Case History: *A 26 years-old female patient, who presented sudden lower abdominal pain. Gynecological status normal except lower left-sided abdominal tenderness upon palpation (normal delivery six months ago). Abdominal ultrasound showed a 15 cm cystic expansion that is attached to the pancreas and extends to left abdominal cavity. CA19-9 and CEA were within normal range. Computer tomography scan showed a 13x9x9 cm multicystic fluid containing retroperitoneal mass that is attached to the tail of the pancreas, which was interpreted as an unusual pseudocyst or lymfangioma/lymfangiomyoma. A cystic 20x13x10 cm retroperitoneal mass was removed three months later, which macroscopically showed sponge-like appearance (**Snapshot 1**). The specimen contained tail of the pancreas and spleen (280 g) that were attached to the tumor.*

Histologically there were dilated thin-walled channels of varying size, which were lined by flat endothelial-like cells that showed no atypia (**Virtual slide and Snapshot 2**). Occasionally clusters of lymphocytes were observed in the stroma between the channels and inside the lumina (**Snapshot 3**). In addition to the retroperitoneal fat, these channels were found in the peripancreatic fat right next to the acinar structures (**Snapshot 4**) and also attached to the splenic capsule.

Immunohistochemical staining showed strong and homogeneous positivity for CD31 (**Snapshot 5**) and strong but focal positivity for podoplanin (D2-40) (**Snapshot 6**). HMB-45 immunostaining was negative.

Diagnosis: Lymphangioma

Lymphangiomas are benign lymphatic-vessel like tumors. They occur most commonly in children and in cutaneous location. No clear gender predilection has been demonstrated. The pathogenesis of lymphangiomas is thought to be related most commonly to vascular malformation, but in some adult cases the underlying cause may be trauma-associated (lymphangiectasiae). Intra-abdominal lymphangiomas occur in all ages and can be extensive in size (1).

Differential diagnosis includes lymphangiomyoma(tosis), which shows HMB-45 positivity and smooth muscle proliferation in the walls of the lymphatic vessels. In addition, cystic mesothelial proliferations and myxoid liposarcoma can simulate lymphangioma, but these lesions do not show positivity for vascular markers in the luminal walls of the cavities (1).

Retroperitoneal (and pancreatic) lymphangiomas are rare benign tumors and less than 200 cases have been reported (2). They can clinically present as abdominal distension or pain, and even as acute abdomen (3). CT scan usually demonstrates the cystic lesions, but only one third were diagnosed conclusively in preoperative setting (4). Complete resection is recommended, in which case the prognosis is excellent (4, 5).

References:

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