

Macrocytic serous cystadenoma of the pancreas in a young patient: case report

Abstract

Serous cystadenomas of the pancreas are rare cystic tumours. The microcystic form of serous cystadenoma is the most common and easy to diagnose radiologically. On the other hand, the macrocystic form of serous cystadenoma can be confused with other macrocystic lesions of the pancreas with malignant potential. The aim of this work is to highlight the limitations of paraclinical examinations in establishing a precise preoperative diagnosis. We report the case of a 34-year-old female patient admitted with abdominal pain and a unilocular macrocystic serous cystadenoma of the pancreas, diagnosed on histological examination of the operative specimen. Paraclinical examinations, including ultrasound, CT scan, MRI and echo-endoscopy, were inconclusive. The difficulty lies in preoperative diagnosis.

Key words: pancreas, serous cystadenoma, macrocyst

Introduction:

Cystic tumors of the pancreas represent around 10-15% of all pancreatic cysts (1) and around 1% of pancreatic cancers (2). PTCs comprise several entities, including mucinous and serous cystadenomas. In their classic form, serous cystadenomas (SC) of the pancreas are benign microcystic tumors (3),(5). However, oligocystic or macrocystic forms are even rarer and pose a diagnostic and therapeutic problem (2,3,6). The aim of this study is to report on the management of a young girl with a unilocular macrocystic serous cystadenoma and to review the literature.

Case Presentation

Patient, aged 34, presented with left hypochondrium pain of the heavy type in a context of general preservation, on clinical examination patient conscious 15/15 th, hemodynamically and respiratorily stable, on abdominal examination presence of epigastric and left hypochondrium tenderness, rectal examination without particularities. Abdominal CT showed a multiloculated cystic formation in the corporocaudal region of the pancreas measuring 11 cm, suggestive of a mucinous cystadenoma (figure 1). Abdominal MRI: large cystic mass of the body and tail of the pancreas, 10 cm in diameter, with fine septations suggestive of a serous cyst (figure 2). The procedure consisted in a corporocaudal pancreatectomy (figure 3). Anatomical pathology revealed a serous cystadenoma without malignancy.

cystic mass

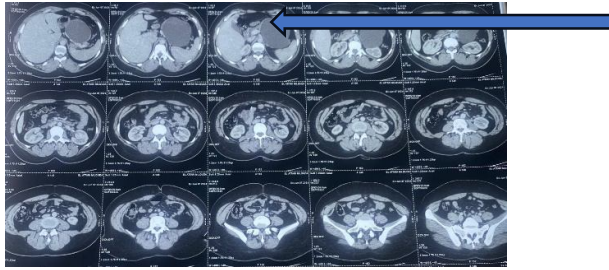


Figure 1: abdominal CT scan

Cystic mass



Figure 2: Abdominal MRI



Figure 3: surgical part

Discussion

Cystic lesions of the pancreas are divided into cystic tumors (CTT) and true cysts of the pancreas, with epithelium in 10-15% of cases, and pseudocysts (PC) without epithelial lining, accounting for almost 90% of cases (3),(5),(7),(8). The main histological types are CS, mucinous cystadenocarcinoma (CM),

solid and pseudopapillary tumors, and papillary and mucinous intracanal tumors. Macrocystic serous cystadenoma (MSC) is a rare entity, difficult to diagnose radiologically and echo-endoscopically, accounting for 6-10% of MSCs. It has a clear female predominance, reaching 80% of cases in some series (9). This female predominance might suggest the existence of a hormonal or genetic factor, but this lesion expresses neither estrogen nor progesterone receptors (10). They express cytokeratins AE1/AE3, 7, 8, 18, 19, EMA, NSE, Inhibin- α , MUC 1 and MUC 6 (9),(10),(11). These last four antibodies determine the serous nature of the lesion (12),(13). In its typical form, CS is found predominantly (75-86%) in women with an average age of 60-65 years (3,14). It is an asymptomatic tumour in 33% to 62% of cases, and may be discovered by chance (3),(15),(16). Abdominal pain is the most frequent clinical manifestation of macrocysticCS(7),(17),(18). In some cases, CS is revealed by compression of a neighbouring organ(3),(6),(7) In its classic form, CS presents a microcystic, multilocular, glycogen-rich architecture containing more than 6 cysts and measuring less than 2 cm, with a specific central zone that is sometimes calcified (3),(19),(14). In microcystic forms, diagnosis of CS is relatively straightforward on imaging. Macrocystic forms, however, pose a differential diagnostic challenge with PK, CM and sometimes other solid tumors (3),(19),(8),(20),(21). Indeed, PKs are ten times more frequent than other cystic lesions, and occur in a particular context suggestive of pancreatitis, alcoholism or trauma (3),(6),(8). Macrocystic CS of the pancreas is a rare benign tumor, which rarely degenerates, and may be confused clinically and radiologically with CM with malignant potential (3),(6),(15),(21). The ultrasonographic or CT presentation is often characteristic, in the form of a well-limited, honeycomb-shaped multilocular mass, approximately 25 mm in long axis, occasionally (10-15% of cases) centered by a central, stellate, focally calcified scar. Hypervascularization of the lesion is revealed by contrast injection (22). Imaging is usually sufficient to confirm the diagnosis; cytological and/or histological confirmation is required in only 19% of cases (22). In such cases, the pathologist can analyze a sample under echo-endoscopic guidance, as in our two observations, where precise typing was not possible. According to Belsley et al., serous epithelial cells are found and identified in only 21% of cases (22). Serous cystadenoma may share great morphological similarities with a well-differentiated neuroendocrine tumor of the pancreas, but the negativity of Synaptophysin and Chromogranin A in serous cystadenoma makes it possible to distinguish between these two entities (23),(24). These tumors are considered benign, with an excellent prognosis (9). The only criterion for malignancy is the presence of extra-pancreatic metastases, most often in the liver, leading to reclassification as cystadenocarcinoma. Vascular or peri-nerve invasion, or local invasion of the duodenum or stomach, are not considered malignant criteria (9),(25). Thus, active surveillance may be proposed to the patient, if all the following criteria are met (22),(26):

- typical radio-clinical presentation
- slow progression
- an asymptomatic mass measuring less than 4 cm
- absence of diffuse pancreatic lesions.

The risk of death is mainly due to postoperative complications (9).

Conclusion:

Unilocular macrocystic serous cystadenoma is a rare, benign TKP. It poses a preoperative diagnostic problem, as it can be confused with cysts with a high malignant potential. Surgical resection is always indicated in the case of a symptomatic patient or when there is diagnostic doubt. Conservative

treatment can only be undertaken in a patient with asymptomatic TKP in a setting with appropriate clinical, biological and morphological monitoring facilities.

Ethical Approval:

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

Consent

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

Disclaimer (Artificial intelligence)

Option 1:

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

Option 2:

Author(s) hereby declare that generative AI technologies such as Large Language Models, etc. have been used during the writing or editing of manuscripts. This explanation will include the name, version, model, and source of the generative AI technology and as well as all input prompts provided to the generative AI technology

Details of the AI usage are given below:

- 1.
- 2.
- 3.

Références :

1. Fujiwara H, Ajiki T, Fukuoka K, Mitsutsuji M, Yamamoto M, Kuroda Y. Macrocystic serous cystadenoma of the pancreas. J Hepatobiliary PancreatSurg 2000; 7: 92-6.
2. Le Borgne J, de Calan L, Partansky C. Cystadenomas and cystadenocarcinomas of the pancreas: a multiinstitutional retrospective study of 398 cases. Ann Surg 1999; 230: 152-61.
3. Farrell JJ : Prevalence, Diagnosis and Management of Pancreatic Cystic Neoplasms : Current Status and Future Directions. Gut Liver 2015 ; 9 : 571-89

4. Del Chiaro M, Verbeke C, Salvia R et al. : European experts consensus statement on cystic tumours of the pancreas. *Dig Liver Dis* 2013 ; 45 : 703-11
5. Chatelain D, Hammel P, O'Toole D et al. : Macrocystic form of serous pancreatic cystadenoma. *Am J Gastroenterol* 2002 ; 97: 2566-71
6. Jais B, Rebours V, Malleo G et al. : Pancreatic serous cystadenoma related mortality is almost nil. *Pancreatology* 2014 ; 14 : S4
7. Muscari F, Suc B, Escat J, Fourtanier G : Les tumeurs kystiques du pancréas. *J Chir (Paris)* 2002 ; 139 : 312-23
8. Bosman F.T, Carneiro F, Hruban R.H, Theise N.D. IARC WHO classification of the Tumors of the Digestive system. 4th edition, 2010.
9. Yasuhara Y, Sakaida N, Uemura Y, Senzaki H, Shikata N, Tsubura A. Serous microcystic adenoma (glycogen-rich cystadenoma) of the pancreas: study of 11 cases showing clinicopathological and immunohistochemical correlations. *Pathol Int.* 2002;52:307-12
10. Salomao M, Remotti H, Allendorf JD, Ponerros JM, Sethi A, Gonda TA, et al. Fine-needle aspirations of pancreatic serous cystadenomas: improving diagnostic yield with cell blocks and α inhibin immunohistochemistry. *Cancer Cytopathol.* 2014;122:33-39.
11. Shah L, Tiesi G, Bamboat Z, McCain D, Siegel A, Mannion C. Tumor-to-tumor metastasis: report of two cases of renal cell carcinoma metastasizing to microcystic serous cystadenoma of the pancreas. *Int J SurgPathol.* 2015;23:48-51.
12. Kosmahl M, Wagner J, Peters K, Sipos B, Klöppel G. Serous cystic neoplasms of the pancreas: an immunohistochemical analysis revealing alpha-inhibin, neuron-specific enolase, and MUC6 as new markers. *Am J SurgPathol.* 2004;28:339-46
13. Wargo JA, Fernandez-del-Castillo C, Warshaw AL : Management of Pancreatic Serous Cystadenomas. *Adv Surg*2009 ; 43 : 23-34
14. Colonna J, Plaza JA, Frankel WL, Yearsley M, Bloomston M, Marsh WL : Serous Cystadenoma of the Pancreas : Clinical and Pathological Features in 33 Patients. *Pancreatology* 2008 ; 8 : 135-41
15. Blaye-Felice S, Cabarrou B, Alassiri A et al. : Kystes du pancréas opérés : concordance entre diagnostic préopératoire et résultat anatomopathologique. *J ChirVisc*2015 ; 152 : A29
16. Cunha JEM, Perini MV, Siqueira SAC et al. : Serous oligocystic adenoma of the pancreas. *Pancreatology* 2003 ; 3 : 482-6
17. Ayadi-Kaddour A, Goutallier-Ben Fadhel C, Rezgui L, Lahmar-Boufaroua A, Khalfallah MT, Mzabi-Regaya S : Le cystadénome séreux macrokystique uniloculaire du pancréas : une variante morphologique à connaître. *Ann Chir* 2003 ; 128 : 177-9
18. Carpizo DR, Allen PJ, Brennan MF : Current management of cystic neoplasms of the pancreas. *The Surgeon* 2008 ; 6 : 298-307
19. Wang Y, Guo K, Zhao M, Song S, Xu Y, Ma G : [Macrocystic serous adenoma of the pancreas : a report of 5 cases]. *Zhonghua Wai Ke Za Zhi* 2010 ; 48 : 1405-8
20. Pravisani R, Intini SG, Girometti R et al. : Macrocystic serous cystadenoma of the pancreas : Report of 4 cases. *Int J Surg* 2015 ; 21 : S98-101
21. Belsley NA, Pitman MB, Lauwers GY, Brugge WR, Deshpande V. Serous cystadenoma of the pancreas: limitations and pitfalls of endoscopic ultrasound-guided fine-needle aspiration biopsy. *Cancer.* 2008;114:102-110
22. Hruban RH, Pitman MB, Klimstra DS. Atlas of Tumor Pathology. Washington, DC: American Registry of Pathology and Armed Forces Institute of Pathology; 2007. Tumors of the pancreas.

23. Mohr VH, Vortmeyer AO, Zhuang Z, Libutti SK, Walther MM, Choyke PL et al. Histopathology and molecular genetics of multiple cysts and microcystic (serous) adenomas of the pancreas in von Hippel-Lindau patients. *Am J Pathol.* 2000;157:1615-1621
24. Brandonea N, Poizata F, Thomassin-Piana J, Turrinib O, Popovicic C, Terris B. Un cystadénocarcinome séreux du pancréas dans une variante solide. *Annales de Pathologie.* 2016;36:125-132
25. Lombardo C, Iacopi S, Menonna F, Napoli N, Kauffmann E, Bernardini J, et al. Pancreatic resection in patients with asymptomatic serous cystadenoma. *Pancreatology.* 2018;18:577-584

UNDER PEER REVIEW