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Case Report

Extra-skeletal Ewing's Sarcoma of Caecum Causing Intussusception: First such Reported Case

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Abstract:

Introduction: Ewing's sarcoma is a tumor of primitive cells which primarily affects long bones. Extra-skeletal Ewing's sarcoma has been rarely reported in small bowel. Its presentation can be varied but it is an extremely unlikely cause of an Ileo-Colic Intussusception as seen in present case.

Case Presentation: A 32 years old male patient presented with symptoms suggestive of bowel obstruction. CT scan was suggestive of Ileo-colic intussusception. Patient underwent diagnostic laparoscopy and right hemicolectomy. Histopathology examination of specimen revealed Ewing sarcoma of colon.

Conclusion: Extra-skeletal Ewing's sarcoma is a rare entity. To the best of our knowledge this is the first reported case in English literature, of a Caecal Ewing's sarcoma causing Intussusception. Further study of such cases may help in better understanding and establishing treatment protocol for this condition.

Keywords: Extra-skeletal Ewing's Sarcoma; Caecal; Intussusception

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Introduction

Ewing's sarcoma is a small round cell tumor with sarcoma specific gene mutations [1].Ewing's sarcoma and PNET (Primitive Neuroectodermal Tumors) are now clubbed together [2] by pathologists due to their genetic similarities[3]. These are mainly located in extremities, Spine and chest wall [4,5]. Uncommon location of Ewing's Sarcoma like Esophagus [6], Stomach [7], Prostrate [8], Jejunum [9] have also been reported. Ewing's sarcoma of bowel causing intussusception is an extremely rare occurrence. To the best of our knowledge this is the first reported case in English literature, of a CaecalEwing's sarcoma causing Intussusception.

Case presentation

A 32 years old male patient presented with complaints of pain abdomen, more in periumbilical region since 1 month. He also had complaints of passing altered blood in stool since one week. On examination patient's vitals were stable. He had tenderness on right side of abdomen, however there was no palpable lump.

Ultrasonography revealed a well-defined hypoechoic lesion in right iliac region. A CT abdomen was done which suggested Ileo- Colic intussusception with likely polyp at leading edge (Fig 1).

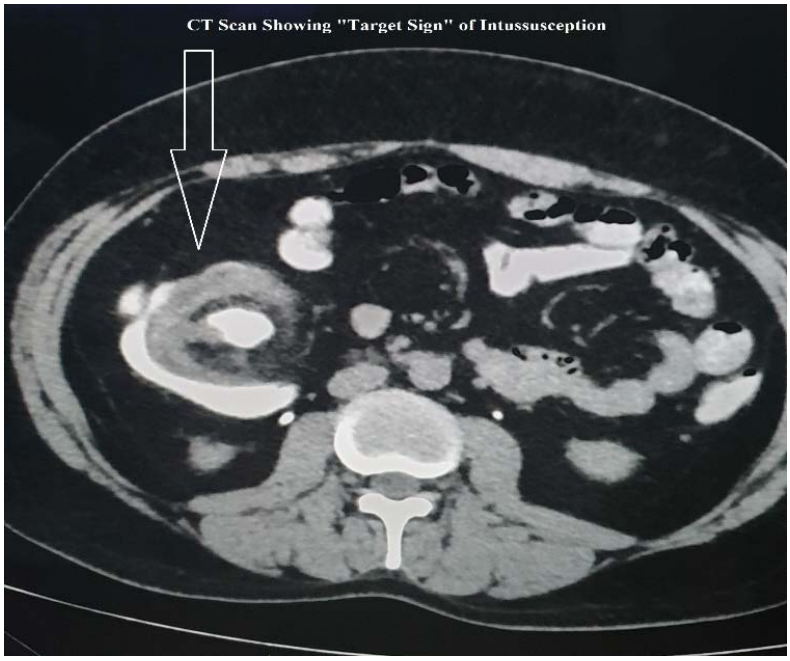

Fig. 1 CT Scan of abdomen showing "Target Sign" of Intussusception

A decision was taken to proceed with diagnostic laparoscopy. The intussusception segment at Ileo-Caecal region with mass was identified (Fig 2) and was brought out via open incision (Fig 3).

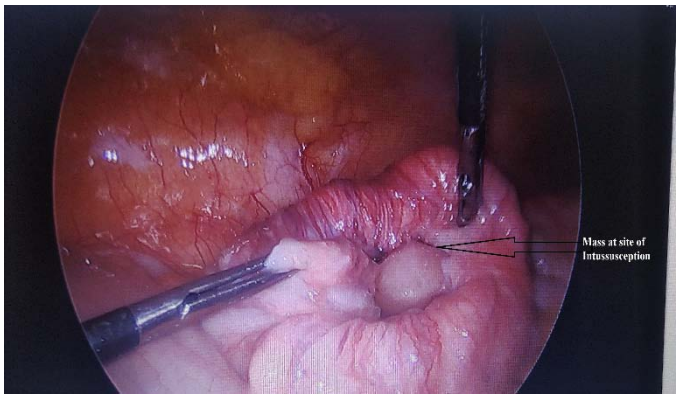

Fig. 2 Intra operative image showing Mass at the site of Intussusception

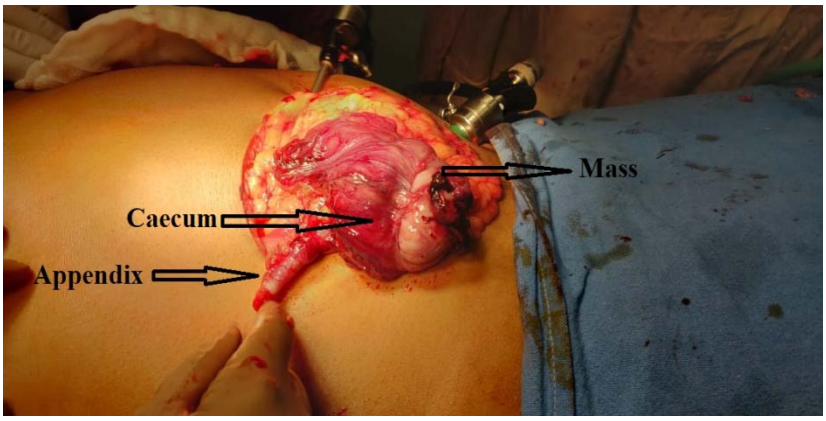

Fig.3 Caecum brought out

Right hemicolectomy with end to end anastomosis was done. Specimen revealed a mass in Caecum (Fig 4) and was sent for histopathological examination.

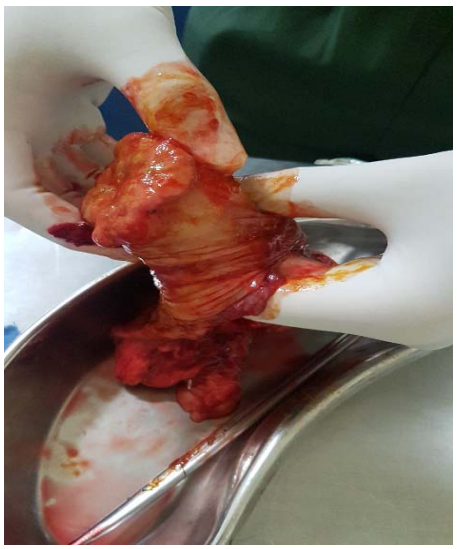

Fig. 4 Cut open specimen revealing the tumor mass

Histopathology reported a Caecaltumor comprising of sheets of round tumor cells with round to oval vesicular nuclei (Fig 5,6). Tumor involved all layers of Caecal wall and there was brisk mitosis. Proximal and distal margins were free and fourteen isolated lymph nodes were also free of tumor.Tumor cells were positive for CD99, FLI1, BCL 2 and Synaptophysin.

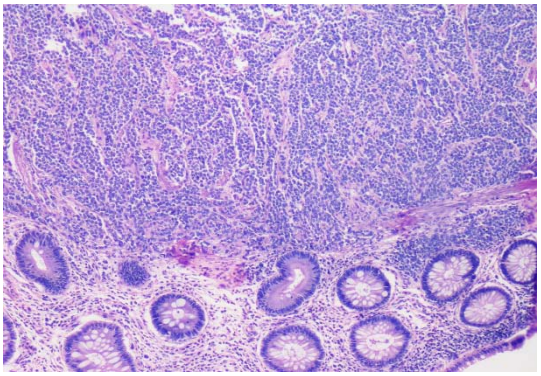

Fig. 5

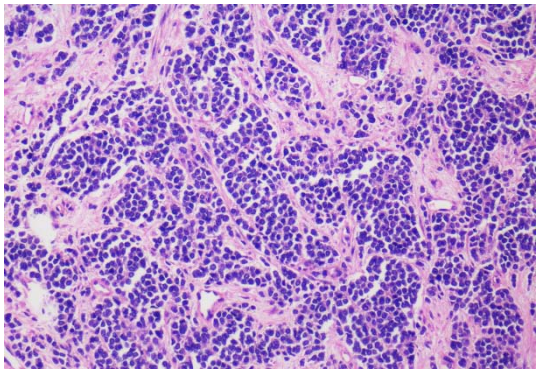

Fig. 6

Fig 5,6 Microscopic slides showing sheets of round tumor cells invading all layers of Caecum

A PET CT done also did not reveal any other primary site of tumor. Patient recovered well and was discharged on fifth post-operative day. Patient was advised chemotherapy but was lost to follow up.

Discussion

Ewing's Sarcoma was first described by James Ewing in 1921. He described them as Endothelioma of bone [10]. These tumors commonly affect skeletal system of younger patients. Rarely when they occur at extra-skeletal sites, the patients are usually older and their 5-year survival rate is also higher [11,12]. On genetic level multiple translocations are found on EWSR1 gene on chromosome 22, most common translocation being (t 11;22) with overexpression of FL1 protein. This is found in 85% of patients [13].

Ewing's of intestine is extremely rare. Yagnik et al in a report published this year documented 31 reported cases of Ewing's of small bowel [14]. Presentations ranged from abdominal lump, bowelobstruction, hematemesis and bowel perforation [14].

A Chinese report in 2017 documented 4 cases of Colo-Rectal Ewing's, none of them in Caecum and none causing intussuspection [15]. Of these four Colo-Rectal cases two had no metastasis and had disease free survival of 20 months and one year after surgery. One patient with peritoneal metastases expired seven months after surgery and chemotherapy and fourth patient with liver metastasis had disease free survival of 7 years after surgery and chemotherapy [15].

Many studies have shown that outcome of localised extra-skeletal Ewing's sarcoma is better than skeletal and metastatic disease, with five-year survival rate ranging up to 75% [16].

Though there are no universal guidelines of management of localised bowel Ewing'ssarcoma, complete excision with margin is advocated [15].

The role of radiotherapy is debatable,NCCN (National Comprehensive Cancer Network) guidelines recommend that any Ewing's Sarcoma be treated with surgery plus chemotherapy [17].

Conclusion

Extra-skeletalEwing's sarcoma being of extremely rare phenomenon doesn't have complete understanding and standardised treatment protocol. This is the first reported case of Caecal Ewing's sarcoma causing intussusception. More such cases need to be reported and studied to help in framing guidelines for their investigation and treatment.

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