

Primary Gastric Extraosseous Osteosarcoma with Extensive Multivisceral Invasion: A Case Report

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Abstract

Introduction: Extraosseous osteosarcoma (ESOS) is a rare soft-tissue sarcoma that produces osteoid without anatomical relationship to the skeleton. Primary gastric ESOS is exceptionally rare and poses major diagnostic and therapeutic challenges. **Case Presentation:** A 78-year-old man with six months of nonspecific gastrointestinal symptoms had a 14 × 16 × 21 cm gastric mass with extensive local involvement. Endoscopic ultrasound-guided fine-needle aspiration was non-diagnostic. After massive upper gastrointestinal hemorrhage with hemodynamic instability, emergency laparotomy and R2 debulking were performed. Histopathology showed a high-grade osteoid- and chondroid-producing malignant mesenchymal tumor; SATB2 was positive. PET-CT showed no skeletal primary, supporting gastric ESOS. **Conclusion:** Primary gastric ESOS may present as a giant locally invasive mass with non-diagnostic biopsy. Complete resection should be pursued when feasible, while emergency presentation may necessitate non-curative surgery.

Keywords: Gastrointestinal Hemorrhage; Osteosarcoma; Sarcoma; Soft Tissue Neoplasms; Stomach Neoplasms.

1. Introduction

Extraosseous osteosarcoma (ESOS) is a rare malignant mesenchymal neoplasm characterized by osteoid or immature bone matrix production outside the skeleton, without anatomical attachment to bone or periosteum¹. According to the World Health Organization classification, diagnosis requires demonstration of tumor-produced osteoid, absence of skeletal attachment, and exclusion of metastasis from a primary bone osteosarcoma².

ESOS accounts for approximately 1% of soft-tissue sarcomas and occurs predominantly in older adults, with the deep soft tissues of the extremities and retroperitoneum among the most common sites^{3,4}. Visceral involvement is uncommon, and primary gastric ESOS is exceptionally rare. A recently published primary gastric ESOS case was described as the first clearly documented example identified after a comprehensive literature search⁵. Earlier clinicopathologic series likewise show that ESOS usually arises in non-

visceral soft-tissue sites⁶. We report a giant high-grade gastric ESOS presenting with massive gastrointestinal hemorrhage and requiring emergency R2 debulking.

2. Case Presentation

A 78-year-old man with hypertension, type 2 diabetes mellitus, prior cerebrovascular accident with residual hemiparesis, coronary artery disease after stenting, benign prostatic hyperplasia, and bilateral carotid stenting presented with a six-month history of nausea, vomiting, abdominal pain, and back pain. He had previously been evaluated at several centers, with provisional diagnoses including a benign soft-tissue tumor, retroperitoneal mass, and gastrointestinal stromal tumor (GIST).

Contrast-enhanced computed tomography (CT) demonstrated a giant heterogeneous upper-abdominal mass measuring $14 \times 16 \times 21$ cm, with central necrotic and calcified areas, extensive contact/infiltration toward the left hepatic lobe, body and tail of the pancreas, duodenum, and omentum, and compression of the portal confluence and inferior vena cava (Figure 1A, B). Upper gastrointestinal endoscopy showed grade C esophagitis and a suspected perforated ulcer/fistula at the duodenal bulb. Endoscopic ultrasonography demonstrated a 10×8 cm solid lesion; endoscopic ultrasound-guided fine-needle aspiration was non-diagnostic.

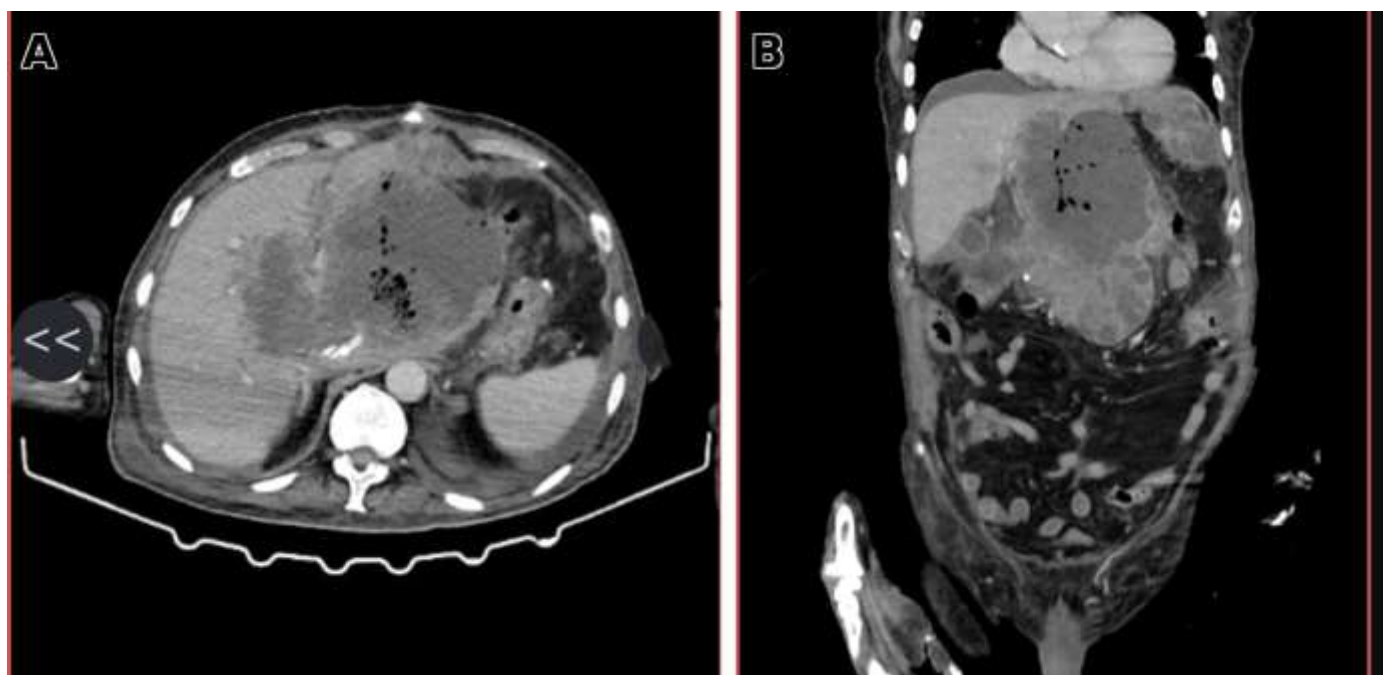


Figure 1. Contrast-enhanced computed tomography (CT) images of the upper-abdominal mass. (A) Axial section: a giant heterogeneous mass with central necrotic and calcified areas. (B) Coronal section: heterogeneous giant upper-abdominal mass with extensive local involvement.

Whole-body ^{18}F -FDG PET-CT showed an upper-abdominal mass conglomerate involving the gastrohepatic, peripancreatic, and mesenteric regions, with heterogeneous intense uptake ($\text{SUV}_{\text{max}} 15.11$), intralesional air and necrosis, and hepatic surface involvement (Figure 2). No distant skeletal lesion or primary bone tumor was detected.

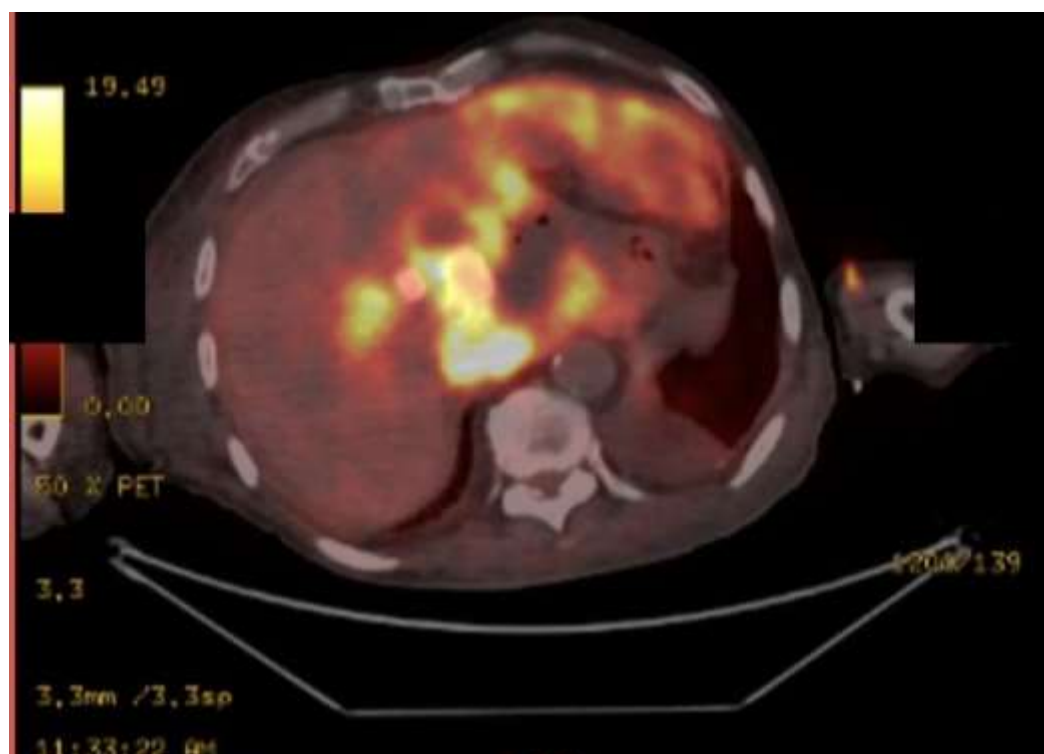


Figure 2. 18F-FDG PET-CT image showing intense, heterogeneous tracer uptake (SU-Vmax 15.11) within the upper-abdominal mass conglomerate; no distant skeletal lesion or primary bone tumor was detected.

During follow-up, massive upper gastrointestinal hemorrhage with hemodynamic instability developed, prompting emergency laparotomy for a life-saving indication. Exploration revealed a giant friable tumor occupying most of the upper abdomen, associated with the gastric wall and showing dense adherence and infiltration toward surrounding tissues. Because of the tumor size, extensive local involvement, and emergency conditions, oncologically complete R0 resection was not considered feasible. Debulking surgery was therefore performed with maximal removal of the dominant tumor mass and limited excision of involved or adherent peritumoral tissues. No formal gastrectomy, pancreatic resection, hepatectomy, or other major organ resection was performed. Gross residual tumor remained; therefore, the procedure was classified as an R2 debulking resection.

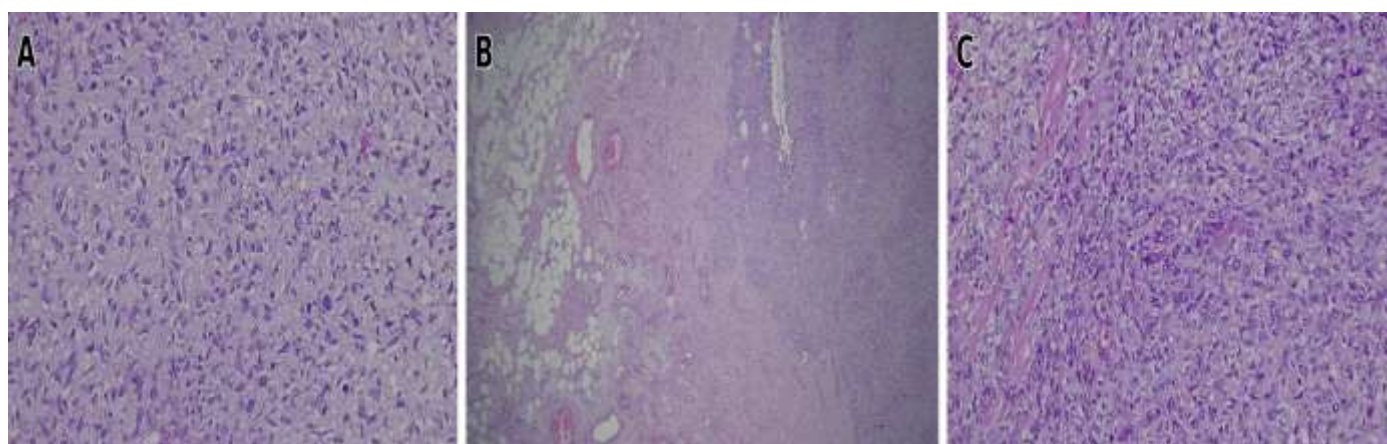


Figure 3. Histopathological appearance (hematoxylin and eosin staining). (A) Pleomorphic malignant mesenchymal tumor cells with hyperchromatic nuclei, abundant eosinophilic cytoplasm, and frequent mitoses (×200). (B) Tumor invasion front infiltrating surrounding tissues (×100). (C) Extensive osteoid matrix produced between pleomorphic tumor cells in the stroma (×200).

Histopathological examination demonstrated a high-grade malignant mesenchymal neoplasm with extensive osteoid and chondroid matrix formation, marked cytologic atypia, frequent mitotic and atypical mitotic figures, extensive necrosis, and infiltration of surrounding tissues (Figure 3A-C). Immunohistochemistry showed strong nuclear SATB2 positivity (Figure 4), S100 positivity in chondroid areas, and negative staining for MDM2, CD117, DOG1, and keratin, supporting the diagnosis of high-grade extraosseous osteosarcoma. One peripancreatic lymph node showed metastatic tumor involvement. Because the debulking specimen was fragmented and the surgical margins could not be anatomically oriented, microscopic margin status could not be reliably assessed.

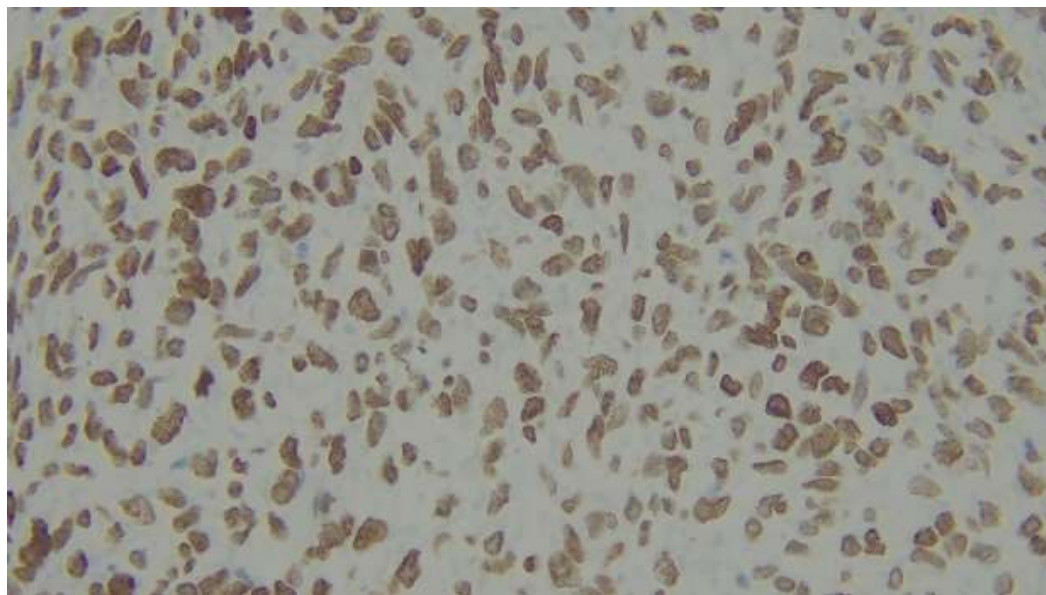


Figure 4. Diffuse and strong nuclear SATB2 positivity in tumor cells (SATB2 immunohistochemistry, $\times 200$).

The postoperative course was uneventful and the patient was discharged. Adjuvant systemic therapy was discussed by the multidisciplinary oncology council but was not administered because of advanced age, performance status, and comorbidities. At six postoperative months, the patient was alive, although clinical and radiological progression had been observed.

3. Discussion

ESOS is a biologically aggressive soft-tissue sarcoma occurring mainly in older adults^{3,4,6}. Primary gastric ESOS is extraordinarily uncommon. Meng et al. recently reported a 65-year-old woman with a 13-cm primary gastric ESOS treated by emergency radical distal gastrectomy with R0 resection and no recurrence at nine months⁵. In contrast, the present case involved a 21-cm tumor, extensive local infiltration, life-threatening upper gastrointestinal hemorrhage, and inability to achieve complete resection (Table 1).

Preoperative diagnosis may be difficult because extensive necrosis, fibrosis, and intratumoral heterogeneity can make limited biopsies non-diagnostic. In the present case, definitive diagnosis required examination of the surgical specimen. The differential diagnosis included GIST, leiomyosarcoma, dedifferentiated liposarcoma, sarcomatoid carcinoma, metastatic osteogenic sarcoma, and osteosarcomatous sarcomatoid mesothelioma. Tumor-derived osteoid remained the key diagnostic feature, while the immunohistochemical profile and absence of a skeletal primary supported gastric ESOS.

Table 1. Comparison of reported primary gastric extraosseous osteosarcoma cases.

Characteristic	Meng et al. (2026) ⁵	Present case (2026)
Age / sex	65 / Female	78 / Male
Primary site	Stomach	Stomach
Tumor size	11 × 9.5 × 13 cm	14 × 16 × 21 cm
Presentation	Abdominal pain/distension, nausea/vomiting, bowel obstruction	Nausea/vomiting, abdominal/back pain; later massive upper GI hemorrhage
Surgery	Emergency radical distal gastrectomy, mass resection, regional lymph node dissection	Emergency R2 debulking; no formal gastrectomy, pancreatectomy, hepatectomy, or other major organ resection
Pathology	High-grade extraskeletal osteosarcoma; SATB2 positive	High-grade extraosseous osteosarcoma; SATB2 positive
Resection status	R0	R2; microscopic margins not reliably assessable because specimen was fragmented/non-oriented
Adjuvant treatment	Recommended but not administered	Recommended but not administered
Follow-up / outcome	9 months; no recurrence or distant metastasis	6 months; alive with clinical/radiological progression

Abbreviations: GI, gastrointestinal; R0, microscopically margin-negative resection; R2, macroscopic residual disease.

For localized ESOS, wide surgical excision with negative margins is generally regarded as the principal treatment and is associated with improved local disease control^{3,7}. In this patient, extensive local disease and active life-threatening hemorrhage made a formal multivisceral R0 resection impractical in the emergency setting; surgery was therefore directed toward hemorrhage control and reduction of tumor burden through R2 debulking.

The benefit of chemotherapy in ESOS remains uncertain. Retrospective data have suggested possible benefit from osteosarcoma-based, particularly platinum-containing regimens, but the evidence is inconsistent⁸. In a large international series of localized ESOS, radiotherapy was associated with reduced local recurrence, whereas chemotherapy did not reduce systemic recurrence and neither modality was associated with improved overall survival⁹. Thus, adjuvant treatment should be individualized in multidisciplinary discussion, especially in elderly patients with major comorbidities.

This report is limited by its single-case design, fragmented R2 specimen, inability to assess microscopic margins reliably, absence of extended molecular profiling, and short follow-up. Nevertheless, it illustrates the diagnostic difficulty and operative decision-making that may arise when a rare gastric ESOS presents as a massive locally advanced tumor with acute hemorrhage.

4. Conclusion

Primary gastric extraosseous osteosarcoma is an exceptionally rare and aggressive malignancy. Diagnosis requires recognition of malignant osteoid production and exclusion of a skeletal primary and relevant mimics. Complete resection with negative margins should be pursued when feasible; however, extensive local disease and emergency presentation may prevent curative surgery. The present case demonstrates the clinical complexity of a giant gastric ESOS requiring emergency R2 debulking after life-threatening gastrointestinal hemorrhage.

Patient Consent Statement: Written informed consent for publication of the clinical details and accompanying images was obtained from the patient.

Ethical Approval: Institutional ethics committee approval is not required for publication of an anonymized single-patient case report at our institution. All procedures were conducted in accordance with institutional ethical standards and the Declaration of Helsinki and its later amendments.

Author Contributions (CRediT): Conceptualization: Y.K., C.K.P.; Investigation: Y.K., S.G., U.T., İ.A., Z.Z., H.G., C.K.P.; Resources: Y.K., S.G., U.T., İ.A., Z.Z., H.G., C.K.P.; Formal analysis: Ç.D., K.E.A.; Data curation: Y.K., S.G., U.T.; Visualization: Y.K., Ç.D., K.E.A.; Writing – original draft: Y.K.; Writing – review & editing: S.G., U.T., Ç.D., K.E.A., C.K.P.; Supervision: C.K.P., U.T. All authors have read and approved the final version of the manuscript and accept full responsibility for its content.

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Data Availability Statement: All data generated or analysed during this case report are included in this article. Additional anonymized information is available from the corresponding author upon reasonable request.

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