

A Rare Case of Osteosarcoma Metastasis to the Small Bowel Causing Intussusception in a Pediatric Patient: A Case Report and Review of Literature

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INTRODUCTION

Osteosarcoma is the most common malignant tumor of the bone. The lungs are a predominant site of metastases from osteosarcoma. Metastatic osteosarcoma to the abdomen is very rare (1). A few cases involving metastatic osteosarcoma (OS) causing small bowel intussusception have been reported. (1-9) A review article by Serpico et al. identified 42 patients with metastatic OS to the abdomen (1). To the best of our knowledge, 12 OS patients with abdominal metastasis to the small bowel were reported to have intussusception. Of these, only 3 cases were reported previously in pediatric patients (2,3,4).

Although small bowel intussusception often resolves spontaneously, the presence of a leading point is a clear indication of surgical intervention. Patients with oncological disease comprise a subgroup in which medical caregivers should maintain a low threshold for investigating the presence of metastasis as a leading point.

Here we present a rare case of a pediatric female patient with osteosarcoma who developed metastasis-induced small bowel intussusception.

CASE PRESENTATION

We report a case of a 13-year-old female who was diagnosed with stage IV shoulder osteosarcoma at the age of 12 years. The patient had progressive left shoulder pain, where MRI showed a left proximal humerus bone mass. A bone biopsy was done and revealed osteosarcoma. A staging CT scan showed multiple bilateral lung nodules measuring up to 0.5 cm suggestive of metastatic lung nodules. PET CT scan showed no other hypermetabolic lesions suggestive of other metastases. The patient had no other medical illnesses and no past surgical history. She underwent neoadjuvant chemotherapy following the MAP protocol (Methotrexate, Doxorubicin, Cisplatin) and subsequent local control via surgical resection, with the final pathology revealing 95% necrosis. Post-operative chemotherapy was administered, and a follow-up CT showed resolution of the lung nodules. The patient presented with a prolonged course of febrile

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neutropenia approximately 6 months post-presentation; CT chest demonstrated the emergence of a lung lesion with central cavitation, a small gas locule, and a surrounding ground glass shadowing which was suspected to be an atypical or fungal infection. The patient received medical treatment, but due to the progression of this area on subsequent CT scans, the patient underwent a lobectomy 9 months after the initial diagnosis, with the pathology confirming completely resected osteosarcoma.

Two months later, the patient presented to the emergency room with severe abdominal pain but without other associated symptoms. Her vital signs were within normal limits, and her blood workup was also normal, except for a hemoglobin level drop to 6.9 g/dl from a baseline of 10 g/dl. Her WBC count was $5.7 \times 10^3/\text{ul}$ and her platelet was $257 \times 10^3/\text{ul}$. Liver function, renal function, and electrolytes were unremarkable. The patient had no melena, no fresh bleeding per rectum, and no hematuria. She stated that she lost her menses while on chemotherapy, but after she finished it, her menses became very heavy, with significant bleeding in the last cycle. Her last menses was one week prior to her presentation to the ER. Anemia was attributed to heavy menstruation. The patient received blood in the emergency room, abdominal pain improved with conservative treatment and was discharged. Four days later, she returned to the emergency room with abdominal pain accompanied by bilious vomiting, she had bowel movement one day prior to the presentation. On physical exam the abdomen was soft, and she had epigastric tenderness with no distension. She had no fever but had mild tachycardia up to 104 bpm. Her HB was 10.3 g/d and her WBC was $6.3 \times 10^3/\text{ul}$. Her renal function and electrolytes were unremarkable. An abdominal CT scan revealed a 5 cm proximal jejunal intus-

susception (Figure 1), with no signs of ischemia or significant bowel dilation. The patient was admitted to the hospital and kept on nil per os (NPO) status with serial abdominal examinations to monitor for obstruction or peritonitis signs. A follow-up ultrasound demonstrated persistent intussusception.

The patient underwent a diagnostic laparoscopy, which identified a small bowel intussusception 10 cm distal to the duodenojejunal junction. The intussusception could not be reduced laparoscopically, leading to conversion to laparotomy. The small bowel was reduced, revealing a mass as the leading point (Figure 2). Small bowel resection and anastomosis were performed. The immediate post-operative period was unremarkable.

The pathology examination revealed a 6.5 cm long bowel segment with an unremarkable outer surface except for a centrally puckered area. Upon opening, the lumen exposed an intraluminal exophytic hemorrhagic necrotic tumor measuring 2.6 cm in maximum dimension, confined to the mucosa and away from both surgical resection margins (Figure 3). Microscopic examination showed a polypoid growth within the lumen, with no tumor involvement of the wall or serosa. The tumor comprised highly pleomorphic spindle cells with bizarre hyperchromatic nuclei, brisk mitotic activity, and numerous atypical mitotic figures. Osteoid material was also present within the tumor (Figures 4a, b). Metastatic osteosarcoma was diagnosed. A PET CT scan was done and showed no other hypermetabolic lesions suggestive of metastasis. The family and the patient declined further chemotherapy. Two months later, she developed multiple bilateral lung metastases, in addition to liver and bone metastases, and was started on palliative treatment. The patient passed away 5 months after the reported event.

Figure 1. An abdominopelvic computed tomography showing jejunojejunal intussusception (arrows).

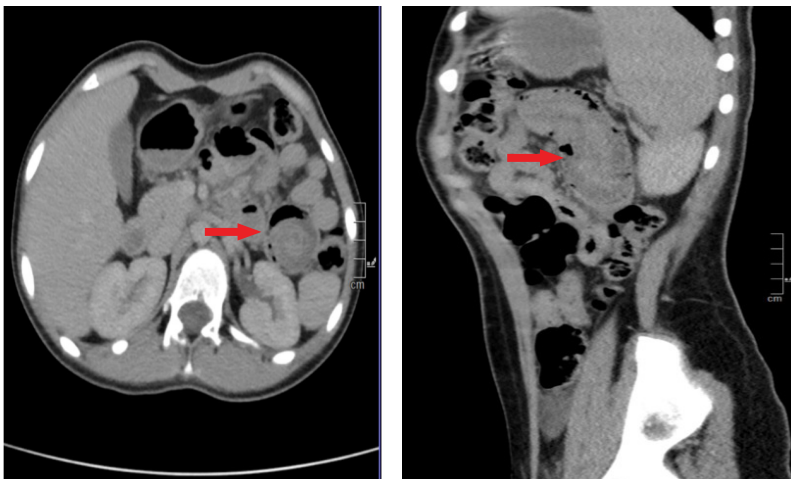


Figure 2. The leading point was noted to be a mass causing tethering (blue arrow) of the bowel wall.

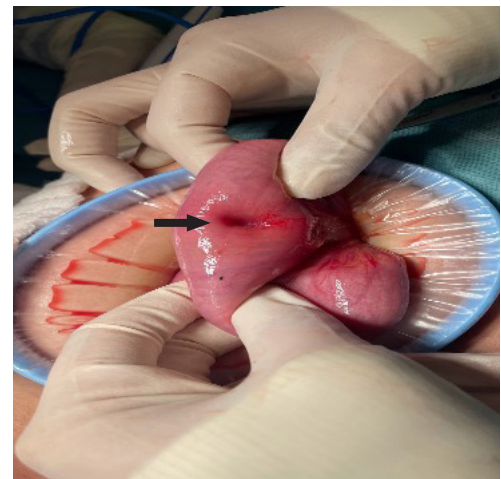


Figure 3. Gross small bowel specimen was opened and an intramural mass was noticed (arrow)

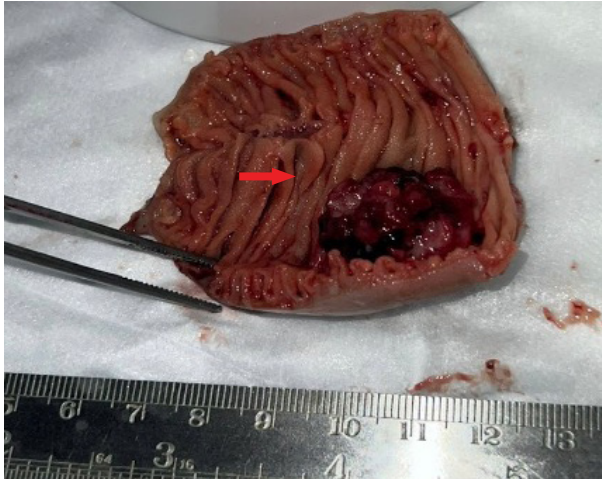


Figure 4. Microscopic images of the polypoid lesion within the bowel lumen with intact wall and serosa. B: Microscopic images of the osteoid material and atypical mitotic figures.

Figure 4a

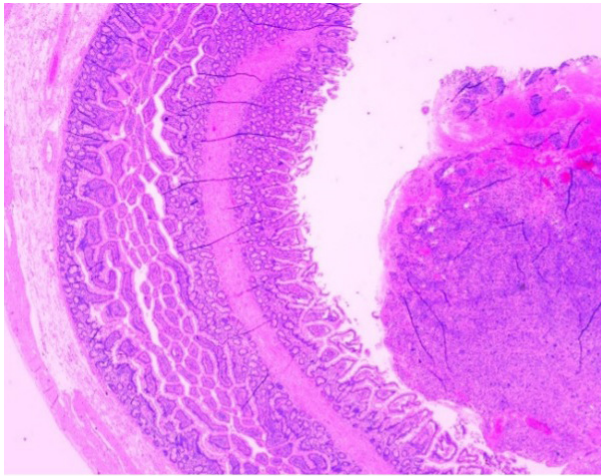
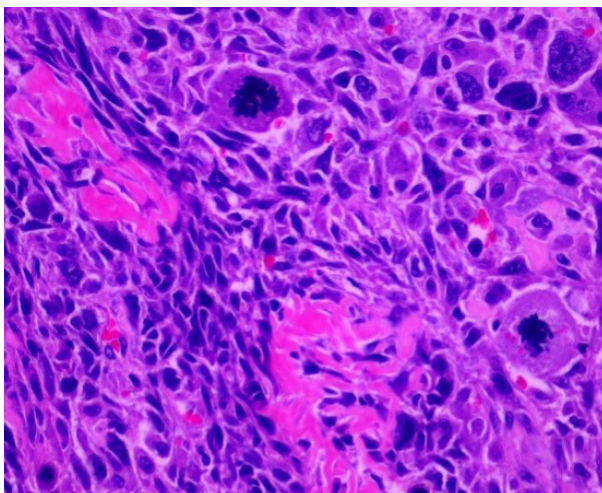


Figure 4b



DISCUSSION

OS most commonly metastasizes to the lung. Other sites include bone, pleura, and heart. Metastatic osteosarcoma to the small bowel is a rare entity, yet there is an increasing number of cases being reported in the literature (1-9). Metastasis to the gastrointestinal tract could represent an alteration in the natural history of OS due to adjuvant chemotherapy and increased long-term survival (10). Serpico et al. reviewed cases of metastatic osteosarcoma to the abdomen from 1963-2017, they identified 42 patients. 10 patients presented with intussusception. To the best of our knowledge, 12 patients reported in the literature with OS metastasis to the small bowel causing intussusception (1-9). Patients' ages range from 8 to 67 years old (2,5) at the time of osteosarcoma diagnosis and from 11 to 67 years old at the time of small bowel metastasis with intussusception. One patient experienced a small bowel intussusception within five days of osteosarcoma diagnosis, which can be considered metastasis at presentation (5). The remaining patients had small bowel intussusception presentation occurring 1.5-15 years after the initial diagnosis (6). Most reported cases involved metastasis to other sites, indicating a poor prognosis. Patients with OS metastasis to the abdomen have a 5-year survival of 10.9% (1). The time from the diagnosis of abdominal metastasis to death is 7.4 months on average (1). Only three cases of metastatic osteosarcoma causing intussusception in pediatric patients have been reported to date (2,3,4). (Table 1). All three pediatric patients had another metastasis as well. Our reported cases developed multiple metastases after the reported event and passed away 5 months after the GIT metastasis, which follows the reported pattern of aggressive disease with poor prognosis reported in the literature.

Intussusception is more common in pediatric patients than adults. In pediatric patients, most intussusception cases are idiopathic, with only 10% having a leading point, compared to 90% of adult patients with intussusception (7). Small bowel intussusception is usually interpreted as a benign and self-limiting event and represent normal small bowel motion. Patient with signs of obstruction, ascites, or intussusception length greater than 3.5 cm are more likely to need surgical intervention (11). In our reported case, the fact that this patient was under osteosarcoma surveillance raised suspicions of a small bowel malignancy as a leading point. Furthermore, since the intussusception was more than 3.5 cm and did not resolve spontaneously, surgical intervention was indicated.

Table 1. Osteosarcoma metastasis causing small bowel intussusception in pediatric patients

Author	Age at diagnosis of OS	Site of primary tumor	Presence of another metastasis	Age at metastasis.	Presentation	Outcome	Chemotherapy received
Hung, Giun-Yi 2000 ⁽²⁾	8 years	Femur	Lung, brain	13 years	Abdominal pain	Had postop shock and passed away.	Neoadjuvant chemo and radiotherapy + Adjuvant Chemotherapy: methotrexate, cisplatin, ifosfamide, epirubicin.
Chandramohan, K. 2003 ⁽³⁾	9 years	Femur	Scapula	11 years	Abdominal pain, vomiting	Developed multiple metastasis a month later and started on palliative treatment.	3 courses of Cisplatin; Adriamycin, Ifosfamide, Mesna.
Abbo, Olivier 2013 ⁽⁴⁾	12 years	Femur	Lung	17 years	Abdominal pain, RLQ tenderness Recurred with obstruction	Not reported.	Induction Doxorubicine, methotrexate, and cisplatin (AOST .331 protocol). Ifosfamide and VP16 and cis-retinoic acid post lung metastasis
Current case	12 years	Shoulder	Lung	13 years		Developed multiple lung, liver and bone metastasis and Converted to palliative treatment	Chemotherapy per MAP protocol

Intussusception can present with intermittent abdominal pain with or without vomiting or obstruction (Table1). On pathology examination, our patient had a hemorrhagic lesion, which could have contributed to the drop in hemoglobin, although this is hard to prove given no melena was reported, and no hemocult test was done.

Aboo et al. reported a case of a patient with a known history of osteosarcoma who presented with small bowel metastasis causing intussusception, which spontaneously reduced at the time of laparoscopic intervention. However, as intussusception recurred, laparotomy was attempted, and a mass was identified as the leading point. Subsequently, a small bowel resection and anastomosis were performed (4). This raises a fundamental question: if a patient with a previous history of malignancy presents with small bowel intussusception that was managed by reduction, should surgical intervention with bowel resection be attempted, or should clinicians at least investigate for metastasis using PET scans or CT scans? This remains a matter of controversy. The author argues that, in this subgroup of patients with a previous history of malignancy with or without prior metastasis, the presence of a pathological leading point as a cause of intussusception should justify early surgical intervention.

CONCLUSION

Small bowel intussusception in pediatric patients is mostly idiopathic. Patients with osteosarcoma presenting with signs of intestinal obstruction are a special subgroup where metastasis to the small bowel should be considered as a possible leading point and investigated properly. Surgical intervention is needed to relieve the obstruction. However, OS metastasis to the small bowel carries a poor prognosis.

REFERENCES

- 1 Serpico, Robert, Jeffrey Brown, Alan Blank, Kevin Jones, R. Lor Randall, and John Groundland. "Metastasis of Osteosarcoma to the Abdomen: A Report of Two Cases and a Review of the Literature." *Case reports in oncology* 14, no. 1 (2021): 647-658.
 - 2 Hung, Giun-Yi, Tzeon-Jye Chiou, Yuh-Lin Hsieh, Muh-Hwa Yang, and Winby York-Kwan Chen. "Intestinal metastasis causing intussusception in a patient treated for osteosarcoma with history of multiple metastases: a case report." *Japanese journal of clinical oncology* 31, no. 4 (2001): 165-167.
 - 3 Chandramohan, K., Thara Somanathan, P. Kusumakumary, P. G. Balagopal, and Manoj Pandey. "Metastatic osteosarcoma causing intussusception." *Journal of pediatric surgery* 38, no. 10 (2003): E1-E3.
 - 4 Abbo, Olivier, Kalitha Pinnagoda, Lionel A. Micol, Maya Beck-Popovic, and Jean-Marc Joseph. "Osteosarcoma metastasis causing ileo-ileal intussusception." *World journal of surgical oncology* 11, no. 1 (2013): 1-3.
 - 5 Kuwabara, Hiroko, Kazuhiko Fujita, Masako Yuki, Isao Goto, Toshiaki Hanafusa, and Yuro Shibayama. "Cytokeratin-positive rib osteosarcoma metastasizing to the small intestine." *Indian Journal of Pathology and Microbiology* 57, no. 1 (2014): 109.
 - 6 Herndon, Geoffrey G., Chirag R. Patel, Bart Rose, Shi Wei, Gene P. Siegal, and Sameer Al Diffalha. "Osteosarcoma with Telangiectatic Features Metastatic to the Small Bowel." *Human Pathology: Case Reports* 24 (2021): 200499.
 - 7 Yoo, Seong Ho, Min A. Kim, Kyu Joo Park, Joon Koo Han, Sang Hoon Lee, Chong Jai Kim, and Eui Keun Ham. "Intestinal Metastasis of Osteosarcoma Presenting with Intussusception: A Case Report." *Journal of Pathology and Translational Medicine* 36, no. 4 (2002): 271-273.
 - 8 Wootton-Gorges, Sandra L., Rebecca Stein-Wexler, and Danel C. West. "Metastatic osteosarcoma to the small bowel with resultant intussusception: a case report and review of the literature." *Pediatric radiology* 33 (2003): 890-892.
 - 9 Rejin, Kebudi, Özgüven A. Aykan, Görgün Ömer, Yekeler Ensar, Bilgiç Bilge, Ayan İnci, and Özger Harzem. "Intra-abdominal metastasis in osteosarcoma: survey and literature review." *Pediatric Hematology and Oncology* 28, no. 7 (2011): 609-615.
 - 10 Panizo-Santos, Angel, Iosu Sola, María Dolores Lozano, Enrique de Alava, and Javier Pardo. "Metastatic osteosarcoma presenting as a small-bowel polyp: a case report and review of the literature." *Archives of pathology & laboratory medicine* 124, no. 11 (2000): 1682-1684.
 - 11 Munden, Martha M., John F. Bruzzi, Brian D. Coley, and Reginald F. Munden. "Sonography of pediatric small-bowel intussusception: differentiating surgical from nonsurgical cases." *American Journal of Roentgenology* 188, no. 1 (2007): 275-279.
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