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RARE CASE PRESENTATION OF DERMOID CYST OF PANCREAS

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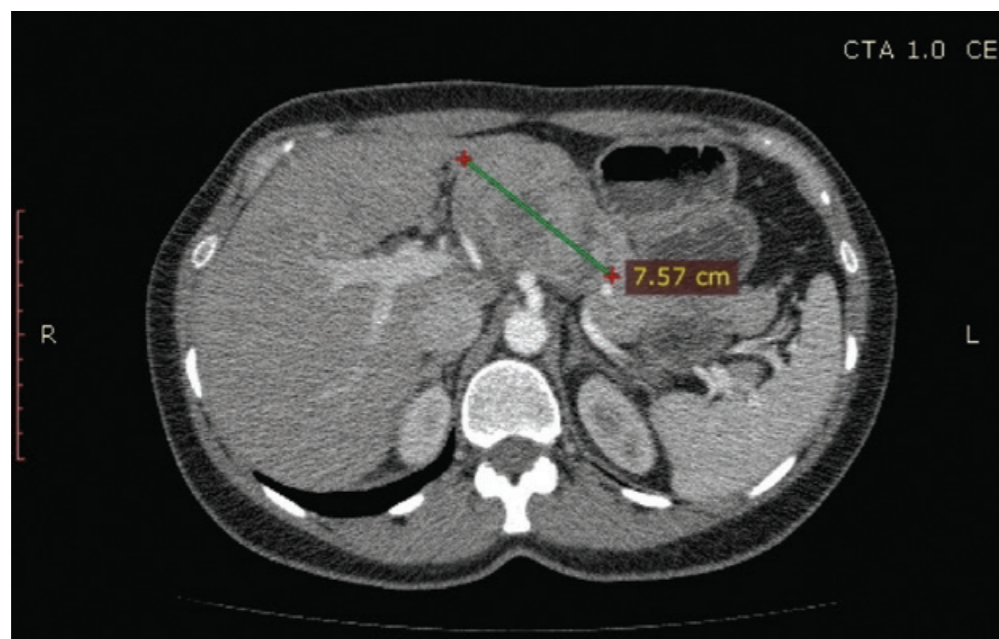
ABSTRACT

Teratomas are classified as either mature or immature, and according to tissue density can be classified to solid or cystic, also known as “dermoid cyst.” Immature teratomas are generally solid, histologically undifferentiated malignant tumors. Mature cystic teratomas (MCTs) in the pancreas are extremely rare with small number of cases reported in the literature. The most common location of the mass is in the head/ body of the pancreas. Diagnosis of teratomas is challenging, since there are no peculiar imaging findings, or characteristic serum markers. We present a case of a 42-years-old woman with a mature cystic teratoma in the head of the pancreas incidentally found at ultrasound imaging, who after surgical treatment remained asymptomatic. This case emphasizes the necessity of specific diagnostic tools and increased awareness about MCT, when a pancreatic cystic lesion is disclosed.

Key Words: *dermoid cyst, laparotomy, pancreatic cyst.*

Case Report

A 42-years-old female patient was referred to the ultrasound department due to mild back pain for several weeks. Medical history did not reveal any relevant diseases or habits. On thorough physical examination, her abdomen was soft and no palpable abdominal mass was present. The laboratory findings including serum tumor markers: carbohydrate antigen 19-9 (CA19-9) and carcinoembryonic antigen (CEA), were within normal range. Nonetheless, abdominal ultrasound disclosed well-defined retroperitoneal mass measuring 7.9 cm × 3.5 cm × 5.6 cm, adjacent to celiac trunk. The ultrasound appearance displayed mixed tumor pattern containing cystic and solid components, with absent vessels on color Doppler examination. Under the suspicion of pancreatic cystic tumor, contrast-enhanced CT scan was performed, revealing 7.5 × 4.6 cm bilobulated, homogeneously attenuating, low density retroperitoneal tumor (Figure 1).

Figure 1. Abdominal CT scan

She was referred to the surgical department and few weeks later, laparoscopic treatment was performed. The precise origin of the tumor was determined intraoperatively; the cyst was found to arise from the pancreatic head, adherent to the stomach, celiac trunk and retroperitoneum. Radical tumor excision was achieved, and operative specimen measuring $8.8 \times 4.3 \times 5$ cm revealed soft tissue mass encapsulated in a thin wall, containing yellow-whiteish material with a caseous appearance. Histological examination defined cystic lesion, lined by mature squamous epithelia with regular layer stratification, yet with different width. Beneath the surface, within the keratinous debris mixed sweat and adipose glands, rare hair follicles and multinuclear giant cells of “foreign body type” were present, suggesting mature cystic teratoma of pancreas. There was no evidence of malignancy. The final diagnosis was dermoid cyst of pancreas. Postoperative course was uneventful; patient was discharged 7 days after surgery. At 12 and 24-months follow-up, she was asymptomatic, in excellent condition, without any evidence of recurrence.

Discussion

Pancreatic dermoid cysts are exceptionally rare, about 50 cases have been reported in the literature since 1918, when the first case was published by Kerr and al., in 55 years old female patient (1). Pancreas is the slightest possible primary site of extragonadal teratomas. Diagnosis of teratomas is challenging due to absence of precise preoperative diagnostic tools or pathognomonic findings.

The most common physical finding is a palpable abdominal mass and/ or abdominal tenderness. Sometimes symptoms arise from the “mass effect” or pressure of the tumor lesion on neighboring organs/ tissues. Tumor usually arises in the head (44%) or body of the pancreas. Tail is on the third place by frequency, with only 12% (2). The lack of pathognomonic findings and insufficient diagnostic tests, make these pancreatic tumors hard to be recognized (3). An

imaging classification system has been proposed based on morphologic features of the lesion (4). Apart from patient’s clinical presentation, treatment decision is influenced by tumor size and location, tumor histological features, patient’s age and performance status and surgical risk.

Teratomas are germ cell tumors arising from a pluripotent stem cell, derived from at least two of the three germinative layers (ectoderm, endoderm and mesoderm). These tumors originate from some deviant germ cells (at the time of neural groove closure) that have been stopped during embryonal migration towards the gonads. Commonly teratomas are classified as mature, immature and specialized or monodermal. This classification is taking into account tissue components and tumor maturity. The nature of mature teratoma is benign, since its components are mature tissues, likewise immature teratoma comprises of undeveloped tissues that are mitotically active and generally considered as malignant. The third type of monodermal or specialized teratomas are usually subtype of mature teratomas. Further division is according to tissue density, either solid or cystic, later known as “dermoid cyst”.

Dermoid cysts, the most commonly develop within the ovaries and testis, however extragonadal localization in cranium, neck, mediastinum, omentum, retroperitoneum and sacrococcygeal region have been described (5,6).

Patients with pancreatic cyst may complain on nonspecific gastrointestinal symptoms such as abdominal pain, pain in the back, nausea, vomiting, weight loss, anorexia, fatigue, and fever (7). Laboratory findings are usually non-specific, unless the lesion obstruct the flow of biliary or pancreatic fluid. Serum levels of CEA and CA19-9, which are traditionally used for the evaluation of cystic neoplasms of the pancreas, are considerably low in patients with dermoid cysts, like in our case. Dermoid cysts of the pancreas are true cysts with wall lined by a single layer of keratinizing stratified squamous epithelium, while the underlying tissue content may have elements of adnexal tissue, sebaceous glands, lymphoid tissue, and even inflammatory cells (8).

The differential diagnoses of pancreatic cystic lesions include pseudocysts, benign cystic tumors like mucinous and serous cystadenoma, intraductal papillary mucinous neoplasm (IPMN), as well as neoplastic cysts and solid pseudopapillary tumor. Although ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) are helpful, findings are not usually pathognomonic (9,10). Mucinous cystic neoplasm (MCN) are true cystic neoplasms developed separate from the ductal system. Serous cystadenomas might be microcystic or macrocystic. Intraductal papillary mucinous neoplasm (IPMNs) composed from mucin-producing columnar cells and grosser than 10 mm have a malignant potential. The lesions show papillary proliferation, cyst formation, and varying degrees of cellular atypia (11). Solid pseudopapillary tumors are rare type of pancreatic lesions that are histologically characterized by the presence of degenerative pseudopapillary, loosely cohesive cells with grooved nuclei and aggregates of large hyaline globules (12). A completely different entity like lymphoepithelial cysts, the mostly located in the pancreas (tail and body, followed by the head and neck), relates dermoid cysts, and may be distinguished by the absence of hair follicles or sebaceous glands, which are more frequently

associated to the later one. Moreover, mucinous epithelium, hair follicles and sebaceous glands are found in dermoid cysts, rather than in lymphoepithelial or epidermoid cysts. One more particular feature in favor of dermoid cyst is presentation with suppurative infections, which is not so frequent in other “squamous lined” pancreatic cysts (13).

Final diagnosis of MCTs is established with histopathological evaluation, that includes complete sampling of the cystic wall. 7 – 10% of retroperitoneal teratomas have been reported to be malignant (14). Therefore, whenever a pancreatic cyst is intraoperatively suspected for being a teratoma, total resection is imperative (15). When the course of the disease is poor, or the tumor is inaccessible for total excision, partial excision and external drainage, can be performed (16).

Fortunately, in our case total excision of MCT was successfully and even more favorably, laparoscopically performed.

Conclusion

Dermoid cysts of pancreas are the mostly benign lesions. Macroscopically the tumors can be mobile or fixed, firm or cystic, and smooth or nodular. Only small percentage of mature teratomas may develop into malignant forms. The differential diagnosis includes all other cystic lesions of the pancreas, serous and mucinous cystadenomas, papillary cystic neoplasms, and pancreatic pseudocysts. Management of cystic pancreatic lesions depends of their nature and size; the patient in our case was appropriately referred to surgery for further treatment. We can conclude that “awareness of existence of cystic teratomas of pancreas” is the clue for the correct diagnosis of these tumors.

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Здружение на лекарите по
анестезија, реанимација
и интензивно лекување



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