

CASE REPORT

Open Access



# Primary extraskeletal osteosarcoma of rectal mesentery: a rare case and literature review

Zhikui Huo<sup>1</sup>, Yao Sun<sup>1</sup>, Jinghui Chang<sup>1</sup>, Guo-Dong Li<sup>1</sup>, Jian Shi<sup>1</sup>, Cheng Quan<sup>1</sup>, Li-Na Zhang<sup>1</sup>, Ting-Ting Yang<sup>1</sup>, Feng-Jia Shang<sup>1</sup> and Yong-Ping Yang<sup>1\*</sup>

## Abstract

**Background** Extraskeletal osteosarcoma (ESOS) is a rare kind of sarcoma with a low preoperative diagnosis and a poor prognosis. ESOS arising from abdominal mesentery is extremely rare. Increasing diagnostic methods and standardizing treatment protocols are crucial issues of ESOS.

**Case presentation** We report the case of a 52-year-old female ESOS patient. She had a history of ovarian carcinoma (stage IIIC) surgery two years before, with five cycles of chemotherapy. A mass was found during postoperative examinations. A R0 surgical resection was performed. Post-operational pathological report plus intra-surgery findings supported a diagnosis of ESOS. She is still alive 10 months post-operationally, with routine blood and radiographical examinations.

**Conclusion** Enhancing awareness of this extremely rare disease together with advancements in diagnostic methods will hopefully enable earlier recognition and initiation of treatment. Protocols for standardizing treatments require a larger multi-center collaboration and more data analysis.

**Keywords** Extraskeletal osteosarcoma, Rectal mesentery, Surgical resection, Poor prognosis

## Introduction

Extraskeletal osteosarcoma (ESOS) is a rare kind of sarcoma that is considered a subtype of osteosarcoma, presenting a primary malignant mesenchymal tumor in which neoplastic cells produce osteoid substances in variable amounts [1]. It is first described by Wilson H. in 1941 [2]. Depending on some reports, the incidence of ESOS is around 1–2% of all soft tissue sarcomas and approximately 4–5% of all osteosarcomas [3–6]. Unsimilar to skeletal osteosarcoma, ESOS is another type of osteosarcoma, which is characterized by occurring in extraskeletal organs and soft tissues without attachment to the skeletal bones and periosteum [3, 7]. The locations

of ESOS have already been reported in several body system organs, such as the head, neck, heart, liver, gallbladder, kidney, and bladder; however, the most frequent reported site is the thigh [8–11]. This case we reported is an ESOS occurring in abdominal rectal mesorectum, an extremely rare site that has been reported only twice surrounding the rectum before, to our knowledge, following a literature review giving emphasis to abdominal mesotissue, such as small intestinal mesentery and large bowel mesentery [5, 12].

Due to a lack of clinical specificity and imaging features, ESOS makes it difficult to make a clear diagnosis pre-surgery. A definitive diagnosis conclusion is always made with a combination of clinical, radiographical, and pathological findings, especially immunohistochemical examination post-surgery. Compared with a large quantity of reports about abdominal carcinomas, there are few reports about ESOS in the abdomen. As a result, so far there has not come to make a common sense about

\*Correspondence:

Yong-Ping Yang  
yongpingyang8301@jlu.edu.cn

<sup>1</sup> The Department of General Surgery, The Second Hospital of Jilin University, Changchun 130041, China



treatment protocols for such abdominal ESOS, much less an overall survival data for this type of ESOS. Considering this situation, we reported this case of mesorectal ESOS, including its treatment timeline and re-imaging examination results post-surgery, with the hope of making further recognition of mesorectal ESOS generation, imaging features, treatment strategy discussion, and overall survival data supplement.

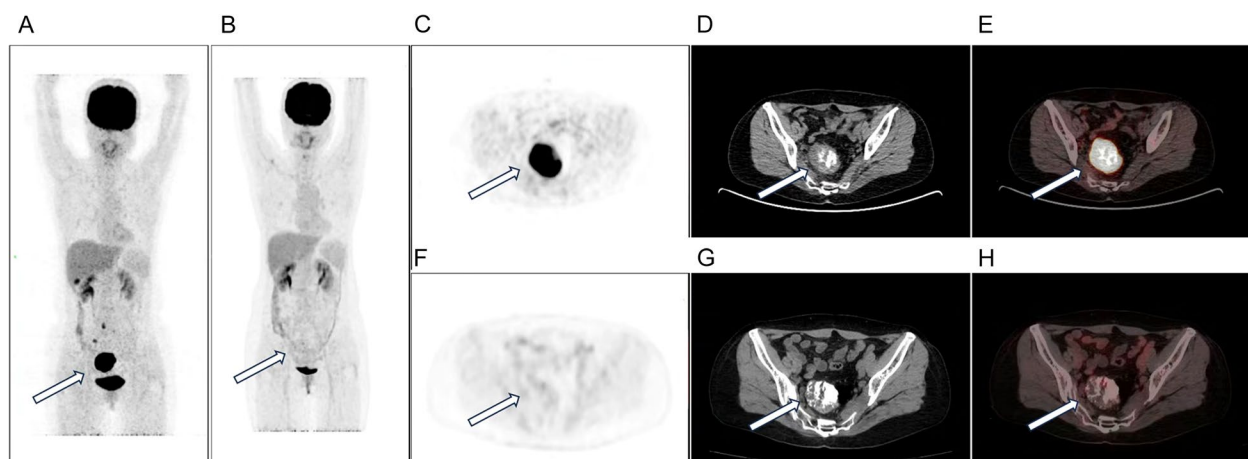
### Case presentation

A 52-year-old female patient was admitted to our hospital with a complaint of 4-week lower abdominal pain. She denied weight loss and pain or swelling in the extremities. No family history of malignancy was reported. On June 2, 2020 she had been diagnosed with ovarian carcinoma (stage IIIC), and a surgery of hysterectomy with bilateral adnexectomy, omentectomy pelvic, and lymph node dissection had been performed. The pathohistological examination result showed high-grade ovarian serous carcinoma and serous tubal intraepithelial carcinoma. Postoperative systemic chemotherapy of paclitaxel (175 mg/m<sup>2</sup>) and carboplatin (300 mg/m<sup>2</sup>) had been performed for 6 cycles. Routine post-operation examinations had been arranged. On February 7, 2022, an <sup>18</sup>F-FDG-PET-CT (Fig. 1) examination had revealed an occupying lesion in the pelvic cavity with high glucose metabolism that had been supposed to be a kind of malignancy. A hypermetabolic nodule between the inferior margin of the liver and ascending colon had been observed which had been suspected to be peritoneal metastasis. After a thorough discussion by gynecological doctors, this patient had undergone chemotherapy of paclitaxel (175 mg/m<sup>2</sup>) and carboplatin (300 mg/m<sup>2</sup>)

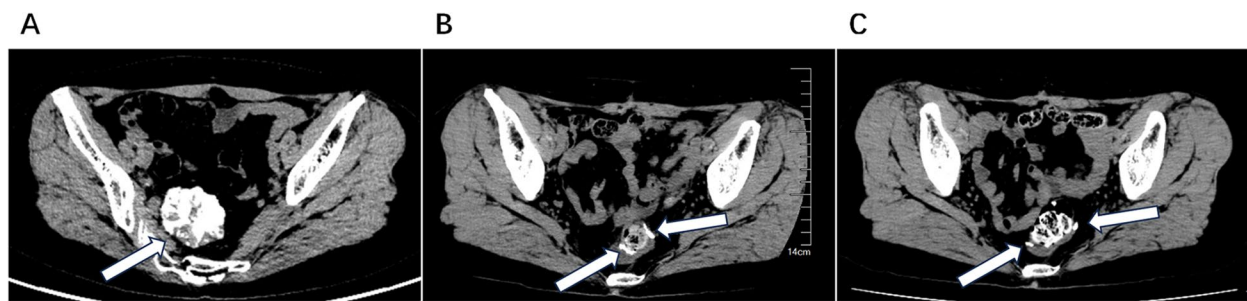
combining with bevacizumab (10 mg/kg), day 1–2 every 3 weeks, with a suspect of recurrence of the originated tumor. On May 18, 2022, a followed-up <sup>18</sup>F-FDG-PET-CT (Fig. 1) re-examination had revealed that the occupying lesion reduced slightly with a decreased glucose metabolic activity. She had finished chemotherapy + bevacizumab for 6 cycles until presented to our hospital.

Initial laboratory investigations showed no significant findings, with a normal level of the serum alkaline phosphatase: 74 U/L (50–135 U/L). A pelvic CT scan (Fig. 2A) revealed a mass measuring 5.0 cm×4.0 cm, which was inside the rectal mesentery. Cardiovascular and respiratory examinations were unremarkable. No morphological or immunohistochemical specimen was obtained preoperatively.

Management consisted of an integrated multidisciplinary treatment (MDT) consultation. Given the patient's ovarian disease history, the condition is considered ovarian in nature, lacking signs of gastrointestinal obstruction or altered bowel function. Consequently, a routine colonoscopy was omitted. The patient's family concurs with this decision. After communication with the patient, surgery consent was received. October 19, 2022, a surgery of R0 resection was achieved, with the discovery of a hard, rough, and off-white mass in the mesorectum, an invading part of the rectal wall, and poor movement due to surrounding adhesion. The mass was 6 cm×5 cm×4 cm in size. Also, a mass gray-white in color was seen in the ascending mesocolon. The size of another mass was 1.0 cm×1.5 cm×0.3 cm. A resection of pelvic mass and partial proctectomy were performed; meanwhile, an ascending mesocolon mass local resection was also performed. In order to maximize radical oncology, the



**Fig. 1** <sup>18</sup>F-FDG-PET-CT 5 and 8 months pre-surgery. (A, C–E) PET-CT 8 months pre-surgery, confirmed abnormal uptake inside the calcified mass. (ESOS with arrows) (B, F–H) PET-CT 5 months pre-surgery, after four cycles of chemotherapy and bevacizumab finished. The mass has shrunk significantly. (ESOS with arrows)



**Fig. 2** CT scan pre-surgery and 4 and 9 months post-surgery. (A) A homogeneous soft-tissue mass with internal calcifications or ossifications in rectal mesentery (with a white arrow) (B) A CT scan 4 months post-surgery showed no local recurrence (bowel lumen anastomosis site with white arrows) (C) A CT scan 9 months post-surgery showed no local recurrence (bowel lumen anastomosis site with white arrows)

technique of total mesorectal excision was used intraoperatively. A second nodule resected from the ascending colonic mesentery was not detected on preoperative evaluation. It was found for intraoperative exploration and postoperative pathology, diagnosed as fibrotic, vitelliform nodules.

After a differential diagnosis excluding heterotopic mesenteric ossification and gastrointestinal stromal tumor, the morphological and immunohistochemical findings were consistent with a diagnosis of osteosarcoma. By immunohistochemical staining, the neoplastic cells were positive for vimentin, SATB2, CD56, and P16 but negative for CK(AE1/AE3), WT1, P53, PAX-8, ER, PR, CK7, CK20, and CK8/18. Ki-67 positive rate was about 40% (Fig. 3). The mass in the mesorectum was demonstrated as focal ossification nodules. February 20 and February 28, 2023, and June 19 and July 21, 2023, CT scans and colonoscopy examinations were arranged (Fig. 4). No recurrence or distant metastasis findings were confirmed. (Fig. 2B,C. Fig. 5) April 10, 2024: PET-CT identified probable multiple hepatic and peritoneal metastases involving the mesentery and descending parietal colon (Fig. 5A,B). On April 17, 2024, a liver puncture was performed, which did not show any cancer cells. On July 30, 2024, pathology results showed carcinoma with poor differentiation. Immunohistochemical staining results: 1. CK (AE1/AE3) (+), ER (small +), PR (-), PAX-8 (-), P53 (-), WT1 (-), P16 (+), TTF-1 (-), NapsinA (-), CK7 (+), Ki67 (85% positive), GATA (partially +), CK20 (-), Hepatocytes (-), SATB2 (-). Pathology consultation was requested. The pathologist considers it likely that the cancer originated from the ovary.

Patient underwent a liver-occupying puncture on April 17, 2024, and the pathology did not show any cancer cells. Three cycles of chemotherapy with the liposomal adriamycin + carboplatin regimen were given on April 28, 2024, May 28, 2024, and June 26, 2024. Follow-up abdominal CT and PET-CT on July 23, 2024, etc.

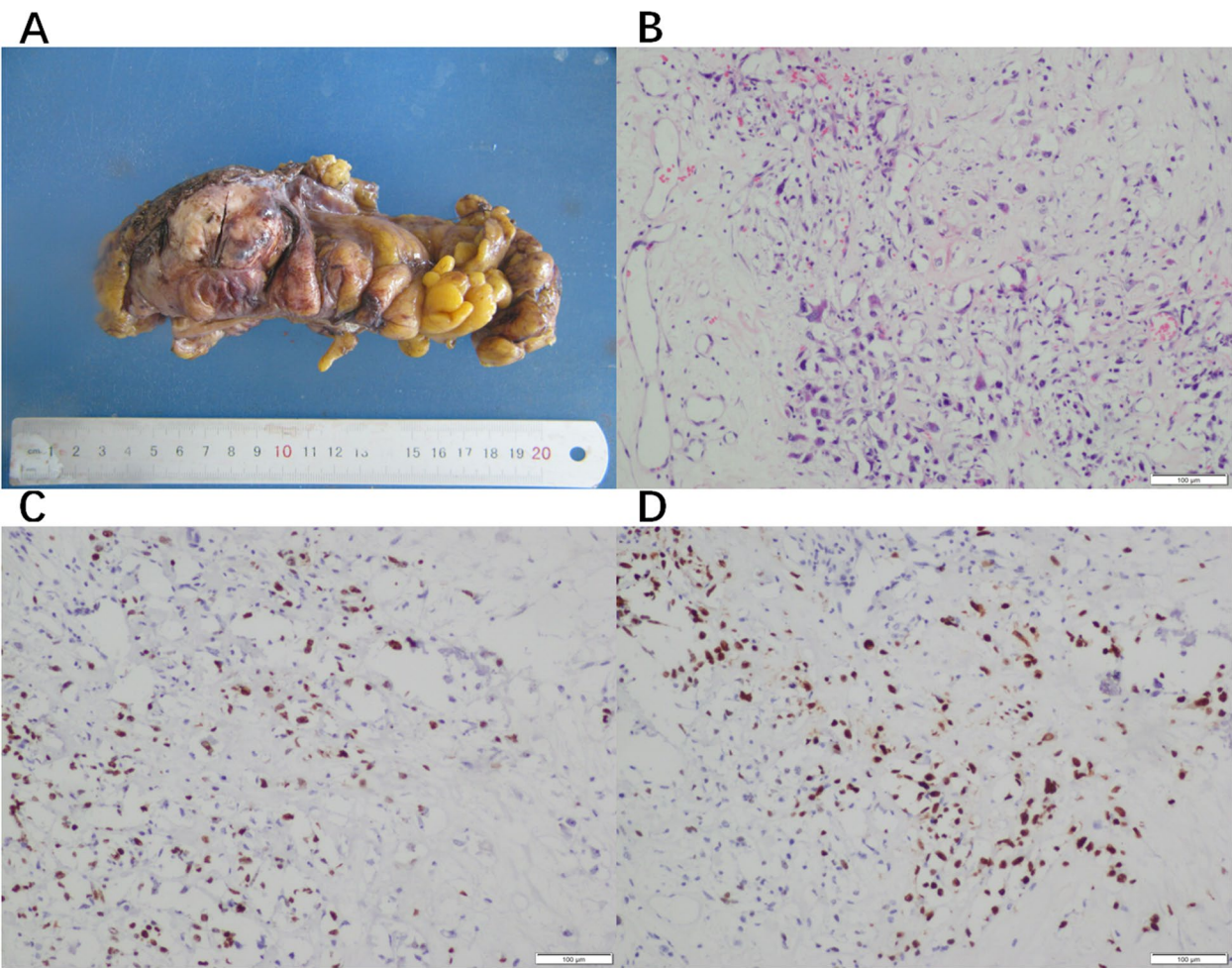
suggested disease progression. The patient underwent puncture of hepatic occupancy on July 30, 2024, with pathology returning carcinoma, poorly differentiated. Immunohistochemical staining results: 1. CK (AE1/AE3) (+), ER (little +), PR (-), PAX-8 (-), P53 (-), WT1 (-), P16 (+), TTF-1 (-), NapsinA (-), CK7 (+), Ki67 (85% positivity), GATA (partial +), CK20 (-), Hepatocyte (-), SATB2 (-). On August 4, 2024, chemotherapy with a gemcitabine + oxaliplatin + amlotinib regimen was administered; after chemotherapy, grade 4 leukopenia and grade 4 thrombocytopenia occurred, and symptomatic treatments such as elevation of leukocytes were given. On September 3, 2024, the dosage of the second cycle of the gemcitabine + oxaliplatin + amlotinib regimen was reduced, and chemotherapy with the gemcitabine + oxaliplatin + amlotinib regimen was administered on September 26, 2024, and October 22, 2024, and on October 22, 2024, chemotherapy with the gemcitabine + oxaliplatin + amlotinib regimen was administered. Gemcitabine + Oxaliplatin + Amlotinib regimen.

## Discussion

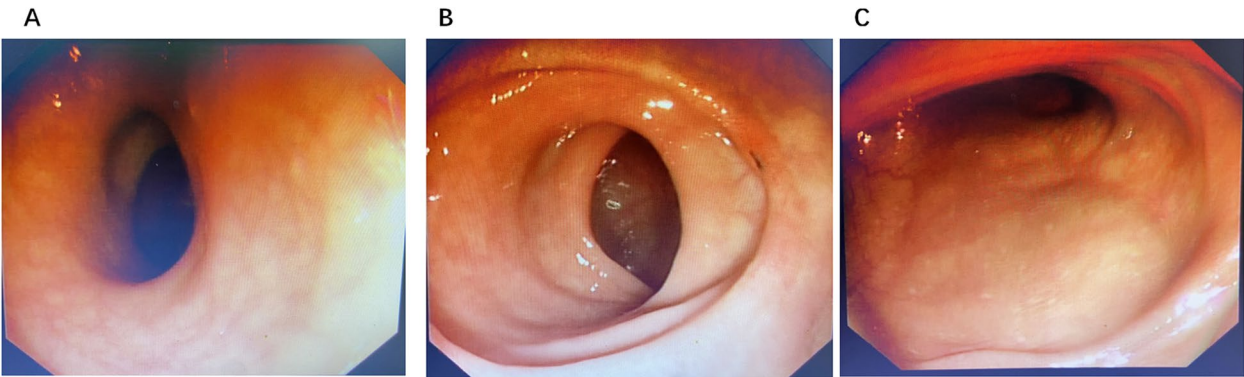
ESOS, as a malignant neoplasm, was first reported in 1941; however, few cases have been reported in contrast to skeletal osteosarcoma so far. ESOS arising from abdominal mesentery are extremely rare. To the best of our knowledge, there have been only 14 cases reported so far [13–26] (Table 1). Among them, the majority of cases arose from small bowel mesentery (10/14), 2 cases arose from sigmoid colon mesentery; besides, and only 1 case was report arising from transverse colon mesentery, besides 1 case did not clearly reported mass sites. This case we are presenting with is an ESOS arising from rectal mesentery; seemingly there has been no case reported before.

The case is extremely rare, and there is no common-sense guideline. And because the preoperative diagnosis was more difficult, there were other primary malignant

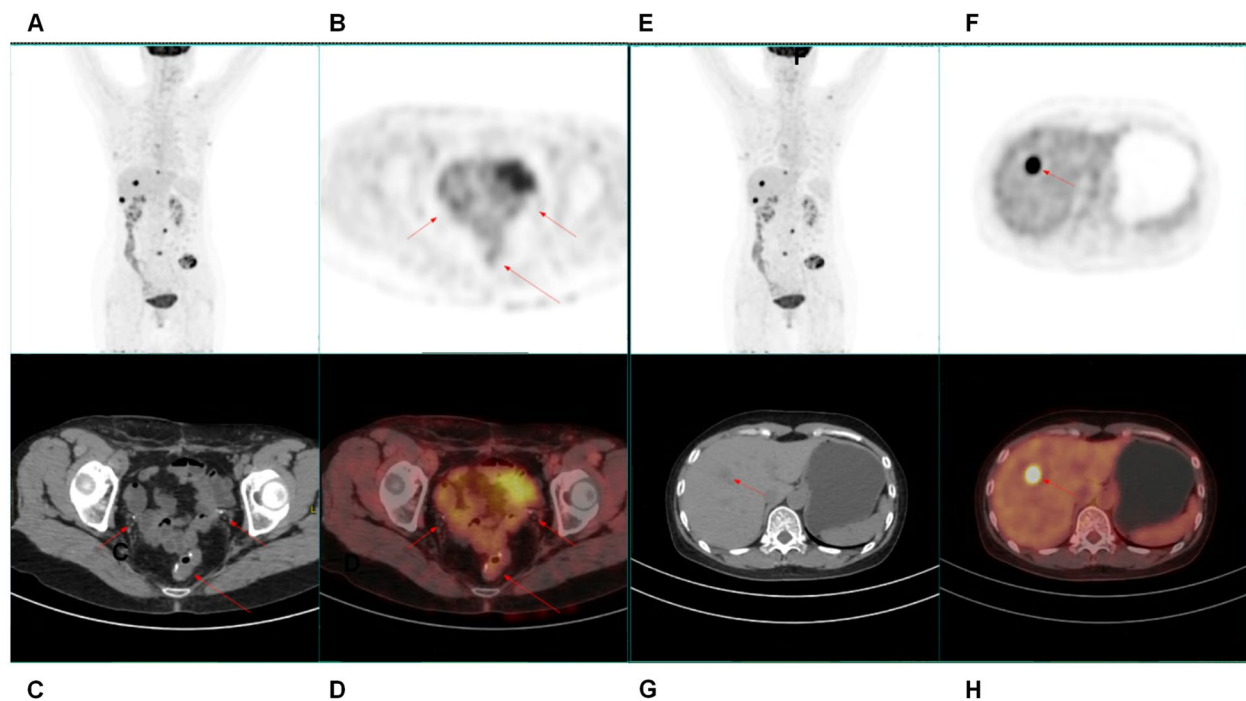




**Fig. 3** Morphological and immunohistochemical results post-surgery. **(A)** 6cm × 5cm × 4 cm mass located in the mesorectum, invading part of the rectal wall. **(B)** The tumor cells demonstrated atypical mitotic figures. (haematoxylin and eosin stain, original magnification × 200) **(C)** Immunohistochemical staining revealed the tumor cells were approximately 40% positive for Ki67 (original magnification × 200). **(D)** Immunohistochemical staining revealed the tumor cells were positive for SATB2 (original magnification × 200)



**Fig. 4** Colonoscopy 9 months post-surgery. **(A-C)** No recurrence evidence found



**Fig. 5** PET-CT was performed 13 months after surgery. (A-D) Increased metabolic activity within the liver and adjacent descending colon nodes suggestive of malignant metastases. (Metastatic lesion with arrows)

tumors in this case, which was a challenging point in this case.

Extraskeletal osteosarcomas (ESOS) is a kind of very rare disease. According to a literature report, the incidence of ESOS is around 1–2% of all soft tissue sarcoma [27]. Allan et al. reported the incidence of ESOS in soft tissue sarcoma is 1.94% (26/2100), while Lorentzon et al. reported that is 1.65% (4/242) [3, 4]. As reported by some groups, the age of patients with skeletal osteosarcoma is always below 30 years old; in contrast, ESOS is primarily with an older mean age of 47.5 to 61 [6, 7, 27–32]. Meanwhile, Nathalie E. J. van den Broek et al. argued that ESOS typically affected patients with an age between 50 and 70 [17]. According to a clinical features analysis by Sheila Thampi et al., the mean age for patients with ESOS is 60.7 years (range from 9 to 96; median age is 64 years) compared to 31.4 years (range from 0 to 99; median age is 20 years) for those with skeletal osteosarcoma [28]. Summarizing all above reports, regardless of the statistic details, it is a definite conclusion that patients with ESOS significantly tend to have an older age in contrast to patients with skeletal osteosarcoma. This age distribution characteristic between ESOS and skeletal osteosarcoma is also proved by the SEER database [28]. Depending on the data supported by Table 1, the mean age for patients with abdominal mesentery ESOS is 60.14 (range from 40

to 75; median age is 62 years). The patient in our case is 52-year-old female.

Gender distribution varies of ESOS in each cohort study have not a consensus. The reason for that perhaps is the low incidence of ESOS and cohort data insufficiency. Female predominance for ESOS was observed by Sheila Thampi et al., as did Choi et al. [7, 28]. However, the majority of case series described a male predominance [6, 11, 27, 29]. In some groups analysis, a ratio of males with ESOS versus females with that is 1.9:1 [17, 30]. From Table 1, we observed that, among abdominal mesentery ESOS, males are 8 cases (8/14), females are 6 cases (6/14).

According to previous literature reporting, distant metastatic disease, larger tumor size, TNM stage >2, axial tumor site, positive margins, and older age are supposed to be adverse prognostic factors [6, 7, 27, 29]. Among these prognostic factors, distant metastatic disease at diagnosis can be confirmed to have an unfavorable prognostic impact by some studies [6, 28]. Distant metastases are the most common cause of death, which is in agreement with current literature, with lung being the most common site, followed by regional lymph nodes and liver [27, 30, 33–35]. Soft tissue, skin, and peritoneal metastasis have also been reported [17, 31]. Covello et al. supported the theory that skin metastasis could be a sign of widespread disease [35]. Larger tumor size

**Table 1** Literature review of EOSOs of the mesentery cases

Case	Gender	Age	Site	Size(cm)	Pathological grade	Regional lymph node involved	Resection margin status	Local recurrence and/or distant metastasis	Treatment strategy	Prognosis
Gerald Fine et al. [19]	M	53	Mesentery. Tumor adherent to peritoneum and bowel	NA	NA	Positive	NA	NA	surgery	Dead 55 days postoperatively
Parvez H.Shirazi et al. [26]	F	56	Small bowel mesentery	NA	NA	Positive	R1	Liver	surgery	Dead
H. N. Choudur et al. [14]	M	45	Small bowel mesentery	15 in length	High-grade	Negative	R0	Negative	surgery + chemotherapy(six cycles of doxorubicin and cisplatin.)	Alive
L.C. Heukamp et al. [13]	M	61	Small bowel mesentery	20×10×10	High-grade	NA	R1	Negative	surgery + chemotherapy (The 1st cycle contained doxorubicin and cisplatin. The 2nd cycle contained cyclophosphamid and cisplatin. From the 3rd cycle onwards contained ifosfamid and doxorubicin.) + surgery	Dead 10 months 2 weeks initial postoperatively
Kyung Hwa Lee et al. [15]	M	67	Transvers colon mesentery	18.5×13×9.5	NA	NA	R1 suspected	Peritoneum, lung and liver 3 months post-operatively	surgery + chemotherapy(the 1st cycle of chemotherapy contained ifosfamide and adriamycin.)	Dead 4 months postoperatively
Muhammad I. Hussain et al. [16]	M	40	Small bowel mesentery	15×10x8	NA	NA	R1 suspected	Liver preoperatively	surgery + chemotherapy	Alive still 14 months post-operatively
Xiang Salim et al. [24]	M	63	Small bowel mesentery	15×9×9	NA	NA	R0	Mesenteric and pelvic mass 1 months post-operatively	surgery + chemotherapy (adriamycin)	Dead 18 months postoperatively
Sun-Ju Oh et al. [20]	F	70	Small bowel mesentery	15.1×10.3×7.5	NA	NA	NA	Negative	surgery	Dead 2 months postoperatively
Nathalie E.J. van den Broek et al. [17]	F	71	Sigmoid colon mesentery	14×9×9	NA	NA	R0	Greater omentum, sigmoid colon preoperatively, abdominal wall, wide-spread intra-abdominal lesion 5 months postoperatively	surgery	Alive still 5 months postoperatively
Alexandros Diamantis et al. [22]	M	73	Small intestine mesentery	22×12×10	NA	Positive suspected	R0	Negative	surgery + chemotherapy(Adriamycin and Ifosfamide)	Alive still 3 years postoperatively

Table 1 (continued)

Case	Gender	Age	Site	Size(cm)	Pathological grade	Regional lymph node involved	Resection margin status	Local recurrence and/or distant metastasis	Treatment strategy	Prognosis
Shingo Ito et al. [18]	F	46	Small intestine mesentery	3.8×2.5×1.3	NA	NA	R0	NA	surgery	Alive still 10 months post-operatively
Xinyang Nie et al. [23]	F	75	Sigmoid colon mesentery	11.5×7×6.5	NA	Negative	R0	Descending colon and the anastomosis 9 months postoperatively	surgery	Alive still 9 months postoperatively
Gabriel Yihan Tong et al. [21]	F	66	Small intestine mesentery	5 in length	High-grade	NA	R0	NA	surgery	NA
S. A. Ahmed et al. [25]	M	56	Small intestine mesentery	NA	NA	NA	NA	NA	surgery	NA



is another adverse prognostic factor, which is supported by present and previous literature [23, 28, 29, 36]. It can be a definitely concluded observed by many groups that overall survival rate falls down as tumor size increases. Bane BL et al. observed all patients (6/7 patients, 85.7%) alive with no evidence of disease had tumor measuring less than 5 cm that were amenable to perform a complete surgery excision with margin negative. Whereas the overall survival rate fell down to 12.5% (2/16 patients) for tumor size greater than or equal to 5 cm [27]. A larger SEER database analysis was also able to confirm this former adverse prognostic impact of larger tumor size, with a matrix distribution of 10 cm, by using Kaplan–Meier (KM) methods with 95% confidence intervals and log-rank tests and the Fine-Gray proportional subhazard model [28]. Ahmad et al. observed that there was a significant survival decrease for tumor size > 10 cm through an univariate analysis; however, no such decrease remained in multivariate analysis [29]. This result anticipates that tumor size or volume is one of the main adverse prognostic factors but not the only one. Some other adverse factors may also occupy their positions to a certain extent.

ESOS is categorized into three main pathological subtypes depending on different matrix components: osteoblastic, chondroblastic, and fibroblastic [35]. Telangiectasis, small cell, and mixed type are another three pathological subtypes [27, 37]. One of the most common of these is the osteoblastic variant, and the typing in our individual case was of the same type. There have been several inconsistent cohort studies on the prognostic significance of the different pathologic subtypes. Chung EB et al. observed patients with ESOS with fibroblastic components fared better than the rest [11]. Whereas Lee JY et al. observed patients with the subtype of chondroblastic survived longer than those with osteoblastic subtype by an univariate analysis [30]. However, some clinical studies did not find the prognostic difference among these three main subtypes [27, 33]. Therefore, it seems that to understand the ESOS prognostic factors will likely require a greater understanding of the cell of origin of ESOS. In the presented case, the patient was a patient suffering from gynecologic cancer with postoperative recurrence. Predicting her prognosis remains a challenging task.

Given the rare incidence of ESOS, varied clinical characters, and differing treatment approaches among groups, there is little evidence regarding standardized protocols for ESOS. Based on data from the literature, current treatment options are based on wide resection, followed by postoperative adjuvant radiotherapy and chemotherapy [38]. According to the study, expanding the surgical scope reduces the local recurrence rate but has no significant effect on prolonging survival time [30]. Although

multimodal therapy approaches were applied, including surgical resection, chemotherapy, and radiotherapy, previous literature reported dismal overall survival for patients with ESOS [7, 11, 27, 29–33]. According to the reports, 3-year overall survival of ESOS is approximately 61%, and 5-year overall survival is only 25% [7, 39]. However, complete surgical resection and negative surgical margin so far have been anticipated to be crucial effective treatments that have already performed comprehensively in carcinoma and osteosarcoma, though there is no adequate reliable data and a lack of multi-center cohort analysis trials on these two yet. Goldstein-Jackson et al. found that complete surgical resection was the only statistically significant prognostic factor for a better overall survival result in their univariate analysis [31]. Although a complete surgical remission is performed by most studies, local recurrences and even distant metastasis are still obstacles, which are the main unfavorable impacts on overall survival. As previous literature reported, the local recurrent rate is approximately 20%–69% [29–31, 33, 40]. In this individual case, a total mesorectal resection technique was used, and postoperative pathology suggested that no lesions were seen at the bilateral margins and that the liver occupying the line of perforation pathology suggested non-ESOS. To some extent, it suggests that the surgery has the potential to achieve radical resection. More data are still needed to confirm this.

Chemotherapy remains controversial about regimens and effects [23, 29, 31]. Ahmad et al. reported they received an unsatisfied effective rate by applying doxorubicin-based regimens, while cisplatin-based chemotherapy was not active either [29]. However, S. Y. Goldstein-Jackson et al. obtained more favorable results with a more aggressive multiagent chemotherapy strategy [31]. Therefore, they recommend that patients with ESOS be treated with polychemotherapy regimens including doxorubicin, ifosfamide, cisplatin, and possibly methotrexate and adequate surgery. Wang et al. observed that there was no significant survival benefit between the group of patients with methotrexate, adriamycin, and cisplatin-based chemotherapy regimens and the group of patients with adriamycin or ifosfamide chemotherapy regimens. Even no survival difference between those who received chemotherapy and those who did not receive was found in his study [37]. In our presenting case, a multi-regimen of chemotherapy and gene-targeting drug bevacizumab were given before a thorough surgery, and a  $^{18}\text{F}$ -FDG-PET-CT followed, which confirmed “occupying lesion reduced slightly with a decreased glucose metabolic activity.” It seemingly suggests that a proper multi-regimen may be effective for abdominal mesentery ESOS; furthermore, bevacizumab could be certified to be one of the useful medicine supplements for ESOS. The patient



underwent a postoperative chemotherapy regimen of liposomal camptothecin in combination with carboplatin for recurrence of ovarian malignancy. Perhaps these drugs may also have some effect on ESOS.

Radiotherapy is a predisposing factor for the development of bone and soft-tissue sarcomas [41]. Whereas palliative radiotherapy is considered a substitute approach for the patients with ESOS who have no opportunity to complete a surgical resection, who tolerate chemotherapy, or who are in advanced stage. Radiotherapy has been reported to be beneficial in reducing the volume of tumors and local recurrence but not beneficial in increasing the overall survival rate [7, 37].

This is a rare case and the type of pathology could not be clarified preoperatively to clarify the chemotherapy regimen. When the diagnosis is difficult, MDT does have helpful implications. However, in this case, the patient was fully communicated with preoperatively and agreed that the results of intraoperative exploration would determine the subsequent treatment. When the diagnosis is difficult, there should be a sense of suspicion of ESOS. In the case of preoperative diagnostic difficulties, further examination is needed, and if necessary, puncture biopsy is performed if the site is suitable. Particular attention is paid to those with a history of ESOS.

ESOS should also be considered when imaging demonstrates an intraperitoneal solid cystic or calcified mass [38]. The imaging features of ESOS lack distinctive characteristics. The diagnosis of ESOS should be made in the context of the clinical presentation as well as imaging and pathologic findings and only after the possibility of a primary bone tumor or metastasis of a bone tumor to the soft tissues has been ruled out. Combining clinical and imaging findings, it is necessary to differentiate it from liposarcoma, gastrointestinal mesenchymal tumor, or calcified hemangioma. In conclusion, this report illustrates that ESOS originating from the rectal mesentery should be considered in the differential diagnosis of intra-abdominal malignant mesenchymal tumors. The optimal treatment of mesenteric ESOS remains a topic that requires in-depth study.

## Conclusion

ESOS is an extremely rare and aggressive malignancy that is characterized by containing a component of osteoid substance that carries a poor prognosis. This presenting case may be the first case reported about ESOS in rectal mesentery. Although a complete surgical resection is largely accepted, the chemotherapy effectiveness is still controversial, with various groups opting for osteosarcoma or soft tissue sarcoma regimens. Radiotherapy is routinely absent or as a substitute for surgery.

Increased awareness of this rare disease with advanced examination will hopefully be able to make an earlier diagnosis. In the future, the feasibility of a multicenter and/or international collaboration is required to set a standard treatment protocol for ESOS.

## Abbreviations

ESOS Extraskelatal osteosarcoma  
MDT Multidisciplinary treatment

## Acknowledgements

The authors thank the patient's family for their cooperation. The authors thank those patients who contributed to the article but did not meet the criteria for authorship.

## Authors' contributions

Yong-Ping Yang and Zhikui Huo conceived of this study, designed it. Yong-Ping Yang, Zhikui Huo, Sun Yao, Jinghui Chang, Guo-dong Li, Jian Shi, Cheng Quan, Li-Na Zhang, Ting-Ting Yang and Feng-Jia Shang drafted the manuscript. Yong-Ping Yang, Zhikui Huo, Jian Shi, Min Wang, and Guo-dong Li participated in design of this study, acquired the data and analyzed the data. Yong-Ping Yang participated in manuscript preparation and critical revision. All the authors read and approved the manuscript. participated in design of this study, acquired the data and analyzed the data. Yong-Ping Yang and Yao Sun participated in manuscript preparation and critical revision. All the authors read and approved the manuscript.

## Funding

This study was funded by Jilin Province HealthScience and Technology Capacity Enhancement Project.

## Data availability

No datasets were generated or analysed during the current study.

## Declarations

### Ethics approval and consent to participate

The authors certify that they obtained all appropriate patient consent forms. They were assured that the patient's names and initials would not be published and due efforts would be made to conceal the identity of the patient, although anonymity could not be guaranteed.

### Consent for publication

We got the consent of the patient and family members before using the patient's data in this study.

### Competing interests

The authors declare no competing interests.

Received: 27 October 2024 Accepted: 19 January 2025

Published online: 29 January 2025

## References

- Aslan M, Samdanci ET. Very expensive neoplastic lesion mimicking a paraganglioma in the area of parapharyngeal: extraskelatal osteosarcoma. *Braz J Otorhinolaryngol*. 2022;88(2):279–82.
- Wilson H. Extraskelatal ossifying tumors. *Ann Surg*. 1941;113(1):95–112.
- Allan CJ, Soule EH. Osteogenic sarcoma of the somatic soft tissues. Clinicopathologic study of 26 cases and review of literature. *Cancer*. 1971;27(5):1121–33.
- Lorentzon R, Larsson SE, Boquist L. Extra-osseous osteosarcoma: a clinical and histopathological study of four cases. *J Bone Joint Surg Br Volume*. 1979;61-B(2):205–8.

5. Iannaci G, Luise R, Sapere P, Costanzo RMA, Rossiello R. Extraskelatal osteosarcoma: A very rare case report of primary tumor of the colon-rectum and review of the literature. *Pathol Res Pract*. 2013;209(6):393–6.
6. Mc Auley G, Jagannathan J, O'Regan K, Krajewski KM, Hornick JL, Butryn-ski J, et al. Extraskelatal osteosarcoma: spectrum of imaging findings. *Am J Roentgenol*. 2012;198(1):W31–7.
7. Choi LE, Healey JH, Kuk D, Brennan MF. Analysis of outcomes in extraskelatal osteosarcoma: a review of fifty-three cases. *J Bone Joint Surg Am*. 2014;96(1):e2–e.
8. Olgyai G, Horvath V, Banga P, Kocsis J, Buza N, Olah A. Extraskelatal osteosarcoma located to the gallbladder. *HPB (Oxford)*. 2006;8(1):65–6.
9. von Hochstetter AR, Hattenschwiler J, Vogt M. Primary osteosarcoma of the liver. *Cancer*. 1987;60(9):2312–7.
10. Banerjee S, Banerjee P. Extraskelatal soft tissue osteosarcoma. *J Indian Med Assoc*. 1987;85(8):242–4.
11. Chung EB, Enzinger FM. Extraskelatal osteosarcoma. *Cancer*. 1987;60(5):1132–42.
12. Shimazu K, Funata N, Yamamoto Y, Mori T. Primary osteosarcoma arising in the colon - Report of a case. *Dis Colon Rectum*. 2001;44(9):1367–70.
13. Heukamp LC, Knoblich A, Rausch E, Friedrichs N, Schildhaus HU, Kahl P, et al. Extraosseous osteosarcoma arising from the small intestinal mesentery. *Pathol Res Pract*. 2007;203(6):473–7.
14. Choudur HN, Munk PL, Nielson TO, Ryan A. Primary mesenteric extraskelatal osteosarcoma in the pelvic cavity. *Skeletal Radiol*. 2005;34(10):649–52.
15. Lee KH, Joo JK, Kim DY, Lee JS, Choi C, Lee JH. Mesenteric extraskelatal osteosarcoma with telangiectatic features: a case report. *Bmc Cancer*. 2007;7.
16. Hussain MI, Al-Akeely MH, Alam MK, Jasser NA. Extraskelatal osteosarcoma, telangiectatic variant arising from the small bowel mesentery. *Saudi Med J*. 2011;32(9):958–61.
17. van den Broek NEJ, Willemsen P, Mattelaer C. A primary extraskelatal osteosarcoma of the mesentery: a case report. *Acta Chir Belg*. 2018;118(2):125–8.
18. Ito S, Terado Y, Shimojima R, Hara Y, Narita K, Tachimori Y, et al. Primary extraskelatal osteosarcoma of the mesentery: A case report. *Int J Surg Case Rep*. 2019;60:111–4.
19. Fine G, Stout AP. Osteogenic sarcoma of the extraskelatal soft tissues. *Cancer*. 1956;9(5):1027–43.
20. Oh S-J, Chang H-K. Unusual giant cell-rich variant of extraskelatal osteosarcoma in the mesentery of small intestine. *Int J Clin Exp Pathol*. 2017;10(11):11225–9.
21. Tong GY, Leow KS, Gunasekaran S, Hue SS-S, Srinivasan S. Primary extraskelatal osteosarcoma of small bowel mesentery presenting with acute bowel obstruction. *J Radiol Case Rep*. 2021;15(12):10–9.
22. Diamantis A, Christodoulidis G, Vasdeki D, Karasavvidou F, Margonis E, Tepetes K. Giant abdominal osteosarcoma causing intestinal obstruction treated with resection and adjuvant chemotherapy. *World J Gastrointestinal Surgery*. 2017;9(2):68–72.
23. Nie X, Fu W, Li C, Lu L, Li W. Primary extraskelatal osteosarcoma of sigmoid mesocolon: a case report and a review of the literature. *World Journal of Surgical Oncology*. 2021;19(1).
24. Salim X, Paton D, Lambers A, Smith RC. First Case of Mesenteric Extraosseous Osteosarcoma in Australia. *Journal of Oncology Medicine & Practice*. 2016;1(1):1–3.
25. Ahmed SA, Mohammed U, Garba ES, Calvin B, Shehu MS. Primary Mesenteric Extraskelatal Osteosarcoma. 2013.
26. Shirazi PH, Rayudu GV, Fordham EW. Extraosseous osteogenic sarcoma of the small bowel demonstrated by 18 F scanning. *Journal of Nuclear Medicine Official Publication Society of Nuclear Medicine*. 1973;14(5):295–6.
27. Bane BL, Evans HL, Ro JY, Carrasco CH, Grignon DJ, Benjamin RS, et al. Extraskelatal osteosarcoma. A clinicopathologic review of 26 cases. *Cancer*. 1990;65(12):2762–70.
28. Thampi S, Matthay KK, Boscardin WJ, Goldsby R, DuBois SG. Clinical Features and Outcomes Differ between Skeletal and Extraskelatal Osteosarcoma. 2014;2014: 902620.
29. Ahmad SA, Patel SR, Ballo MT, Baker TP, Yasko AW, Wang XM, et al. Extraosseous osteosarcoma: Response to treatment and long-term outcome. *J Clin Oncol*. 2002;20(2):521–7.
30. Lee JS, Fetsch JF, Wasdhal DA, Lee BP, Pritchard DJ, Nascimento AG. A review of 40 patients with extraskelatal osteosarcoma. *Cancer*. 1995;76(11):2253–9.
31. Goldstein-Jackson SY, Gosheger G, Delling G, Berdel WE, Exner GU, Jundt G, et al. Extraskelatal osteosarcoma has a favourable prognosis when treated like conventional osteosarcoma. *J Cancer Res Clin Oncol*. 2005;131(8):520–6.
32. Torigoe T, Yazawa Y, Takagi T, Terakado A, Kurosawa H. Extraskelatal osteosarcoma in Japan: multiinstitutional study of 20 patients from the Japanese Musculoskeletal Oncology Group. *J Orthop Sci*. 2007;12(5):424–9.
33. Lidang Jensen M, Schumacher B, Myhre Jensen O, Steen Nielsen O, Keller J. Extraskelatal osteosarcomas: a clinicopathologic study of 25 cases. *Am J Surg Pathol*. 1998;22(5):588–94.
34. Van Rijswijk CSP, Lieng J, Kroon HM, Hogendoorn PCW. Retroperitoneal extraskelatal osteosarcoma. *J Clin Pathol*. 2001;54(1):77–8.
35. Covello SP, Humphreys TR, Lee JB. A case of extraskelatal osteosarcoma with metastasis to the skin. *J Am Acad Dermatol*. 2003;49(1):124–7.
36. Hoch M, Ali S, Agrawal S, Wang C, Khurana JS. Extraskelatal osteosarcoma: a case report and review of the literature. *Journal of radiology case reports*. 2013;7(7):15–23.
37. Wang H, Miao R, Jacobson A, Harmon D, Choy E, Hornicek F, et al. Extraskelatal osteosarcoma: A large series treated at a single institution. *Rare Tumors*. 2018;10.
38. Wang XC, Zhang L, Lin JB, Huang XY, Liang JH, Zhong JP, et al. Imaging diagnosis and differential diagnosis of extraskelatal osteosarcoma. *BMC Cancer*. 2024;24(1):11.
39. Heukamp LC, Knoblich A, Rausch E, Friedrichs N, Schildhaus HU, Kahl P, et al. Extraosseous osteosarcoma arising from the small intestinal mesentery. *Pathol Res Pract*. 2007;203(6):473–7.
40. Sordillo PP, Hajdu SI, Magill GB, Golbey RB. Extraosseous osteogenic sarcoma. A review of 48 patients. *Cancer*. 1983;51(4):727–34.
41. Wiklund TA, Blomqvist CP, Raty J, Elomaa I, Rissanen P, Miettinen M. Postirradiation sarcoma. Analysis of a nationwide cancer registry material. *Cancer*. 1991;68(3):524–31.

## Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.