Case Report

CT of primary undifferentiated pleomorphic sarcoma of small intestine with distant metastases presenting with intussusception

Saneesh P S*, Pooja Verma, Shahina Bano, G R kumar Hemanth
Radiodiagnosis Department, PGIMER & RML Hospital, New Delhi, India

ARTICLE INFO

Article history:
Received 23 July 2019
Revised 5 October 2019
Accepted 9 October 2019
Available online 9 November 2019

Abstract

Undifferentiated pleomorphic sarcoma of bowel is very rare, only a few cases have been reported till date. A 29 year old male patient presented with acute abdominal pain due to intussusception, contrast enhanced computed tomography abdomen showed multiple endoluminal homogenously enhancing polyoid lesions with distant metastasis. Radiologically our primary diagnosis was malignant gastrointestinal stromal tumor, however, histopathological evaluation turned out to be a small bowel undifferentiated pleomorphic sarcoma. Considering the multiplicity of lesion and early distant metastasis radiologist should also consider sarcoma involving bowel as one of the differential diagnosis.

INTRODUCTION

Undifferentiated pleomorphic sarcoma (UPS) was previously known as malignant fibrous histiocytoma (MFH). UPS usually occurs in adults with the most common location being extremities followed by retroperitoneum. However, primary UPS of bowel is extremely rare, with 24 cases reported in literature till date. It is a rapidly growing tumor with a propensity for early distant metastasis. We report a case of primary UPS of bowel which presented as multifocal endoluminal polyoidal growth in ileum and jejunal with lung, mediastinal, and skull metastasis.

Case report

A 29-year-old male patient presented to the emergency department with complaints of acute abdominal pain. He had a history of intermittent vague abdominal pain, vomiting, melena and right-sided scalp swelling since 1 month duration and 3 episodes of convulsions in last week. The patient underwent ultrasonographic evaluation, which revealed jejunojejunal and ileoileal intussusceptions (Fig. 3A and B). Erect abdominal radiograph showed features of small bowel obstruction however there was no features of pneumoperitoneum. Chest radiograph revealed mediastinal widening with prominent left hilum. The hemogram revealed anemia and increased total leukocyte count. The patient was taken up for emergency open laparotomy, which revealed multiple polyoidal growths in small intestine and few lesions at the lead point.
were resected and sent for histopathological examination. On postoperation day 3, patient underwent computed tomography evaluation, which was done in 128 Dual energy Siemens CT scanner. 1000 mL of positive oral contrast was given to the patient, followed by intravenous contrast. Contrast-enhanced computed tomography (CECT) abdomen showed multiple intraluminal polypoidal growths in ileum and jejunum, largest measuring 7.5 × 4 cm. Lesions showed mild homogenous postcontrast enhancement (Fig. 1A and B). No evidence of necrosis or hemorrhage noted within the lesion. There were no features of acute intestinal obstruction, mesenteric, and retroperitoneal lymphadenopathy. CECT chest showed large conglomerated nodal mass lesion involving anterior, medial, and posterior mediastinum (Fig. 1C and D). On lung window, multiple nodules were noted in bilateral lung parenchyma (Fig. 2A and B). Other organs such as liver, adrenals, kidney, and spleen were normal. CECT Brain revealed enhancing subgaleal and extradural soft tissue lesion with lytic bone destruction of right parietal bone (Fig. 2C and D). Histopathological and immunohistochemistry reports revealed undifferentiated pleomorphic sarcoma. On gross examination, received part of intussusceptions measured 52 cm in length and diameter of cut ends were 0.5 cm and 4.5 cm. Serosal surface was covered by exudates and showed multiple perforations at a distance of 9 cm from the first cut end and multiple indurated ulcers were identified ranging from 2 to 5 cm. Intussusception segments showed congestion and gangrenous mucosa. Microscopy revealed tumor cells in submucosa arranged in sheets, short intersecting fascicles. Cells are large, oval to spindle with marked nuclear pleomorphism, coarse chromatin 1-4 conspicuous nucleoli and moderate amount of eosinophilic cytoplasm (Fig. 3C). Many tumor giant cells and brisk mitotic activity were seen along with the necrotic area. Tumor cells were infiltrating transmurally.
into serosa and overlying mucosa. The background showed lymphoplasmacytic infiltrate. Adjoining intestine showed transmural hemorrhagic necrosis. Immunohistochemistry showed PanCK negative, Desmin negative, SMA negative, and CD117 negative.

**Discussion**

UPS previously referred as malignant fibrous histiocytoma, with extremities being the most common location (lower extremities—49%; upper extremities—19%) followed by retroperitoneum (16%) [1], can involve any organ such as head, neck, dura, brain, heart, lungs, pancreas, aorta, spleen, and liver. Visceral organ involvement is very rare. The prognosis of UPS of extremities is generally poor with metastasis arising in 30%-50% cases, but better as compared to abdominal involvement, because of early detection of lesion and easy accessibility for biopsy and histopathological evaluation in UPS of extremities and late presentation, diagnostic dilemma and poor accessibility for biopsy in abdominal UPS. Small bowel UPS can present as abdominal pain, vomiting, fever, weight loss, malena, abdominal distension, palpable abdominal mass, and intussusception. The usual CT imaging finding of UPS is well-defined homogenous mass with or without

Fig. 2 – Axial lung window images A and B showing multiple nodules noted in anterior segment of Right upper lobe (→) and superior segment of left lower lobe with feeding vessel sign (←→). Axial CECT brain image C showing enhancing subgaleal and extradural soft tissue lesion (←→) with lytic destruction of right parietal bone (←→) in bone window (D).
Fig. 3 – USG abdomen in sagittal section (A) and axial section (B) showing small bowel intussusceptions. Photomicrograph (C) showing: Cells are large, oval to spindle, with marked nuclear pleomorphism, coarse chromatin and conspicuous nucleoli and moderate amount of eosinophilic cytoplasm. Many tumor giant cells and brisk mitosis.

eiscrosis and hemorrhage [2,3]. In our case, it presented as multifocal mildly enhancing endoluminal polypoidal mass in ileum and jejunum. Our case was in concordance with the previous studies, the most common site of bowel involvement is ileum and jejunum, however, few cases of duodenal and ampulla of Vater involvement was reported [6].

In our case, the primary diagnosis was malignant gastrointestinal stromal tumor (GIST), however other differential diagnosis includes, lymphoma and adenocarcinoma of small intestine. Malignant GIST usually exhibit exoluminal or endoluminal growth with areas of necrosis/hemorrhage and heterogeneous enhancement in postcontrast imaging. However, considering the multiplicity of the lesion, homogenous enhancement pattern and early distant metastasis to lungs, mediastinum, and skull was not favoring for malignant GIST. GIST usually metastasizes to liver and peritoneum. The second possibility we considered was lymphoma, but in our case, there was no evidence of associated retroperitoneal or mesenteric lymphadenopathy. Lymphomas usually does not cause intestinal obstruction, more or less it causes aneurysmal dilatation of the bowel loops. We also considered the possibility of small intestinal adenocarcinoma as a differential, but usual imaging appearance of adenocarcinoma is short segment irregular circumferential wall thickening or heterogeneously enhancing luminal growth with areas of necrosis and hemorrhages and associated significant mesenteric
lymphadenopathy. Other possible diagnosis includes pleomorphic leiomyosarcoma, liposarcoma, and rhabdomyosarcoma, but these are indistinguishable from UPS on imaging.

In our case, the patient was completely asymptomatic 1 month before. He had a history of vague abdominal symptoms for 1 month duration with sudden onset of abdominal pain due to intussusception. CT imaging revealed multiple endoluminal bowel lesions with distant metastasis to lung, mediastinum, and skull at the same time. As considering the short duration of clinical presentation, multiplicity of bowel lesions and associated multiple metastatic foci. UPS bowel is a tumor having rapid growth patterns and early distant metastasis. As compared with previous studies, intestinal UPS can present as early distant metastasis with metastatic rate of 42% (1).

The treatment modality of UPS is early surgical resection with negative margins. Adjuvant radiotherapy is well effective for UPS of extremities; however, it remains unclear in cases of UPS involving the bowel. Chemotherapeutical agents like Doxorubicin and Ifosfamide is reserved for unresectable and metastatic cases [4,5].

UPS have a very poor prognosis with 2 year survival rate of 60%. The recurrence rate is 44% and the metastasis rate of 42% [1].

Conclusion

We report a case of UPS of bowel presenting with acute abdominal pain due to intussusception. CT imaging revealed multiple endoluminal polypoidal lesions in ileum and jejunum with early distant metastasis to lung, mediastinum, and skull. Our primary diagnosis was malignant GIST, other differential diagnosis of lymphoma and adenocarcinoma of small bowel were considered. However, histopathology and immunohistochemistry revealed it as a case of UPS of small intestine. Considering multifocality of primary lesion and early distant metastasis, the radiologist should always consider the possibility of sarcoma involving bowel as one of the differential diagnosis along with other common malignant lesions.

References

[1] Weiss SW, Enzinger FM. Malignant fibrous histiocytoma. An analysis of 200 cases. Cancer 1978;41(6):2250–66.
[2] Goldman SM, Hartman DS, Weiss SW. The varied radiographic manifestations of retroperitoneal malignant fibrous histiocytoma revealed through 27 cases. J Urol 1986;135(1):33–8.
[3] Park JH, Yeon JW, Han EM, Jang SK, Kang SM, Ahn IO. Primary malignant fibrous histiocytoma of the mesentery: a case report. J Korean Radiol Soc 2007;57(6):549–52.
[4] Nascimento AF, Raut CP. Diagnosis and management of pleomorphic sarcomas (so-called “MFH”) in adults. J Surg Oncol 2008;97(4):330–9.
[5] Sleijfer S, Seynaeve C, Verweij J. Using single-agent therapy in adult patients with advanced soft tissue sarcoma can still be considered standard care. Oncologist 2005;10(10):833–41.
[6] Kim YR, Lee YH, Yoon KH, Yun KJ. CT findings of primary undifferentiated pleomorphic sarcoma in the small bowel: a case report. J Korean Soc Radiol 2015;73(5):323–7.