Retroperitoneal Schwannoma Presents as a Peripancreatic Cystic Structure: The Utility of EUS/FNA in the Diagnosis

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Abstract

Retroperitoneal schwannomas are extremely rare tumors that arise from the Schwann cell of the peripheral nerves. Patients are typically asymptomatic, however, large tumors may lead to the development of abdominal pain or obstructive symptoms. Herein we describe a case of a 57 year-old male with a known peripancreatic cystic mass presenting with new onset abdominal pain and a CT demonstrating a 40 × 42 mm peripancreatic cystic structure previously 17 × 20 mm. Endoscopic ultrasound with fne needle aspiration was performed and cytological examination revealed a peripheral nerve sheath tumor. The patient underwent uneventful laparoscopic resection. Analysis of the surgical specimen revealed a spindle cell tumor with positive S100, negative CD117/CD34 and pancytokeratin consistent with a schwannoma.

Keywords: Schwannoma; Retroperitoneal; Peripancreatic; Endoscopic ultrasound; Fine needle aspiration.

Introduction

Retroperitoneal schwannoma (RS) is an extremely rare tumor arises from the Schwann cell of peripheral nerves. It represents 0.3-3% of the total schwannomas and 6% of all retroperitoneal tumors [1,2]. These lesions are usually found incidentally on different imaging modalities such as computerized tomography (CT), magnetic resonance image (MRI) or ultrasound (US). Herein, we are reporting a case of RS presenting as a peripancreatic cystic structure diagnosed by endoscopic ultrasound (EUS) with the utility of fine needle aspiration.

Case Report

A 57 year-old African American male with end stage renal disease status post kidney transplant who is known to have a peripancreatic cystic structure that was being monitored clinically presented with new onset abdominal pain. An abdominal CT scan was obtained which revealed a 40 × 42 mm.

Peripancreatic cystic lesion (Figure 1) previously 17 × 20 mm. EUS with FNA demonstrated a 39 × 38 mm well encapsulated cystic structure with solid components without definite communications with the pancreatic duct (Figure 2).

Figure 1: CT scan revealing a well-defned peripancreatic cystic lesion.

A 5 ml of blood tinged fluid was aspirated using a 22 gauge needle, and a subsequent fluid analysis demonstrated: CEA 1.0 ng/ml, Amylase 260 U/l and benign biological behavior on the molecular fluid analysis (Moderate DNA quantity and good DNA quality, negative GNAS point mutation, KRAS point mutation, and Tumor suppressor genes

Figure 2: Linear endoscopic ultrasound demonstrating a 39 x 38 mm well encapsulated peripancreatic structure without communication with the pancreatic duct.

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(LOH) point mutation). Cytological examination consistent with a nerve sheath tumor. A laparoscopic resection was performed which was followed by an uneventful post-operative course. The surgical specimen demonstrated a spindle cell tumor with mild to moderate cytological atypia without increase in mitosis. The tumor was positive S100, negative desmin, keratin, smoothmuscle actin, CD117 and CD 34. An MIB-1 proliferation index is approximately 6% (Figure 3). The histologic and immunophenotypic findings confirmed the schwannoma diagnosis.

Discussion

Gastrointestinal schwannomas are rare tumors that arise from the Schwann cell and represent less than 1% of all gastrointestinal neoplasm [3]. The stomach and colon are the most common sites (60-70% of cases) [3]. RS is an extremely rare benign tumor with a low malignant potential.

A 1.7% of all the reported RS are malignant [4]. They are mostly asymptomatic when found; however, larger tumors may cause abdominal pain or obstructive symptoms. RS presents in various forms; cystic (60%), partially cystic or solid thereby, are indistinguishable from pancreatic cystic neoplasms (i.e. IPMN or mucinous cyst) when they appear as a cystic lesion located within or adjacent to the pancreas [5]. Surgical removal is a reasonable option when symptomatic or if there is uncertainty about the diagnosis [2]. Preoperative diagnosis of the RS can be challenging especially when presenting as cystic structure located within or adjacent to the pancreas. While cross sectional imaging mostly delineates its boundaries, definitive diagnosis requires tissue sampling. IPMN, mucinous cyst, duplication cysts, and solid pseudopapillary tumors can present similarly, however, the presence of positive S 100 and negative CD34, negative CD117 and other immunohistochemistry stains support the schwannoma diagnosis and fairly exclude the others. In our case, the diagnosis was made using a curvilinear EUS with FNA however, the patient underwent surgical removal to rule out malignant potential due to the increased size of the lesion and new onset of abdominal pain. While diagnostic EUS/FNA is recommended for most pancreatic/peripancreatic cystic structures including schwannoma, surgical removal of RS to rule out malignant potential is yet to be proven [6].

Conclusion

RS are extremely rare tumors that arise from the Schwann cell of the sympathetic or parasympathetic nerves [4]. RS are seemingly important in the differential diagnosis of pancreatic/peripancreatic cystic lesions yet its preoperative diagnosis can be challenging. EUS is being utilized to further evaluate retroperitoneal cystic structure with the aim for fluid sampling and analysis and therefor it plays a major role in diagnosis of RS however, its utility to determine the malignant potential versus benign is unclear [7,8].

Figure 3: Surgical specimen demonstrating spindled cell proliferation with positive S100, negative pancytokeratin and CD117.

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