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Extraosseous Ewing's sarcoma of the pancreas

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Dear Editor,

We present the clinical case of a 21-year-old male with abdominal pain in the left hypochondrium radiating to the ipsilateral lumbar area and a weight loss of 2kg over a month, secondary to a large palpable intra-abdominal mass.

An abdominal computed tomography (CT) scan was performed as the first diagnostic test which showed a mass suggestive of pancreatic origin, replacing the body and tail of the pancreas, measuring 14x11x7cm, with solid characteristics and a necrotic-cystic component, infiltrating the spleen, displacing the gastric chamber and enveloping the celiac trunk with narrowing of the splenic artery and thrombosis of the splenic vein. The study was completed with a positron emission tomography (PET) scan confirming a hypercaptant lesion in the femur suggestive of bone metastasis. A directed biopsy (CT-guided) of the abdominal mass was performed, with histological and immunohistochemical findings of small cell tumor positive for CD99, NKX2, and FLI1. Given a high suspicion of a tumor from the Ewing sarcoma (ES) family, the histological piece was sent to a reference center for molecular study. Molecular study by FISH revealed positivity for the rearrangement of the EWSR1/FLI1 gene t(11;22)(q24;q12.2), confirming Extraosseous Ewing Sarcoma (EES). It was staged as stage IV metastatic EES

and chemotherapy treatment was proposed (Vincristine-Doxorubicin-Cyclophosphamide/Ifosfamide-Etoposide regimen).

Discussion

EES is a rare, aggressive soft tissue tumor with a poor prognosis. Its most common locations include the paravertebral region, lower extremities, thoracic wall, and retroperitoneum. The age of presentation is usually teenagers and young adults (10-30 years old). Its pancreatic location is extremely rare, with just over 25 cases described in the literature, and its presents as a painful abdominal mass with rapid growth, often accompanied by jaundice and anemia. CT may show an isodense or hypodense mass depending on the degree of necrosis and one-third of tumors present calcifications. Definitive diagnosis is based on histology, immunohistochemical characteristics and cytogenetic analysis. Treatment is based on chemotherapy along with adjuvant radiotherapy or surgery, with a 5-year overall survival rate of 60-70%, decreasing by less than 25% in cases of metastasis.

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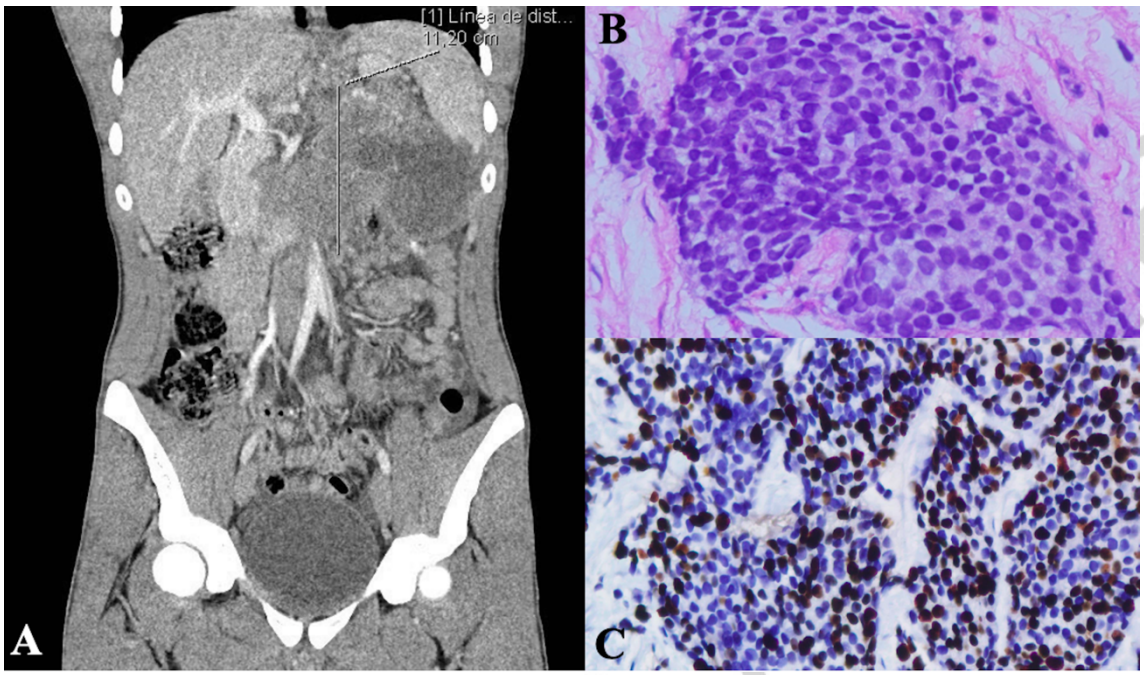

Fig.1. A) CT image in coronal section showing a large heterogeneous mass deppendig from the pancreas and replacing its body and tail. B) and C) Histological images: (H&E 60x) small, round, hyperchromatic, undifferentiated tumor cells and Ki-67 (40x) with a high mitotic index.