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Dermoid cyst of the pancreas: a case report

Cisto dermoide de pâncreas: relato de caso

Carla Kellen da Silva Menezes[1]; Luciana Botinelly Mendonça Fujimoto[2]; Leonardo Simão Coelho Guimarães[3]; Silvana de Albuquerque Silva Damasceno[4]; Jenifer Morais de Melo[5]

ABSTRACT

Dermoid cysts or mature cystic teratomas are mesenchymal neoplasms most commonly found in the ovaries, but which may occur in any location along the pathways of ectodermal cell migration. They are rarely seen in the pancreas, where they show a slight preference for the pancreatic head. We report a case of dermoid cyst of the pancreas in a 69-year-old male patient, discussing the epidemiology, clinical presentation, diagnosis and treatment of this neoplasm. Since preoperative diagnosis is difficult, given its rarity in this site, it is usually diagnosed by histopathology of the specimen.

Key words: dermoid cyst; mature teratoma; pancreas.

INTRODUCTION

Dermoid cyst of the pancreas (mature cystic teratoma of the pancreas) is a very rare mesenchymal neoplasm, with 35 cases described in the world literature[7].

Dermoid cysts, in general, occur in all ages, have no sex preference, and are commonly found in ovaries, but may occur in any pathway of ectodermal cell migration. The pancreas is extremely rare as a primary site[1-5, 7, 8, 12, 13, 15], where there is a slight preference for the pancreatic head[3, 11, 15].

The clinical picture is non-specific[3, 4, 7, 8, 12, 13, 15], and the preoperative diagnosis is difficult. Many times it is suggested by imaging studies, but only confirmed by histological evaluation[9].

Macroscopically, the cyst generally presents a well-delimited and thick wall, with pasty yellow content of caseous aspect, rarely clear and serous. Microscopically, it contains differentiated elements from one or more germ layers[13].

The objective of this report is to present a case of dermoid cyst of the pancreas, located at the tail of pancreas, of difficult preoperative diagnosis, and to review epidemiological, clinical, diagnostic and histopathological aspects of this rare neoplasm.

CASE REPORT

A 69-year-old man, from Manaus, Amazonas, was admitted in December, 2010, to treat a urinary tract infection. A computed tomography (CT) of abdomen was ordered, for evaluation of renal lithiasis.

The CT revealed the presence of an expansive septated cystic lesion, of thicked walls and heterogeneous content in the tail of the pancreas, measuring 7.7 × 6.3 cm in its largest dimensions. The finding was considered not characteristic, maybe a pseudocyst with heterogeneous content, or even a pancreatic primary neoplastic process.

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1. Resident in Surgical Pathology at Hospital Universitário Getúlio Vargas (HUGV) of Universidade Federal do Amazonas (UFAM).

2. Doctorate in Biotechnology from UFAM; associate professor at the Department of Pathology and Legal Medicine of the Medical School of UFAM.

3. Specialist in Gastrointestinal Surgery; full professor of Colégio Brasileiro de Cirurgia Digestiva (TCBCD); gastrointestinal surgeon at HUGV-UFAM; professor of Clinical Surgery at Universidade do Estado do Amazonas (UEA).

4. Physician of the Department of Pathology and Legal Medicine of the Medicine School of UFAM.

5. Graduating student of Medicine at UFAM.

Other imaging and laboratory exams were performed.

The patient had a history of social drinking, ex-smoking, osteoarthrosis of knee and hip, eventually on use of nonsteroidal anti-inflammatory drugs. He referred a car accident 18 years before, with femur fracture and closed blunt abdominal trauma. He denied abdominal pain and previous pancreatitis. The physical examination was normal.

Laboratory investigation revealed serum dosages of amylase, lipase and tumor markers (carcinoembryonic antigen [CEA], carbohydrate antigen 19-9 [CA19-9], cancer-associated antigen 125 [CA 125] and alpha-fetoprotein) within normal ranges, as well as the other hematological and biochemical investigations.

In January, 2010, the patient underwent a new total abdominal CT, which revealed a lesion with the same characteristics as the previous one, but with a discrete increase, measuring 8.8 × 7 cm (**Figure 1**).

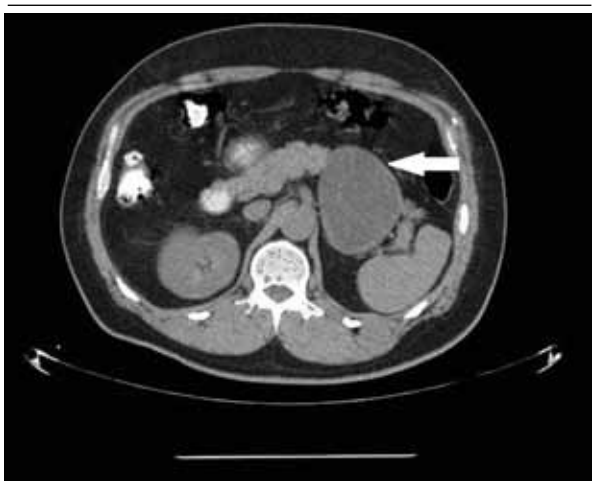

FIGURE 1 – *Total abdominal CT: hypodense expansive lesion of irregular contours, with internal septations, situated in the distal portion of the pancreatic body/tail, measuring 8.8 × 7 cm (APxT), with discrete parietal and septal enhancement (arrow)*

CT: computed tomography; APxT: anteroposterior × transverse diameters.

In March, 2010, a magnetic resonance imaging (MRI) was ordered. It demonstrated a multiloculated cystic lesion, in contact with the gastric wall; the pancreatic tail measured 7.2 × 6.4 × 7.3 cm.

No image showed invasion of adjacent organs, distance metastases or lymphadenopathy.

The possibilities of pseudocyst and malignant neoplasm were eliminated by the surgery team, and the suggested differential diagnoses were mucinous cystic neoplasm, solid pseudopapillary tumor (Frantz's tumor), non-functioning endocrine tumor with cystic degeneration, intrapapillary mucinous neoplasm and lymphoepithelial cyst of the pancreas.

The patient underwent laparoscopic enucleation of the lesion in June, 2010, and the material was sent for histopathological examination, which macroscopically revealed a thick cyst wall, with several micronodular formations on the surface, and abundant pasty homogeneous yellowish content (**Figure 2**).

FIGURE 2 – *Pancreatic dermoid cyst: thickened cystic wall, with several micronudulations on surface and pasty homogeneous yellowish content of caseous aspect*

Microscopic examination depicted cystic formations lined by keratinizing stratified squamous epithelium, with the presence of sebaceous glands, surrounded by mature lymphoid tissue, which contains lymphoid follicles and germinal centers. The cystic content was composed by laminated strands of keratin (**Figures 3** and **4**).

The patient had a favorable evolution, with no complications, being discharged two days after surgery. He remains without complaints.

DISCUSSION

The dermoid cyst of the pancreas, also called mature cystic teratoma of the pancreas, is an uncommon mesenchymal neoplasm, firstly described in 1918[4, 5, 10-13], with 35 cases reported in the world literature[7]. This is the 36th reported case up to now, according to a bibliographic survey.

It can affect all age groups, but according to Lane *et al.*[7], in the 35 reported cases, the mean age of patients was 36.4 years

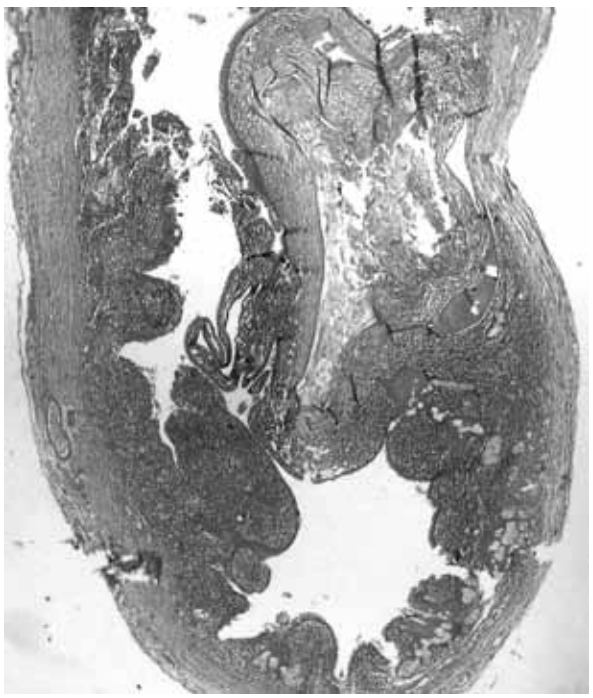

FIGURE 3 – *Pancreatic dermoid cyst (HE 4×): cystic formation lined by keratinizing stratified squamous epithelium, with the presence of sebaceous glands, surrounded by mature lymphoid tissue*

HE: hematoxylin and eosin.

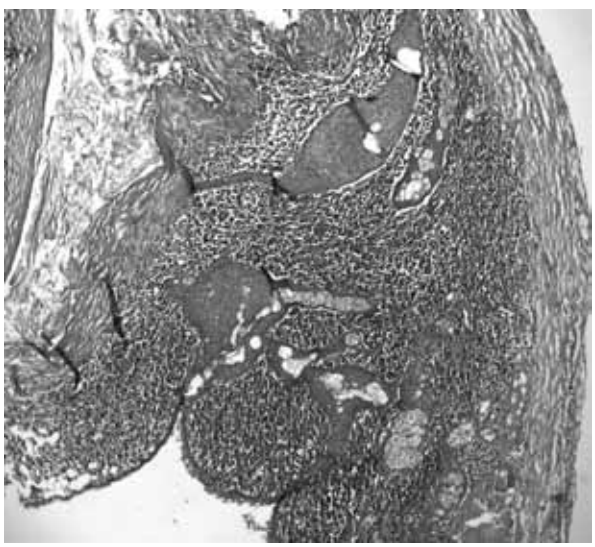

FIGURE 4 – *Pancreatic dermoid cyst (HE 20×): closer view of Figure 3*

HE: hematoxylin and eosin.

(ranging from 4 months to 74 years). Among patients, 20 were male and 15, female.

Degrate *et al.*[3], in a review of 33 cases, affirms that the average size of lesions was 7.5 cm (ranging from 2.2 to 20 cm), similarly to the observed in this case.

Dermoyd cysts are frequently found in the ovaries, but may occur in any pathway of ectodermal cell migration, typically in the midline, as in testes, skull, brain, mediastinum, retroperitoneum, omentum, and bladder. Occurrence in the pancreas, though, is very rare[1-5, 7, 8, 11-13, 15].

They may appear in any site in the pancreas, being lightly more common in the pancreas head[3, 7, 11, 15], what differs from the case presented here, in which the lesion was found in the pancreatic tail.

Clinical presentation is non specific. The most common signs and symptoms are palpable mass in the upper quadrant of the abdomen, abdominal pain, back pain, nausea, vomits, anorexia, weight loss, fatigue and fever. However, there are some cases with no symptoms (18.3%)[6], only incidentally discovered at diagnostic imaging[3, 4, 8, 12, 13, 15], as it happened in our study.

Preoperative diagnosis, due to the rarity of the lesion, is difficult[9], but it may be suspected after image exams, like ultrasonography (USG), CT, and ultrasound-guided fine-needle aspiration (FNA). The radiologic appearance of these lesion is variable, depending on the proportions of the diverse tissues of which they are composed[2-4, 12, 14].

There is also report of the conduction of endoscopic ultrasound-guided FNA[8], but there are no pathognomonic data to recognize a dermoyd cyst, and many times preoperative diagnosis is inconclusive[9, 11, 13], as well as in this case, in which the pancreatic dermoyd cyst was not a differential diagnosis according to the surgery team.

Daghfous *et al.*[2] performed a CT-guided FNA of a pancreatic mass whose content was sebaceous secretion, being the diagnosis of pancreatic teratoma suggested preoperatively. It was later confirmed as mature cystic teratoma of the pancreas, after distal pancreatectomy and histopathological examination, what suggests a good alternative for a preoperative diagnosis.

Tumor markers CEA and CA19-9 are generally lower than those in other pancreatic cystic neoplasms[13], but no consensus exists and further studying is necessary. In the present case, the patient presented normal levels of these and other markers, as previously cited.

The treatment is based on surgical removal, through either open surgery (distal pancreatectomy, cystectomy, external drainage and cystogastrectomy)[13] or laparoscopy, as in this case. This seems to be a good alternative for the treatment of benign pancreatic cyst lesions.

Dermoid cysts are true cysts; thus, their wall consists of stratified squamous epithelium and underlying connective

tissue[3, 5]. This characteristic is important to distinguish them from pancreatic pseudocysts, which correspond to 90% of cystic lesions of the pancreas and are collections of pancreatic secretions enclosed in a fibrous wall with no epithelium lining[6].

Macroscopically, the cyst is composed by a well-limited thick wall, which may contain micronodulations; its content is pasty pale yellow, of caseous aspect, as in this case. Rarely it may be light and serous[13] and sometimes may contain other elements, such as teeth, hair, bone, cartilage and skin appendices[3].

Microscopically, the cyst contains differentiated elements of one or more germ cell layers[13] as, for example, the cystic wall, formed commonly by a stratified squamous epithelium with the presence of sebaceous glands, and immediately adjacent, lymphoid tissue that may contain germinal centers. This layer of lymphoid tissue corresponds to the micronodulations observed in the cystic wall at macroscopy.

Frequently, there is the presence of mucin-producing ciliated columnar epithelium. Hair may also be found inside the cystic wall[11].

Yu *et al.*[14] reported a dermoid cyst of the pancreas in a 2-year-old child. The cystic wall contained smooth muscle, pancreatic tissue, lymphoid tissue, intestinal tissue and glial tissue.

The cystic content corresponds to the great quantity of keratin, sometimes disposed in laminated formation, possibly containing sebaceous secretion and cholesterol clefts[15]. There is the report of a carcinoid tumor arising in a mature pancreatic cystic teratoma[8].

Histopathologically, the differential diagnosis must include all cystic lesions of the pancreas: pseudocysts; neoplastic cysts, as the serous and mucinous cystadenoma; intraductal papillary mucinous neoplasm; and solid pseudopapillary tumor. However, one of the main differential diagnoses is the lymphoepithelial cyst, different from the dermoid cyst for the absence of skin annexes[7, 11, 13].

Although dermoid cysts are benign neoplasms, a small percentage may develop into malignant forms. Hence, a complete sampling of the lesion is necessary to exclude the presence of immature foci[3, 10, 11]. Up to now, all pancreatic teratomas reported were mature, that is, of benign behavior[3].

Pancreatic dermoid cysts are benign neoplasms, of good prognosis, and have been little reported in literature. They must be remembered during preoperative investigation, to enable a more conservative treatment, like the laparoscopic enucleation performed in this patient, which may be used as a good alternative. Besides, they seem to be of easy histopathological diagnosis, but may confound pathologists because they rarely present in this location.

RESUMO

Cistos dermoides ou teratomas císticos maduros são neoplasias mesenquimais comumente encontradas nos ovários, mas que podem ocorrer em qualquer via de migração das células ectodérmicas. No pâncreas, a ocorrência é rara, sendo mais comum na cabeça pancreática. Relata-se caso de cisto dermoide do pâncreas em paciente masculino de 69 anos, discutindo-se epidemiologia, clínica, diagnóstico e tratamento dessa neoplasia, pouco suspeitada no pré-operatório devido à sua raridade nessa topografia; geralmente, é diagnosticada apenas pelo exame histopatológico da peça cirúrgica.

Unitermos: *cisto dermoide; teratoma maduro; pâncreas.*

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MAILING ADDRESS

Carla Kellen da Silva Menezes
Rua 4, 19, apto 2; Residencial Luiza; Parque 10 de novembro; CEP: 69054-735; Manaus-AM, Brazil; e-mail: carlakmenezes@yahoo.com.br.