Retroperitoneal fibrosis with a complex cystic lesion mimicking an inferior vena cava tumor

A 32-year-old woman presented with a dull aching abdominal pain of 2 months duration. She had similar complaints 18 months earlier and on evaluation, she was found to have a 4-cm cystic lesion near the head of the pancreas (Fig. 1). A repeat contrast enhanced computed tomography scan of the abdomen revealed a complex thick-walled cystic lesion measuring ~5 cm in size, located near the head of the pancreas and invading the medial wall of the inferior vena cava (Fig. 2). Subsequent 18-fluorodeoxyglucose positron emission tomography showed intense uptake in the lesion with a maximum standardized uptake value of 9.0 (Fig. 3). Endoscopic ultrasound (EUS) revealed a 5-cm complex septated cystic lesion in relation to the head of the pancreas abutting the inferior vena cava (Fig. 4). EUS guided aspiration of the cyst revealed a serosanguinous fluid that was rich in lymphocytes and no malignant cells were seen. The cyst was completely emptied and fluid cancer antigen 19-9, carcinoembryonic antigen, and triglyceride were within normal limits, whereas fluid amylase and lipase were elevated.

The patient underwent laparotomy and a 6-cm hard lesion arising from the infrahepatic inferior vena cava was observed. There was extensive desmoplastic reaction surrounding the lesion involving the aorta, and superior mesenteric artery. The lesion was removed and reconstruction of the inferior vena cava and left renal vein was performed using a polytetrafluoroethylene vascular graft. The biopsy from the resected specimen revealed a lymphoplasmacytic infiltrate with storiform fibrosis and obliteratorive phlebitis suggesting a diagnosis of idiopathic retroperitoneal fibrosis.

Retroperitoneal fibrosis is a rare entity characterized by nonspecific chronic inflammation of the retroperitoneum that can be caused by trauma, radiation or drugs such as methylsergide [1,2]. Treat-
ment is usually medical in the form of immunosuppression with or without surgical intervention depending upon the stage of the disease and the type of organ involved in the disease process [3]. Formation of a pseudocyst is an unusual complication of retroperitoneal fibrosis [4].

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References

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