

Case Reports

Extraskelatal osteosarcoma, telangiectatic variant arising from the small bowel mesentery

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ABSTRACT

يعد الورم العظمي العفلي (الغرن العظمي) من الأورام الخطيرة جداً والخبيثة التي تصيب الأنسجة الرخوة، ويتصف هذا المرض بإنتاج الأنسجة العظمية الورمية من دون الوصول إلى العظم أو السمحاق. ويندر أن يصيب هذا الورم الأعضاء الحشوية حيث لم يعلن الأدب الطبي الغربي إلا عن 3 حالات مصابة بالورم العظمي العفلي المساريقي. نستعرض في هذا المقال حالة مريض ذكر يبلغ من العمر 40 عاماً، وتم تشخيص حالته بمرض الورم العظمي العفلي الناتج عن مساريق الأمعاء الدقيقة. لقد جاء هذا المريض إلى المستشفى بعد إصابته بنزيف في أسفل المنطقة المعوية المعدية، وقد كان أعلى هذه المنطقة طبيعياً بعد إجراء الفحص الداخلي بالمنظار وتنظير الأمعاء. وأظهرت الأشعة المقطعية وجود كتلة متعددة المواقع ومحددة الشكل في المنطقة السفلية البطنية المقاربة من الأمعاء الدقيقة، ويبلغ حجمها 13×7×7 سم مع انتشار المرض في الكبد. ولقد قمنا باستئصال الورم بكتلة واحدة مع الأمعاء الدقيقة الملائقة، وكشف فحص الأنسجة التي تم استئصالها عن وجود أورام نقيلة من النوع العظمي العفلي. وخلال هذا المقال قمنا أيضاً بمناقشة كيفية تشخيص الورم العظمي العفلي، وكيفية علاجه، والنتائج التي توصلنا إليها.

Extraskelatal osteosarcoma (EOS) is a highly aggressive and rare malignant soft tissue tumor, characterized by the production of neoplastic osseous tissue without attachment to the bone or periosteum. It rarely involves the visceral organs. Only 3 cases of mesenteric EOS have been reported in English literature. Here, we describe a male patient of 40 years, who was diagnosed to have EOS arising from small bowel mesentery. This patient presented with lower gastrointestinal (GI) bleeding. Upper GI endoscopy and colonoscopy were normal. Computed tomography scan demonstrated a well defined multi-lobulated mixed density mass lesion measuring about 13x7x7 cm in lower abdomen adjacent to small bowel loops with liver metastasis. Palliative en bloc resection of tumor with adjacent small bowel was performed. The histopathology revealed a telangiectatic type

osteosarcoma of mesentery. Diagnosis of EOS, its management and the outcome in context of the current literature are discussed.

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Extraskelatal osteosarcoma (EOS) is a highly aggressive and rare mesenchymal tumor characterized by the production of neoplastic osseous tissue without attachment to the bone or periosteum.¹ It accounts for less than 1% of all soft tissue sarcomas and approximately 4% of all osteosarcomas.²⁻⁴ Common sites of involvement are deep soft tissue of thigh (47%), upper extremity (20%), and retroperitoneum (17%).⁴ It rarely involves the intra peritoneal visceral organs. Only 3 cases of mesenteric EOS have been reported in the English literature.⁵⁻⁷ Due to the rare nature of disease, preoperative diagnosis in patients with abdominal mass is difficult and is often confirmed after laparotomy and histopathology. We describe the clinical presentation, diagnostic modalities, treatment, and the postoperative course of a patient of this rare type of EOS.

Case Report. A 40-year old male, smoker, presented with per rectal bleeding and off and on melena for the last 15 days. He had a history of occasional abdominal discomfort for the last 6 months. On general physical examination, he was looking pale but of good built with stable vital signs. Local examination revealed protuberant abdomen with mild peri-umbilical tenderness. No organomegaly or ascites was detected. Bowel sounds were present. Per rectal examination

revealed melena. On investigation, hemoglobin was 6.2 gram percent (normal value 12.5-16.5 gram/dl). Other laboratory findings including blood chemistry, liver, and renal function tests were within normal ranges. Upper GI endoscopy showed mild gastritis. Colonoscopy was normal. Radioactive labeled RBC scan was undetectable of any ongoing bleeding. Computed tomography (CT) scan demonstrated a well defined multi-lobulated mixed density mass lesion measuring approximately 13x7x7 cm in lower abdomen adjacent to small bowel loops. It was surrounded by fat stranding and spiculations were noticed at its superior medical aspect (Figure 1). There was a single enlarged lymph node measuring approximately 2x2.5 cm in portahepatis. Two attenuated lesions 7x7x6 cm and 5x5x3 cm in the right lobe of liver were also observed. Computed tomography scan of the chest demonstrated enlarged mediastinal lymph nodes. At exploratory laparotomy a multi lobulated mass of 15x10x8 cm having solid and cystic components was found, arising from the mesentery of small bowel encroaching up to the mesenteric border of jejunum (Figure 2). There were 2 metastatic lesions in the right lobe of liver. Palliative en-bloc resection of tumor with adjacent small bowel was performed and side to side anastomosis was constructed with GIA-80. Cut section of the mass consisted of grey-white solid area with gritty sensation and large multi cystic areas, containing clotted blood, necrotic debris, and hemorrhagic fluid. Postoperative recovery was uneventful. The histopathology revealed a telangiectatic type osteosarcoma of mesentery (Figures 3 & 4). No primary or metastatic lesion was found on bone scan. He was discharged on his request in good general condition because he preferred to complete the rest of treatment in his own country. While there, he had a wide excision of the metastatic hepatic lesions and completed a full course of chemotherapy. On telephonic follow up,

he remained well and enjoyed a good quality of life for approximately 14 months. One and a half years after his surgery, he presented again in the emergency department of our hospital with the same complaints of per rectal bleeding and melena. After resuscitation he had upper GI endoscopy. It revealed a malignant ulcer in first part of duodenum which underwent heat probe coagulation with satisfactory hemostasis. Computerized tomography scan of the abdomen showed multiple

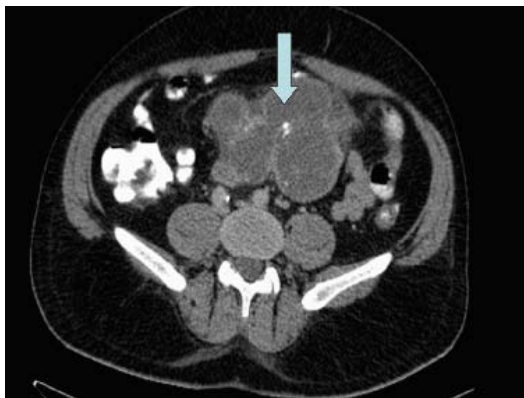


Figure 1 - Computerized tomography scan of the abdomen showing a heterogeneous mesenteric mass with focal calcification.



Figure 2 - Excised specimen comprises of tumor and adjacent small bowel.

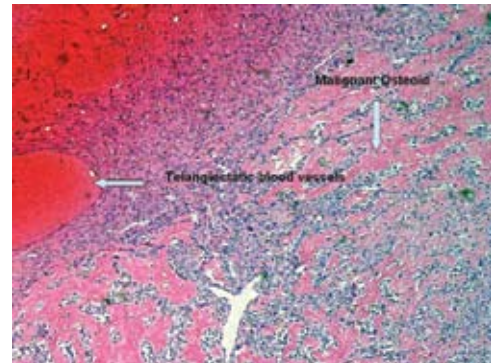


Figure 3 - Low Power view showing the presence of malignant osteoid with telangiectatic blood vessels surrounded by bizarre atypical spindle shaped stromal cells.

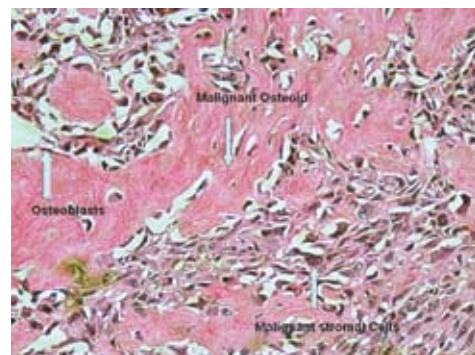


Figure 4 - High power view (40x) demonstrates eosinophilic malignant osteoid rimmed by osteoblasts and surrounded by malignant spindle shaped stromal cells.

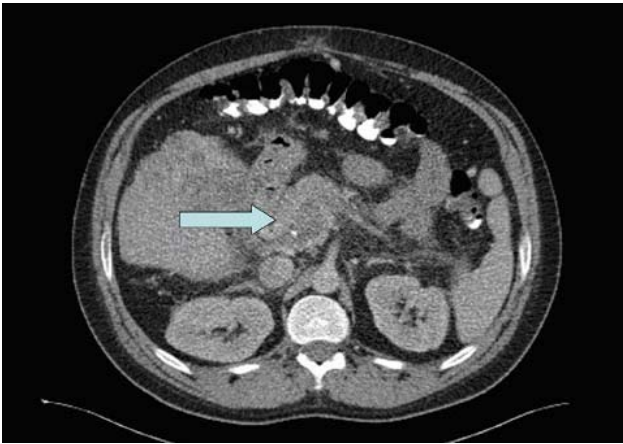


Figure 5 - Computerized tomography scan of the abdomen showing the local recurrence with focal area of calcification involving the duodenum.

hypodense focal lesions with foci of calcifications in both lobes of liver. A metastatic lesion of 7x3.5 cm, involving the first, second and third part of duodenum, surrounded by multiple enlarged lymph nodes, invading the portal vein and hepatic artery and ascites was noticed (Figure 5). Oncology team suggested chemotherapy, as the recurrent tumor was not resectable, but patient requested for discharge and went back to his country.

Discussion. Primary osteosarcomas of bone occur predominantly in the first decades of life while EOS has peak prevalence in the sixth decade of life. Men are slightly more frequently affected with a ratio of 1.9:1.⁸ Our patient was male, in fourth decade, younger than the described age in the literature. The clinical presentation of our patient with GI bleeding was almost similar with the case reported by Lee et al.⁷ Symptoms usually include a slow growing painful mass that is visible on plain radiographs, CT or MRI. Radiological studies typically show large soft tissue masses with focal to massive area of mineralization and lack of osseous involvement.⁴ However, other soft tissue sarcomas and carcinomas can also present with significant area of calcification and thus histopathology is required to establish the definite diagnosis. Based on the excessive production of malignant osteoid, the appearance of osteoblast like tumor cells and exclusion of other sarcomatous differentiation, EOS was diagnosed. Nucleoli display high grade pleomorphism and frequent mitosis including abnormal mitotic figure. On immunohistochemistry, Ki-67 showed high proliferative index depicted by nuclear positivity of tumor cells. The pathogenesis of tumor is unclear. Cases usually arise de novo and no precursor lesions are known. Radiation induced EOS develops at least 4 years, following high

dose radiations. In addition, a history of trauma has been reported in 12-30% of patients.^{4,9} However, there was no history of trauma or radiation in our patient. Extraskeletal osteosarcoma arising from unusual sites, like kidney, urinary bladder, mediastinum, pleura, diaphragm, esophagus, small intestine, liver, gallbladder, mesentery, parotid, breast, uterus, larynx, hand and cerebellum have been reported.² Mesenteric EOS has been described previously only in 3 case reports in the English literature.⁵⁻⁷

Extraskeletal osteosarcoma has poor prognosis, and approximately 75% of the patients die of the disease within 5 years of diagnosis.^{3,4,8} Local recurrence develops in approximately one third of the patients and pulmonary metastasis in 60%.¹⁰ Tumor size is the most important prognostic factor, as the patients with a lesion larger than 5 cm have a worse clinical outcome. Other poor prognostic factors include the presence of metastatic lesions, patient age and increased lactate dehydrogenase and alkaline phosphatase level.²⁻⁵ Our patient with a 15 cm mass and liver metastasis was alive 18 month after the diagnosis, the longest survival among the 3 reported cases of mesenteric EOS. All major sub types of primary osteosarcoma can also be found in EOS. The most common is the osteoblastic variant, followed by fibroblast chondroids, telangiectatic small cell, and well differentiated type.⁵ Wide resection followed by adjuvant chemotherapy and radiotherapy is the available mode of management for these patients. Adjuvant chemotherapy has proved to be effective in all the cases of EOS mesentery. Radical resection appears to be the best therapeutic option. Resection of liver or pulmonary metastasis may prolong the survival in these patients.

In conclusion, EOS can involve the peritoneum and should be considered in the differential diagnosis of mesenchymal tumor of abdominal cavity. It is an uncommon entity, with poor prognosis, and radical surgical resection is the main stay of treatment. More information regarding the appropriate management plan of EOS needs to be discussed and published.

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