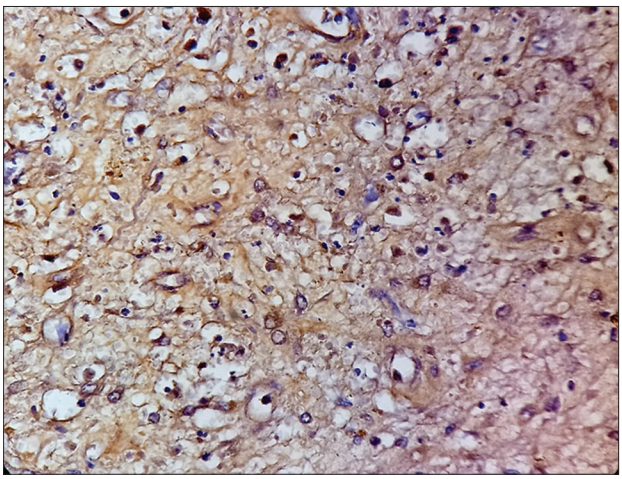

Figure 3: (Original): Immunohistochemistry (S100) shows lipoblasts like cells showing nuclear positivity (×40)

GI liposarcomas are rarely diagnosed on imaging. CT and magnetic resonance imaging usually demonstrate an enhancing heterogeneous soft-tissue mass. Diagnosis is only confirmed on histopathological analysis. In the GI tract, the most commonly reported morphological subtype is dedifferentiated liposarcoma and well-differentiated liposarcoma. On histology, well-differentiated liposarcoma is composed of mature lipomatous proliferation interspersed with thick fibrous septa. Cellular pleomorphism and nuclear atypia in adipocytic and stromal cells are seen. Mitotic figures are rare with variable amounts of lipoblasts. Myxoid liposarcoma is composed of hypocellular bland spindle cell proliferation and monovacuolated lipoblasts, set in a myxoid stromal background with a prominent plexiform capillary network. A t(12;16)(q13;p11) or t(12;22)(q13;q12) translocation is seen in the majority of the cases of myxoid/round cell liposarcoma. Dedifferentiated liposarcomas are characterized by a broad morphological spectrum. Most often the dedifferentiated component is pleomorphic; however, monomorphic spindle cell proliferation can be seen [9].

Treatment for liposarcomas is surgical resection. These neoplasms have significant potential to recur after incomplete surgical excision. Therefore, widely negative margins must be obtained [1]. When a sufficient resection margin cannot be obtained, post-operative irradiation may be performed. Subsequent close follow-up is warranted.

CONCLUSION

We have described a rare case of myxoid liposarcoma of small bowel presenting with intussusception and obstruction. These tumors are rarely diagnosed preoperatively and diagnosis is only confirmed on histopathology. Treatment of GI liposarcomas includes surgical excision with widely negative margins followed by post-operative irradiation.

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