

Primary pancreatic sarcoma

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CASE REPORT

A 53-year-old woman with a past medical history of appendectomy and neurinoma on the left side of the neck in 1986 was admitted for asthenia and itching of recent onset, with absence of abdominal pain or any alterations on physical examination. A blood test showed: alkaline phosphatase, 1,499 IU/L, gamma-glutamyl transpeptidase, 1,163 IU/L, total bilirubin, 1.62 mg/dl, and CA-19.9, 533 IU/ml (normal value < 37 IU/ml). Doppler-US and abdominal CT scans were performed, which revealed a mass measuring 6 cm in diameter located in the head of the pancreas, with dilated intra- and extra-hepatic bile ducts. The lesion had cystic areas with solid septa and papillary epithelium, some of which stained very brightly (Figs. 1 and 2); no evidence of vascular invasion or adenopathies was observed. With a diagnostic impression of resectable cystic neoplasm of the pancreas with no surgical contraindications a cephalic duodenopancreatectomy was performed. In the surgically removed tissue a solid mass of whitish color and elastic consistency was observed. The mass was located in the head of the pancreas. Although not encapsulated, it showed well-defined borders and was easily removed from the adjacent pancreas. An immunohistochemical study showed that it was positive for vimentin marker, and negative for cytokeratins (AE1-AE3), S-100, chromogranin, CD 34, C-KIT, and actin. Furthermore, a low cell proliferation index (MIB-I) was also observed (Fig. 3). These data from the histological study suggested the presence of a low-malignancy-grade fusocellular sarcoma. After a 2-year follow-up, the patient remained asymptomatic and without recurrent disease.



Fig. 1. Abdominal US showed a mass measuring 6 cm in diameter located in the pancreatic area.
Ecografía abdominal donde se observa una masa de 6 cm de diámetro localizada en área pancreática.



Fig. 2. Abdominal CT scan shows a mass in the head of the pancreas, causing displacement and compression of adjacent structures.
TAC abdominal donde se aprecia una masa en la cabeza del páncreas, que desplaza y comprime las estructuras adyacentes.

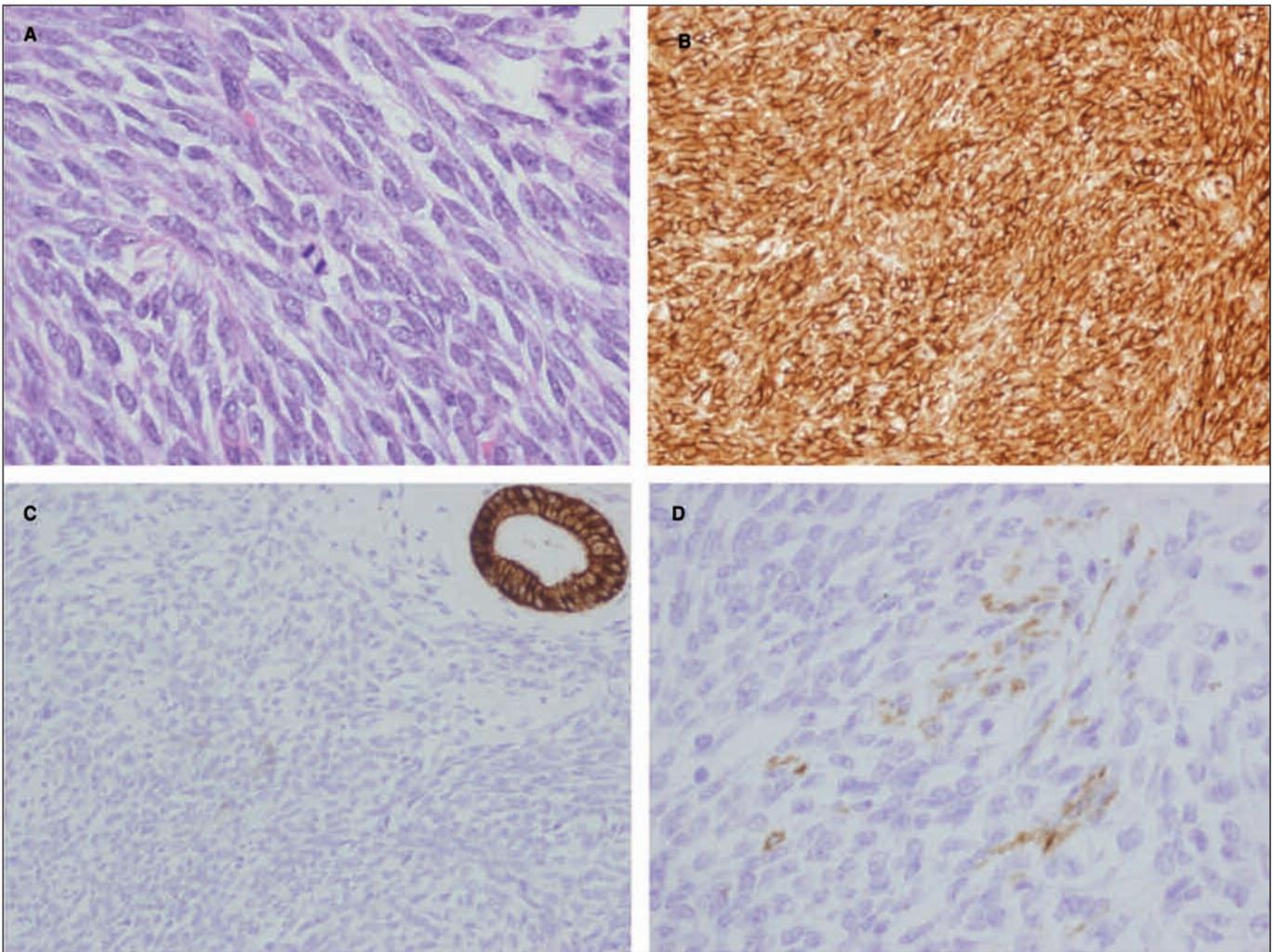


Fig. 3. Immunohistochemical study: A. Hematoxylin and eosin staining with the presence of fusiform cells having elongated nuclei and blunt ends, and showing some mitoses. B. Positive vimentin marker. C. Negative for cytokeratins (AE1-AE3). D. Focally positive for desmin.

Estudio de inmunohistoquímica: A. Tinción de HE donde se observa la presencia de células fusiformes, con núcleos alargados y de bordes romos, apreciándose alguna mitosis. B. Positividad para el marcador vimentina. C. Negatividad para citoqueratinas (AE1-AE3). D. Desmina focalmente positivo.

DISCUSSION

Primary pancreatic sarcomas are an exceptional disease representing less than 1% of pancreatic neoplasms (1-4). They derive from the mesenchymal support tissue in the pancreas, which includes several histological subtypes such as the leiomyosarcoma discussed here (2). They are characterized as having a slow growth rate, so generally they are large when diagnosed, presenting symptoms as a result of the compression produced on neighboring organs. This entity should be differentiated from other types of sarcomas that develop more frequently and originate in the connective tissue surrounding the pancreas, giving rise to a secondary invasion of this organ (2,3). Potential findings in radiographic data include a mass with heterogeneous density, weak staining, peripheral and irregular outline at times, and variable thickness (4). Hence, pancreatic sarcoma can be easily mistaken for pancreatic adenocarcinoma or cystic neoplasm of the pancreas (4,5).

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