CASE REPORT

A Sporadic Desmoid Tumor: an Exceptional Pancreatic Cystic-Solid Mass

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Abstract Desmoid tumors are locally aggressive and non-metastatic neoplasms with a high rate of recurrence. Desmoid tumors of the pancreas are, however, very rare, and only a few cases have been reported in the literature. This paper reports an anecdotal case of a diffuse pancreatic desmoid tumor with the involvement of the pancreatic head, body, and—partially—tail. The patient underwent the Whipple procedure and subtotal pancreatectomy. Histopathological assessment showed that the tissues were partly positive for smooth muscle actin, but not for S100 or PanCK. The Ki67 index of the cells was only 1 %. Unfortunately, the patient died on the 10th postoperative day due to massive upper gastrointestinal bleeding.

Keywords Pancreatic tumor · Cystic-solid tumor · Desmoid tumor · Whipple procedure

Introduction

The desmoid tumor (DT) was first described in the early nineteenth century. The term was derived from the Greek word “Desmos”, describing a tendon-like attribute. DTs constitute 0.03 % of all neoplasms and 3 % of soft-tissue tumors with an annual incidence of around 2–4 cases/million/year in the general population [1]. These neoplasms can arise from all parts of the human body [2]. The intra-abdominal types are most often associated with familial adenomatous polyposis (FAP) or the Gardner syndrome (up to 70 % of the cases), while association with sporadic forms is uncommon [3]. Sporadic forms are regarded as non-traumatic or non-genetic related types. The first case of a pancreatic DT, reported in 1956 by Wilson, resembled a pseudocyst. Only 10 similar cases have been described in the English medical literature thus far [4].

Case Report

A 21-year-old female presented with epigastric pain of 40 days duration. On physical examination, there was mild epigastric tenderness, but no mass could be palpated. Transabdominal ultrasonography showed a cystic-solid mass in the head and body of the pancreas. Computerized tomography (CT) scan delineated the mass as a heterogeneous multi-lobular thick-walled cystic lesion (61×35 mm) with extension to the lesser sac (Fig. 1). Endoscopic ultrasonography (EUS) demonstrated a 6×4-cm cystic-solid mass in the pancreatic head, suspicious of mucinous cystadenoma. The large size of the mass per se was an indication for the surgical removal of the tumor, obviating the need for EUS-guided biopsy. Surgical finding was a large, firm, and whitish mass

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Fig. 1 A large cystic-solid mass in the pancreatic head and body (arrows)

located on the head, neck, and—partially—tail of the pancreas. The major vessels were intact, and the liver and spleen were unremarkable. The patient underwent the Whipple procedure, and owing to the involvement of the body, subtotal pancreatectomy was performed as well. Only the tail was preserved.

The specimen was an irregular, brown, rubbery, and firm mass, about 6 cm in diameter (Fig. 2a). Sliced sections of the tumor showed a few cystic areas containing a brown thick fluid with irregular inner walls. Histopathological examination revealed broad bundles of spindle cells and large amounts of collagen fibers, which had almost totally replaced the pancreatic tissue, including Langerhans' islands (Fig. 2b). The cystic areas had foci of degeneration and collections of foam cells, probably due to secondary pancreatitis. Immunohistochemistry (IHC) stains showed that the spindle cells were partly positive for smooth muscle actin, but not for S100 or PanCK. The Ki67 index of the cells was only 1 %. The tumor was diagnosed as a DT.

After an uneventful postoperative course until the 10th postoperative day, the patient suddenly suffered massive upper gastrointestinal bleeding and died despite efforts to control the bleeding.

Discussion

Approximately, 95 % of pancreatic tumors are primary cancers of exocrine origin, and the remainder arises from neuroendocrine cells. Pancreatic DTs are rare, with only 10 cases having been reported in the last 3 decades in the English medical literature [4]. Almost all the cases occurred in middle-aged patients [5]. In our patient, the tumor had almost completely replaced the pancreas, with only a small part of the tail remained tumor-free.

The risk factors of the DT include a previous history of abdominal surgery and the FAP or Gardner syndrome.

The incidence rate of sporadic intra-abdominal DTs is low (5 %), and fewer than 100 cases have been reported since 1960s [6]. Epigastric pain is the most common complaint, and it seldom radiates to the back, as is the case in pancreatic cancer. Loss of appetite can result in significant weight loss. Painless jaundice as a classic manifestation of pancreatic head cancer is rarely seen owing to the non-obstructing pattern of tumor growth [7].

Fig. 2 **a** Gross specimen of the pancreatic head and body. The cut section shows a glistening white tissue, typical of the desmoid tumor. **b** Histopathology of the desmoid tumor of the pancreas shows the

complete replacement of the pancreatic substance by collagen fibers and bundles of fibroblastic cells (H&E, $\times 100$)

Imaging studies have revealed that the pancreatic DT usually manifests as a well-defined solid or cystic-solid lesion (non-specific). The pancreatic DT expresses markers for fibroblasts like β -catenin, vimentin, and smooth muscle actin, but there are no expressions for stromal cell markers (CD117 and CD34) or neural cell markers (S-100). β -catenin IHC is believed to be useful for distinguishing deep DTs from other benign or malignant fibroblastic and myofibroblastic lesions [8].

According to the literature, most of the reported pancreatic DT patients survived and were disease-free at follow-up for up to 24 months. Unfortunately, our patient died following massive uncontrollable gastrointestinal bleeding on the 10th post-operative day.

Conclusion

Although the incidence of the sporadic DT of the pancreas is low, it should be taken into consideration in the differential diagnosis of patients suffering from a solid or solid-cystic mass of the pancreas.

Authors' Contribution This work was the collaboration between all authors. All authors read and approved the final manuscript.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no competing interests.

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