Pancreatic Embryonal Rhabdomyosarcoma

Narendra Pandit[1] and Durga Neupane[2]

[1]Birat Medical College
[2]BP Koirala Institute of Health Sciences

March 07, 2025

Pancreatic Embryonal Rhabdomyosarcoma

Narendra Pandit[1], Durga Neupane[2]

Professor, MS, MCh, FACS, Department of Surgical Gastroenterology, Birat Medical College and Teaching Hospital, Biratnagar, Nepal

MBBS, Department of Surgery, B.P. Koirala Institute of Health Sciences, Dharan, Nepal

Authors

Narendra Pandit (Corresponding author)

Email: narendrapandit111@gmail.com

Durga Neupane

Email: neupanedurga26@gmail.com

Word count: 500

Keywords: pancreas; rhabdomyosarcoma; IHC; embryo

Key clinical message:

A 32-year-old breastfeeding mother with diabetes mellitus presented with abdominal pain, weight loss, and an 8 cm pancreatic mass. Surgery revealed a high-grade embryonal rhabdomyosarcoma. Despite R0 resection and chemotherapy, she had early liver recurrence within three months. Histopathology confirmed pancreatic embryonal rhabdomyosarcoma with high Ki-67 (90%).

A 32-year-old breastfeeding mother presented with upper abdominal pain, anorexia, vomiting, and weight loss for four months. She had been treated with subcutaneous insulin for diabetes mellitus for the past three years. On examination, she appeared pale and had a vague 8 cm lump in the epigastric region. Blood tests showed low hemoglobin (7.5 gm/dl) with normal liver and kidney function. A contrast CT scan of the abdomen revealed a heterogeneous mass in the neck and body of the pancreas (Figure 1), confirmed by an MRI, with no signs of metastasis. Tumor markers were normal. After stabilizing her with blood transfusions, she was diagnosed with a diffuse pancreatic neoplasm (solid pseudopapillary neoplasm-SPN) and Type 3c diabetes mellitus. She underwent an extended distal pancreatosplenectomy (Figure 2, Figure 3), as the tumor was resectable and preoperative biopsy facilities were unavailable. Postoperative recovery was smooth, received Pneumovac vaccination at discharge, and histopathology was requested. On histopathological microscopy, it revealed proliferation of atypical spindle cells arranged in intersecting short fascicles. The IHC report confirmed high grade sarcoma (embryonal rhabdomyosarcoma) of the pancreas (IHC image is not available as it was done in India). Immunohistochemical staining was positive for vimentin, desmin, myogen and MyoD1; and negative for SOX10, MDM2, p16, CDK4, synaptophysin, CEA, ER, CK and S100. Ki-67 was

90%. Unfortunately, despite R0 resection and adjuvant chemotherapy, she had an early recurrence in liver at multiple sites at three months follow-up.

Rhabdomyosarcoma (RMS) is a rare malignancy originating from embryonic mesenchyme, with the potential to differentiate into skeletal muscle.[1] Although it accounts for about 50% of all soft tissue sarcomas in children, it represents only 3% of such cases in adults, usually occurring in skeletal muscle. Initially described by Webner in 1854, RMS has since been categorized into four histopathological types: embryonal, botryoid, alveolar, and pleomorphic.[1] RMS affecting the biliary tree is particularly uncommon, predominantly seen in infants and children. Shirafkan A and colleagues reported the first case of undifferentiated pleomorphic RMS involving the pancreas, with no other similar cases documented.[2]

Diagnosing RMS before surgery is challenging due to vague symptoms, such as abdominal discomfort, weight loss, and palpable masses. Imaging often reveals non-specific masses with peripheral enhancement, while preoperative biopsies tend to be inconclusive since specialized staining is rarely performed. Pathological confirmation requires identifying atypical spindle cells arranged in intersecting fascicles, alongside muscle-specific markers like myogen, desmin, SMA, and vimentin.

Treatment for adult RMS primarily involves surgical removal of the tumor, followed by chemotherapy and radiotherapy. Studies indicate that adult RMS is chemo-sensitive, with a 75% response rate.[3] The standard regimen includes vincristine, dactinomycin, and cyclophosphamide. Prognosis is influenced by factors like age, tumor histology, location, stage, and local control measures. Older patients and certain histological subtypes show reduced chemosensitivity and tolerance for aggressive therapy.[3] Patients responding to chemotherapy often experience extended metastasis-free intervals. A study by Ogilvie highlighted that combining chemotherapy (using doxorubicin, ifosfamide, and vincristine) with surgery and radiation improved 2-year survival rates to 55% overall and 64% disease-free survival.[3]Nonetheless, long-term outcomes for adult RMS remain poor, with survival rates ranging from 12.5% to 50% within the first year post-treatment.

Conflict of interest: None

Funding: None

Consent: Written informed consent was obtained from the patient for reporting this case image.

Author contribution: Both the authors contributed equally in preparing the manuscript. Final version is approved by both authors.

Figure legends

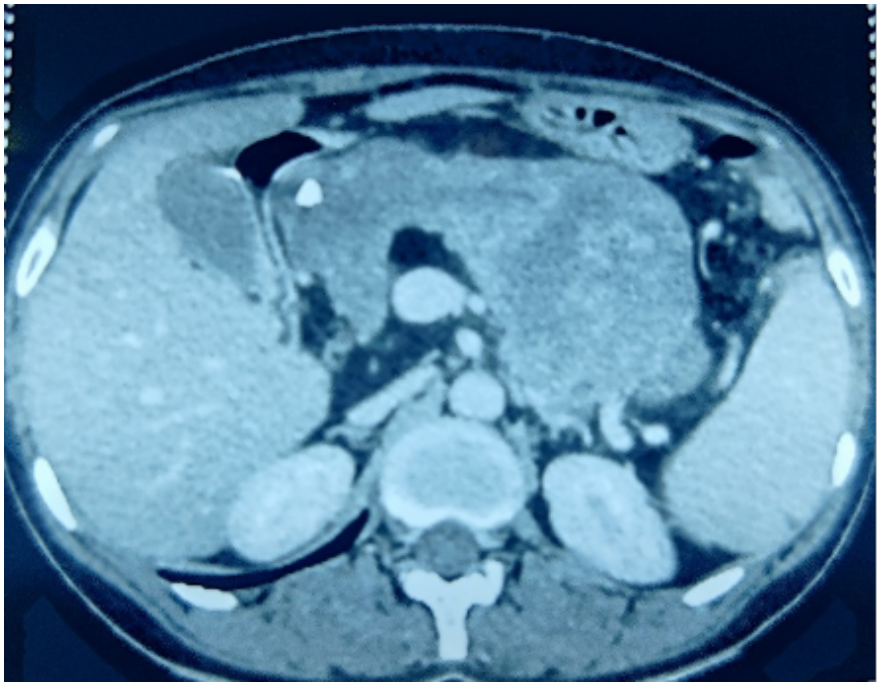
Figure 1. Diffusely enlarged, irregular, lobulated body of pancreas with heterogenous enhancement.

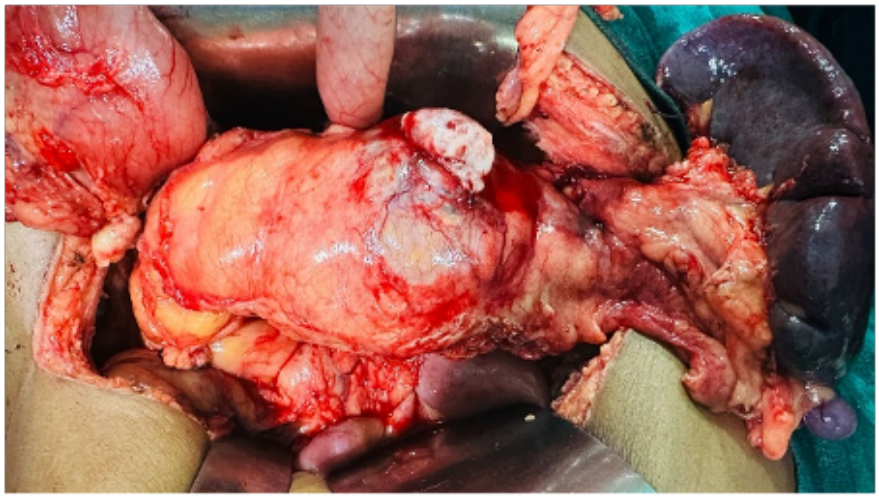
Figure 2: Intraoperative view showing mobilized pancreatic tumor with the spleen.

Figure 3. Cut section of the specimen showing well-capsulated, fleshy tumor.

References

1. A. Hayes-Jordan, R. Andassy, Rhabdomyosarcoma in children, Curr. Opin. Pediatrics 21 (2009) 373–378.
2. A. Shirafkan, N. Boroumand, S. Komak, A. Duchini, Pancretic pleomorphic rhabdomyosarcoma, International Journal of Surgery Case Reports 13 (2015) 33–36.
3. C. Ogilvie, Treatment of adult rhabdomyosarcoma, Am. J. Clin. Oncol. 32 (4) (2009) 436–442





Hosted file

`Figure 3.docx` available at https://authorea.com/users/612370/articles/1275019-pancreatic-embryonal-rhabdomyosarcoma