ORIGINAL ARTICLE

Primary leiomyosarcoma of the pancreas with metastasis to the spleen

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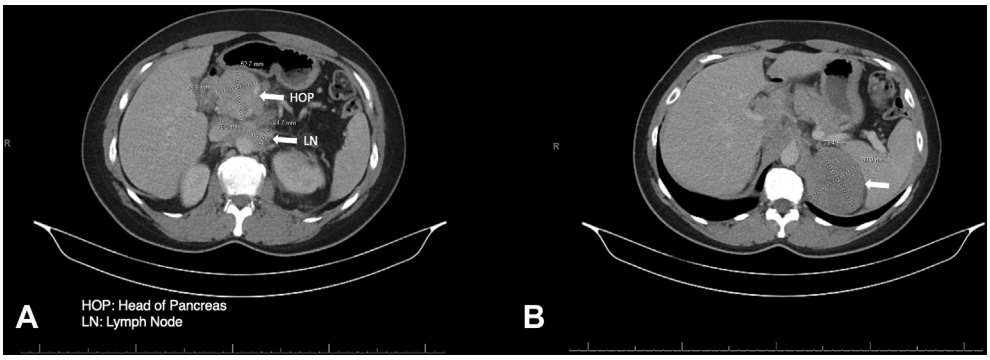

Figure 1. A, CT scan of the abdomen revealing a 4.2-cm mass in the pancreas and periaortic lymphadenopathy. **B,** CT scan showing a 6.7-cm mass in the splenic parenchyma.

A 46-year-old white woman with no medical history was referred for evaluation of intermittent epigastric pain. Her symptoms were not relieved by a 1-month trial of daily proton-pump inhibitors. A CT scan performed 1 week prior to referral revealed a 4.2-cm mass in the pancreatic head along with a 6.7-cm mass in the spleen with associated periportal and periaortic lymphadenopathy (Fig. 1A and B). EUS examination showed a 3.5- × 4-cm well-defined, hypoechoic mass with irregular borders in the uncinate process of the pancreas and a similar 6- × 5-cm round mass within the splenic parenchyma (Video 1, available online at www.videogie.org). An EUS-guided fine-needle biopsy of the pancreatic mass was performed using a 22-gauge Franseen-tip needle (Aquire; Boston Scientific, Marlborough, Mass, USA) without suction. On-site cytology showed neoplastic spindle cells and fibrotic stroma (Fig. 2A). Biopsy of the splenic lesion was then performed to rule out a different concomitant neoplastic process using another 22-gauge Franseen-tip needle (Fig. 2B). EUS-guided biopsy of the spleen has been successfully described in small series.[1,2] A careful Doppler examination of the mass and surrounding structures was done prior to the biopsy to look for intervening vessels. To lower the risk of bleeding and infection, the spleen capsule was punctured only once and normal parenchyma was avoided as much as possible. A meta-analysis of 62 patients has reported a pooled specimen adequacy of 92.8%, with most splenic lesions being lymphoma or lymphoproliferative diseases.[3] In our patient, immunohistochemical staining was positive for desmin, Ki67 (positive for 20% of cells) and smooth-muscle actin but negative for c-kit, DOG1, myogenin, CK20 and CD34; suggestive of leiomyosarcoma, a rare mesenchymal tumor, accounting for 0.1% of primary pancreatic nonislet neoplasms.[4] This neoplasia is highly aggressive and is generally diagnosed as a large, fibrotic mass within the pancreas. It can metastasize both by direct invasion of adjacent organs or to distant sites via hematogenous spread.[5] While 90 cases of primary leiomyosarcoma of the pancreas have been reported in the literature,[6] to our knowledge, this is the first case of a primary pancreatic leiomyosarcoma presenting with metastatic splenic involvement. There is no effective treatment other than complete surgical resection for small lesions.[7] The patient was referred to oncology for chemotherapy with doxorubicin but, given the lack of evidence that this regimen could benefit her, she opted for palliative care. The patient was lost to follow-up soon after.

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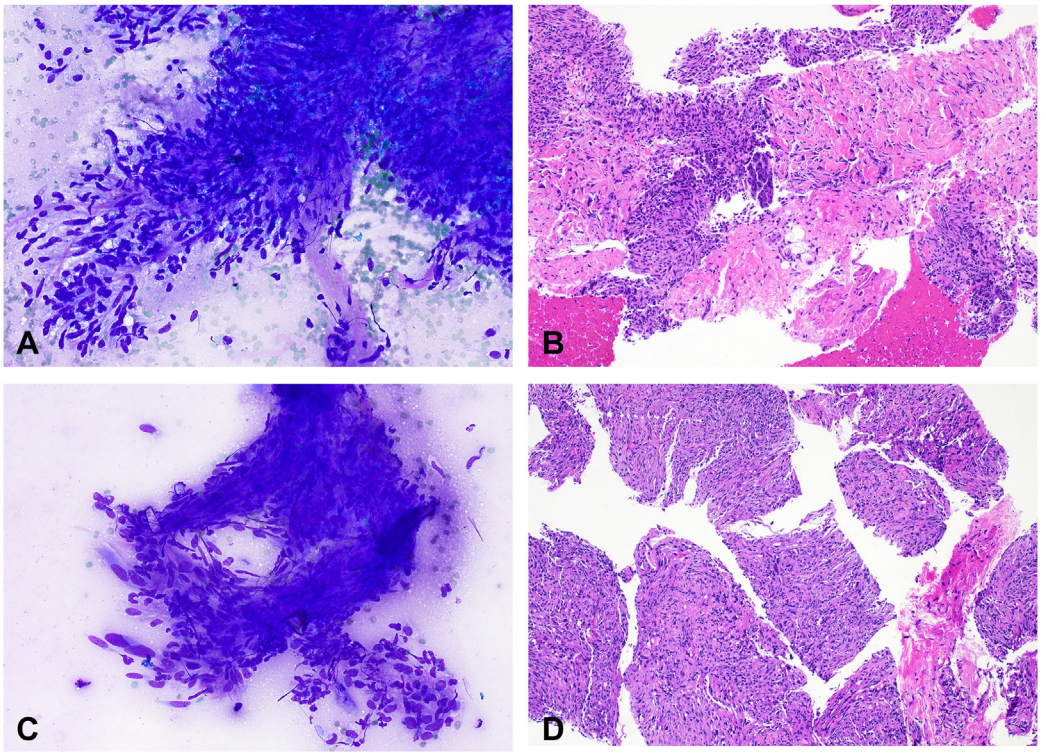

Figure 2. Rapid on-site evaluation cytology of the mass in the pancreas **(A)** and the spleen **(C)** showing neoplastic spindle cells. Histology of both sites **(B and D, respectively)** revealed smooth-muscle pattern (**[A and C]** stain: diff quick. orig. mag. ×20; **[B and D]** stain: H&E. orig. mag. × 10).

DISCLOSURE

Dr Varadarajulu is a consultant for Boston Scientific, Medtronic, and Olympus. All other authors disclosed no financial relationships.

DATA TRANSPARENCY

Individual participant data will not be shared.

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