**Name of Journal:** *World Journal of Gastrointestinal Surgery*

**Manuscript NO:** 58730

**Manuscript Type:** CASE REPORT

**Ewing sarcoma of the jejunum: A case report and literature review**

Shadhu K *et al*. ES of jejunum

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**Received:** August 4, 2020

**Revised:** September 30, 2020

**Accepted:** April 29, 2021

**Published online:** May 27, 2021

**Abstract**

BACKGROUND

Ewing sarcomas (ESs) are highly aggressive malignancy and are predominant in the long bones of extremities of children and young adults with a slight male predilection and rarely presents at extra skeletal locations.

CASE SUMMARY

A 55-year-old woman came to our hospital after finding elevated tumor biomarkers during her physical examination. Her enhanced computed tomography scan showed a jejunal mass. The patient underwent laparoscopic enterectomy. The mass was later diagnosed as ES, evidenced by fluorescence *in situ* hybridization whereby the GLP ES breakpoint region 1 probe was used, showing that more than 10% of the cells showed a red-green-yellow signal proving the breakpoint rearrangement of the ES breakpoint region 1 gene in chromosome 22.

CONCLUSION

We describe a case of localized ES at the jejunum in China based on the literature.

**Key Words:** Ewing sarcoma; Small bowel; Fluorescence *in situ* hybridization; Ewing sarcoma breakpoint region 1 gene; Jejunum; Enterectomy; Case report

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**Citation:** Shadhu K, Ramlagun-Mungur D, Ping XC. Ewing sarcoma of the jejunum: A case report and literature review. *World J Gastrointest Surg* 2021; 13(5): 507-515

URL: https://www.wjgnet.com/1948-9366/full/v13/i5/507.htm

DOI: https://dx.doi.org/10.4240/wjgs.v13.i5.507

**Core Tip:** Ewing sarcomas (ESs) are a highly aggressive malignancy and are predominant in the long bones of extremities of children and young adults. We hereby present a case of extraosseous ES of the jejunum in a female patient. This case highlights the jejunum as a potential site of ES origin and shows that a surgical approach with adjuvant chemotherapy is beneficial.

**INTRODUCTION**

Ewing sarcoma (ES) is a small round-cell tumor with simple sarcoma-specific genetic alterations resulting in a TET/FET family member and E26 transformation-specific family member[1]. ESs are rare small round-cell tumors that arise predominantly in children and young adults with a slight male predilection[2-4]. ES most often arises in the mid-shaft or diaphysis of the long bones of the extremities with the spine making up 8% of the primary sites[5]. Extra osseous ES occurs in the soft tissue of the extremities, paravertebral region, and pelvic cavity[6] and has also been discovered in most organs including the pancreas, liver, adrenal gland, esophagus, and uterus[7-13]. Extra skeletal cases are rare, and these patients generally present at an older age and demonstrate a greater overall 5-year survival than skeletal ES tumors[14,15]. Reports of primary liver involvement have been noted, as well as gastrointestinal sites of origin including the stomach, small intestine, and colorectal[16-19]. Nevertheless, ES is extremely rare in the small bowel. Here, we report a case of primary ES in the jejunum with EWS rearrangement.

**CASE PRESENTATION**

***Chief complaints***

A 55-year-old otherwise healthy female patient came to our hospital after finding out that she had elevated tumor biomarkers during her annual physical examination.

***History of present illness***

She had no other complaints. Her sleep and appetite were normal. Her excretion and egestion were all normal.

***History of past illness***

The patient had a free past medical history.

***Personal and family history***

The patient grew up in her locality, denies any contact with contaminated water or radiation exposure, and denies smoking and alcohol consumption. She had a gestational history of 1-0-0-1. Her menstruation was 16 (5-6/28-30) 50.

***Physical examination***

On examination, the patient’s temperature was 37.0 °C, heart rate was 85 beats per min, respiratory rate was 16 breaths per min, and blood pressure was 110/65 mmHg. The Glasgow coma scale was 15/15 without any pathological signs. Her S1 S2 sounds were regular. Her chest was bilaterally clear; no rhonchi or crackles were heard. Abdominal examination revealed a soft and non-tender abdomen. No mass or distension was observed. Bowel sounds were active.

***Laboratory examinations***

Her carbohydrate antigen 153 (CA-153) level was 38.04 u/mL, CA-199 was 109.5 u/mL, and CA-125 47 was u/mL. The white blood cell count was low at 3.39 × 109/L (normal range 3.50-9.50 × 109/L) and lymphocytes were low at 0.79 × 109/L (normal range 1.10-3.20 × 109/L).

***Imaging examinations***

Her abdominal computed tomography scan showed a contrast-enhanced mass in the small intestine at the left lower quadrant of the abdomen *(*Figure 1*)*.

***Further diagnostic work-up***

The patient underwent minimally invasive exploratory laparotomy. During the exploration, the tumor was located in the distal jejunum. It was well-circumscribed and had a fleshy pink surface similar to that of a gastrointestinal stromal tumor. A segment of the jejunum was resected 5 cm away from the edges of the tumor on both sides and an anastomosis was made using mechanical staple. The patient recovered uneventfully after surgery.

***Pathological report***

Pathological examination showed that the tumor of 3.5 cm × 3.0 cm × 2.3 cm in size was malignant, as there was invasion of the entire wall of the intestine. The resected sample had negative margins (R0) (Figure 2). Immunohistochemical analysis showed CD117 (-), CD34 (-), DOG-1 (-), Ki67 (35%), CK-pan (partly +), CK-L (+), CD56 (-), Syn (+), Villin (-), CK7 (-), Cg A (-), CD99 (+ +), INI-1 (+), Desmin (-), Inhibin-α (-), ER (-), PR (-), Calretinin (-), WT-1 (-), SF (-), HMB45 (-), S-100 (-), and Melan A (-). Fluorescence *in situ* hybridization (FISH) for an ES breakpoint region 1 (EWSR1) gene rearrangement (22q11) was performed using GLP EWSR1 probe, showed that more than 10% of the cells had a red-green-yellow signal, demonstrating the breakpoint rearrangement of the EWSR1 gene in chromosome 22 (Figure 3).

***Post-operative course***

Post-operatively, bone X-rays were done to rule out any primary lesion from her skeletal system (Figure 4). The patient was discharged on post-operative day 8.

***Further work and follow-up***

She was referred to the oncology department for further treatment. The regimen included vincristine, adriamycin, cyclophosphamide, doxorubicin, and addition of ifosfamide and etoposide (VACD-IE), given every 2 wk for 12 cycles. It started 1 mo post-operatively. However, after four cycles, the patient stopped the adjuvant therapy due to a fear of side effects. To date, there has been no sign of relapse and the patient recently showed interest in continuing the adjuvant therapy.

**FINAL DIAGNOSIS**

Extraosseous ES at the jejunum.

**TREATMENT**

Minimally invasive exploratory laparotomy. Referred to the oncology department for further treatment. The regimen included VACD-IE, given every 2 wk for 12 cycles. It started 1 mo post-operatively.

**OUTCOME AND FOLLOW-UP**

After four cycles, the patient stopped the adjuvant therapy due to a fear of side effects. To date, there is no sign of relapse and the patient recently showed interest in continuing the adjuvant therapy.

**DISCUSSION**

ES harbors multiple balanced translocations, and fusions involving the EWSR1 gene on chromosome 22 exist. The most common translocation is t (11;22), EWSR1-FLI1 fusion (85% of cases), causing overexpression of the FLI-1 protein. The second most common translocation is t (21;22), EWSR1-ERG fusion (5%-10% of cases). Numerous other less common variant translocations exist. Lack of reverse transcription-polymerase chain reaction fusion transcripts for EWSR1-FLI1 and EWSR1-ERG does not exclude the possibility of ES because it does not rule out fusion transcripts that may be present below the limit of detection for the given assay (5%)[20]. It most commonly arises from bone but can develop in extra skeletal sites[21]. ES of the small intestine is extremely rare based on the literature[22-24].

Malignant GIST usually expresses CD117, Dog-1, and CD34, which were all negative in this case. Although both synovial sarcoma and ES/PNET could have genetic rearrangements, the regions of these translocations are quite different. In ES/PNET, Chr22 EWS-FLI or EWS-FEV translocations are commonly reported[25]. However, in synovial sarcoma, SYT-SSX translocation is frequently observed[26]. Clear-cell sarcoma could be ruled out by negative immunohistochemistry for HMB45, S-100, and Melan A. A previous study also indicated the necessity of distinguishing from an intraabdominal desmoplastic small round-cell tumor by histological and immunohistochemical characteristics when ES/PNET occurs in the abdominal cavity[27].

Among the 37 cases found, 3 were derived from the esophagus, 9 from the stomach, 5 were of colorectal origin and 20 arose from the small intestine. Twenty-two cases were in males and fifteen were in females. The age range was 9-years-old to 68-years-old. FISH break-apart EWSR1 was positive in 19 cases, negative in 1 case and was not conducted in 17 cases[9,18,22,23,28-51]. Our patient’s characteristics fell within these demographic data. Demographic research has shown that the frequency of EW is higher in United States Caucasian population than in China[52].

ES predominantly affects children and young adults with a peak incidence between 10 and 20 years of age. About 30% of cases occur in adults over the age of 20 and fewer than 5% occur in adults over the age of 40[53].

to date, the outcome of the 5-year survival rate of metastatic patients is usually poor (< 30%) compared to localized ES (65%-75%), despite the use of chemotherapy[54]. Several studies have indicated that localized extra skeletal ES has a more favorable outcome than skeletal tumors[55,56].

According to National Comprehensive Cancer Network guidelines, postoperative radiation therapy should begin within 60 d of surgery and is given concurrently with consolidation chemotherapy[57]. This explains why our patient was referred to oncology department shortly after surgery for further treatments.

Intergroup Ewing’s Sarcoma Study-I and Intergroup Ewing’s Sarcoma Study-II showed that radiation therapy and chemotherapy with VACD was superior to vincristine, adriamycin, cyclophosphamide (VAC)[58]. The 5-year relapse-free survival rate was 60% and 24 % for VACD and VAC, respectively (*P* < 0.001). The corresponding overall survival rate was 65% and 28% (*P* < 0.001). Womer *et al*[59] reported that VACD-IE given on every 2 wk schedule was found to be more effective and no increase in toxicity.

**CONCLUSION**

ES is a highly aggressive small round-cell tumor that arises in adults. We have described a patient with ES occurring in the jejunum. This case report helps solidify jejunum as a potential site for ES origin and surgical approach with adjuvant chemotherapy does prove beneficial. However, this is a single case study and conclusion be made only based on our experience.

**ACKNOWLEDGEMENTS**

We’d like to thank Dr. Ding ZY for the radiological images and Dr. Fang HS for processing the pathological images.

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**Footnotes**

**Informed consent statement:** Written informed consent was obtained from the participants for publication of this article and any accompanying tables/images. A copy of the written consent is available for review by the Editor of this journal.

**Conflict-of-interest statement:** The authors declare that they have no competing interests.

**CARE Checklist (2016) statement:** The authors have read the CARE Checklist (2016), and the manuscript was prepared and revised according to the CARE Checklist (2016).

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**Manuscript source:** Unsolicited manuscript

**Peer-review started:** August 4, 2020

**First decision:** September 17, 2020

**Article in press:** April 29, 2021

**Specialty type:** Surgery

**Country/Territory of origin:** China

**Peer-review report’s scientific quality classification**

Grade A (Excellent): 0

Grade B (Very good): 0

Grade C (Good): C

Grade D (Fair): 0

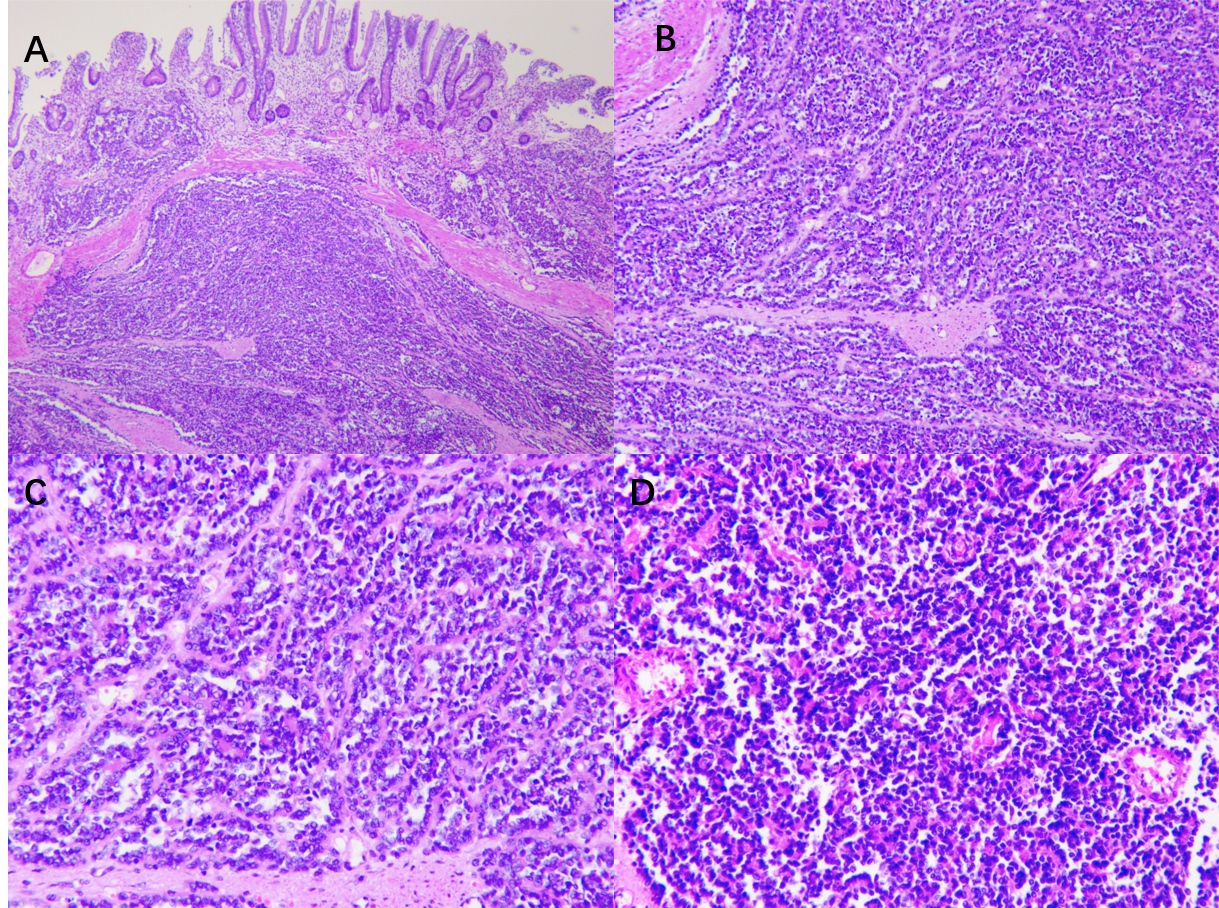
Grade E (Poor): 0

**P-Reviewer:** Altintoprak F **S-Editor:** Zhang L **L-Editor:** Filipodia **P-Editor:** Yuan YY

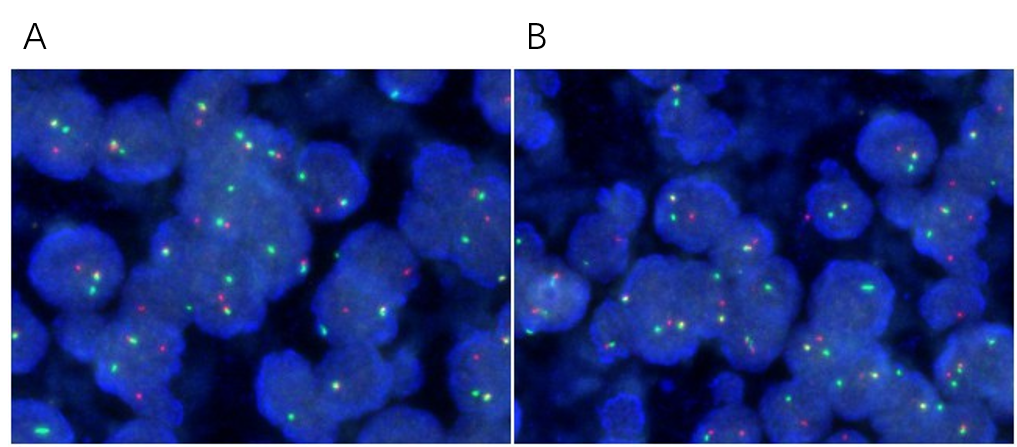
**Figure Legends**



**Figure 1 Transverse spiral computed tomography scan of the abdomen, with intravenous contrast enhancement showing dilation of jejunal wall of the left lower quadrant.**

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**Figure 2 Immunohistochemical analysis.** A: Low magnification of the resected sample using formalin-fixed (magnification: × 40); B: Paraffin-embedded sections of tumor stained with hematoxylin and eosin demonstrating sheets of small (magnification: × 100); C: Round-to-spindle, uniform tumor cells with clear cytoplasm (magnification: × 200); D: Higher magnification of C (magnification: × 200).



**Figure 3 Fluorescence *in situ* hybridization.** A: Fluorescence *in situ* hybridization of the resected tumor showing more than 10% of the cells showed a red-green-yellow signal, proving the breakpoint rearrangement of the Ewing Sarcoma breakpoint region 1 gene; B: More than 10% of the cells from resected sample showing a red-green-yellow signal (magnification: × 200).

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**Figure 4 Post-operative bone X-ray which shows no lesion in skeletal system thereby excluding metastasis.** A: Anterior-posterior view of the chest; B: Posterior-anterior right upper thigh; C: Posterior-anterior left upper thigh; D: Anterior-posterior right upper thigh; E: Anterior-posterior left upper thigh; F: Anterior-posterior lower leg; G: Anterior-posterior lower leg; H: Medial-lateral lower leg; I: Medial-lateral lower leg.



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