Diagnosis, morphopathological profile and treatment of mucinous cystadenoma of the pancreas – a single center experience

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Abstract

Aim: Pancreatic mucinous cystadenoma (MCA) occurs almost exclusively in perimenopausal women and represents between 10% and 45% of cystic neoplasm of the pancreas, being considered a premalignant lesion. Materials and Methods: From 1983 to 2017, 31 patients underwent surgery for MCA of the pancreas in our Center. The median age was 47 years (range 17–81 years). All data were obtained retrospectively. Results: The female/male gender ratio was 14.5/1. Most of the patients (90.3%) were symptomatic. The most common manifestation was non-specific abdominal pain (58.06%), followed by fatigue and vomiting. The median cyst size was 7 cm, with a range between 2 cm and 15 cm. There were 35 procedures in 31 patients (in four patients the resection was preceded by a drainage procedure). From the 28 resections, most of them (89.28%) were performed by an open approach; a minimal invasive approach was used in three patients (robotic – two; laparoscopic – one). Most of the resections (82.14%) were distal pancreatectomies. In all cases, the final diagnosis was based on histological examination that revealed columnar epithelium and ovarian-type stroma. Postoperative complications occurred in 10 (34.48%) patients. Postoperative mortality was 3.44% (one patient) by septic shock secondary to acute postoperative pancreatitis. Conclusions: MCAs represent a rare pancreatic pathology with challenging diagnostic and therapeutic implications. Multi-detector computed tomography (MDCT) scan, endoscopic ultrasound (EUS) and magnetic resonance imaging (MRI)/magnetic resonance cholangiopancreatography (MRCP) are useful in the differential diagnosis with other pancreatic fluid collections and treatment. Oncological surgical resections are recommended. Histopathological examination establishes the final diagnosis. The most common postoperative complication is pancreatic fistula.

Keywords: pancreatic mucinous cystadenoma, surgery, pancreatic cyst.

Introduction

The mucinous cystadenoma (MCA) of the pancreas was recognized for the first time as a separate pancreatic cystic entity by Compagno & Oertel in 1978 [1]. MCAs are usually oligo- or paucilocular and are lined by a cylindrical epithelium and a stroma similar to that of the ovary [2]. MCA occurs almost exclusively in perimenopausal women, female/male (F/M) gender ratio being 20:1 [3] and represents between 10% and 45% of cystic neoplasm of the pancreas [4]. Unless there is a fistula, mucinous cyst generally do not communicate clearly with pancreatic ductal system. Recent studies have shown, however, the existence of microscopic communications demonstrated by injecting dye into the pancreatic ducts [5]. In many cases, MCA tissue architecture can be modified by the presence of cellular atypia. Often uniformly tissue areas with minimal atypia may be interspersed with areas of profound change in appearance.

Despite being a rare lesion, proper and early diagnosis is important, since mucinous cystic lesions of the pancreas are considered to have a high malignant potential [1, 6]. According to the 2010 World Health Organization (WHO) classification of cystic pancreatic tumors, MCAs are considered premalignant lesions [7]. Consequently, oncological surgery is recommended in all the cases. Surgical resection is curative in benign MCAs.

The aim of the present study is to evaluate and compare the diagnostic peculiarities and treatment options in a single center series. To our knowledge, this is the largest series reported in Romania.

Materials and Methods

From 1983 to 2017, 31 patients underwent surgery for MCA of the pancreas in the Center of General Surgery and Liver Transplantation from the “Fundeni” Clinical Institute (Bucharest).

All data were obtained via retrospective review of the patients’ clinical, imagistic and pathology reports. Due to this long period, this series was not homogeneous with respect to diagnosis and management.

MCAs were diagnosed preoperatively by imaging criteria (single or oligoloculated cyst without ductal communication) but the final diagnosis was based on histological examination of the resected specimen (columnar epithelium and ovarian-like stroma). Tissue samples were processed following classical histological techniques (fixation in buffered formalin and paraffin embedment).
Multiple sections were performed in each specimen, in order to identify with high-grade dysplasia MCAs or invasive mucinous cystadenocarcinoma. Thus, from an initial group of 48 patients with cystic pancreatic tumors, 17 were excluded for such lesions.

Serial sections were performed and stained first with Hematoxylin–Eosin (HE) and then immunomarked with cytokeratin (CK) 7 to identify glandular nature of the cyst epithelium, carbohydrate antigen (CA) 19-9 to identify glycoproteins specific to glandular epithelium, Muc5Ac to detect mucin 5Ac of human origin, Muc2 to identify the goblet cells and Ki-67 to assess the benignity of tumors in all cases. The characteristics of all used antibodies are presented in Table 1.

Table 1 – Antibodies used to assess the MCA

<table>
<thead>
<tr>
<th>Antibody</th>
<th>Clone</th>
<th>Source</th>
<th>Dilution</th>
</tr>
</thead>
<tbody>
<tr>
<td>CK7</td>
<td>OV-TL 12/30</td>
<td>DAKO</td>
<td>1:300</td>
</tr>
<tr>
<td>Mouse anti-CA19-9 Ab</td>
<td>116-NS-19-9 Life Technologies</td>
<td>1:100</td>
<td></td>
</tr>
<tr>
<td>Muc5Ac</td>
<td>CLH2</td>
<td>Santa Cruz</td>
<td>1:100</td>
</tr>
<tr>
<td>Muc2</td>
<td>SPM512</td>
<td>ABCAM</td>
<td>1:300</td>
</tr>
<tr>
<td>Mouse anti-Ki-67 Ab</td>
<td>MIB-1</td>
<td>DAKO</td>
<td>1:10</td>
</tr>
</tbody>
</table>

MCA: Mucinous cystadenoma; CK: Cytokeratin; CA: Carbohydrate antigen; Ab: antibody; Muc: Mucin.

**Results**

**Gender and age**

The great majority of the identified lesions was present in females (29 cases – 93.54%). The F/M ratio was 14.5/1. The patients’ age varied in a wide range, between 17 and 81 years. Most of cases were however grouped around the mean age of 47 years in a range determined by a standard deviation of 16, range pushed slightly to the lower limit of the whole age variation interval (Figure 1).

**Symptomatology**

MCAs were asymptomatic and therefore discovered incidentally in three patients. Even large cysts may be asymptomatic. For instance, we had a 63-year-old female patient with a 56/48/46 mm MCA, discovered at the level pancreas head during a periodical abdominal computed tomography (CT) scan performed four years following rectal cancer surgery.

However, most of the patients (28 – 90.3%) were symptomatic. The most common clinical manifestation was non-specific abdominal pain (18 patients – 58.06%), followed by fatigue and vomiting.

**Imaging investigation**

Most of the cases were identified by abdominal ultrasound (US), which provide information about the location, size and the relation of the cysts to the neighboring structures. However, accurate data on location, size and even the type of the cyst were provided by CT and magnetic resonance imaging (MRI) scan.

The preferred imaging method was contrast-enhanced CT scan. The median cyst size, assessed by CT scan, was 7 cm, with a range between 2 and 15 cm. Wall calcifications were described in a single patient (Figure 2a), but intracystic calcifications were also noted. CT examination did not visualize any communication between the cyst and the pancreatic duct to any of the patients. Few patients were assessed by MRI. In one of these cases, for instance, axial MRI and consecutive three-dimensional magnetic resonance cholangiopancreatography (3D-MRCP) reconstruction revealed, in a 48-year-old female patient, an 8 cm MCA of the pancreas body and tail with no communication with the main pancreatic duct (Figure 2, b and c). Subsequently, the tumor was resected by open spleno-pancreatectomy (SP).
Serum tumor markers

Serum tumor markers analysis was available only in six patients, three of them having increased CA19-9 levels (between 255 and 2040 IU/mL).

Morphological assessment

**Gross aspect**

Tumors were located everywhere in the pancreas but most of them, meaning slightly more than 80% were located in the body, the tail or both terminal segments of the pancreas (Figure 3a).

The cyst size, measured on the surgical sample, confirmed the preoperative imaging assessment (see below).

Two-thirds of the cysts had a medium size, varying between 5 cm and 10 cm. However, only five cysts, representing less than 20% of the cases, had more than 10 cm, as reported in the literature (Figure 3, c and d). Moreover, almost 20% of investigated MCAs were small cysts with than 5 cm in diameter (Figures 3 d).

The gross aspect of almost two-thirds of cases was uniloculated, the remaining cysts showing a pauciloculated structure. In Figure 4a, is presented such a large, uniloculated MCA from a spleno-pancreatic resection.

The cysts had a mucinous content (Figure 4b) in almost all cases (28 patients – 90.32%). Hemorrhagic cysts (Figure 4, a and c) were found in two (6.45%) patients and one cyst (3.22%) presented as an abscess (Figure 4d).
Histopathological assessment

In almost all of the cases (29 cases, meaning 93.54%), the cysts walls were covered by a columnar epithelium. However, in two cases, two types of epithelium were observed (Figure 5a) – columnar (black arrows) and cuboidal (red arrows). Some tumors were largely bland with minimal architectural complexity of the epithelium, containing mostly uniform, basally oriented nuclei; other tumors revealed, in turn, a papillary architectural pattern, with pseudostratified hyperchromatic nuclei (Figure 5b – blue arrows). The epithelium expressed diffusely keratin, as well as glycoprotein markers, such as CA19-9. Muc5AC had also a diffuse expression (Figure 5, c–e). In areas with papillary architecture, intestinal-type scattered goblet cells were identified and confirmed by Muc2 staining (Figure 5f). The key morphological feature for the diagnosis of MCA was, however, the presence in all examined cysts of a distinctive subepithelial hypercellular spindle cell stroma of ovarian type, considered as a consistent feature of these tumors (Figure 5, a and b – yellow thick arrows).

Low-grade dysplasia has been described in only three (9.67%) cases, the remaining having a benign aspect. Fresh frozen pathological examinations were performed in three cases, showing a typical MCA appearance. Ki-67 expression was significantly low in all cases (Figure 5g), confirming the benign character of all studied tumors.

Intraoperative assessment

Usually, the diagnosis of an MCA was established preoperatively. However, in six cases, the pre- and intraoperative aspect of the cyst mimicked a pancreatic pseudo-cyst (PP) and a drainage procedure was performed consequently. After definitive diagnosis was established, four of them underwent surgical resection, two being lost from follow-up.

Surgical treatment

There were 35 procedures in 31 patients (in four patients, the resection was preceded by a drainage procedure, as shown in Table 2). Most of the 28 resections (25 cases, meaning 89.28%) were performed by an open approach, the minimal invasive approach being used in three patients: in two of them, robotic surgery was the choice and laparoscopic technique
In the great majority of cases, the resections were distal pancreatectomies including spleen removal (23 cases, meaning 82.14%). In an 81-year-old patient with a large painful cyst, a cysto-jejuno-stoma was performed as palliation. In three cases with large cysts and adjacent inflammation, complex resections were required, involving the stomach, small or large bowel.

### Table 2 – Types of surgical procedures

<table>
<thead>
<tr>
<th>Procedures</th>
<th>Intervention type</th>
<th>No. of cases</th>
<th>Previous procedures (PP)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Open approach</td>
<td>17</td>
<td>No PP 14</td>
</tr>
<tr>
<td></td>
<td>Spleno-pancreatectomy (SP)</td>
<td></td>
<td>CJA 2</td>
</tr>
<tr>
<td></td>
<td>Spleno-preserving distal</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pancreatectomy</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Pancreatoduodenectomy</td>
<td>2</td>
<td>1 ED</td>
</tr>
<tr>
<td></td>
<td>Simple resection</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Robotic (R)</td>
<td>2</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Laparoscopic (L)</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td><strong>Resections</strong></td>
<td>Cysto-jejunostomy (CJA)</td>
<td>1</td>
<td></td>
</tr>
<tr>
<td><strong>Palliative surgery</strong></td>
<td>Cysto-gastrostomy (CGA)</td>
<td>1 lost from follow-up</td>
<td></td>
</tr>
<tr>
<td><strong>Only pre-mucinous</strong></td>
<td>External drainage (ED)</td>
<td>1 lost from follow-up</td>
<td></td>
</tr>
</tbody>
</table>

### Outcome

The average postoperative stay was 19.7 days.

Postoperative complications occurred in 10 of the 29 patients (lost from follow-up patients were excluded), all females, representing almost one-third of the studied cases (Table 3). Most of these complications appeared after spleno-pancreatectomies (seven of the 10 cases).

The pancreatic fistula (PF) was the most frequent complication, occurring in half of these cases. In four patients, it occurred after spleno-pancreatectomies. All patients with PF were conservatively treated and had favorable outcome.

Acute post-operative pancreatitis occurred in two patients. In one case, that was conservatively treated, it had favorable course, while, in the second case, acute pancreatitis and massive upper gastrointestinal (GI) bleeding occurred and emergency surgery was performed but with a poor outcome.

Infectious complications were present in two patients. In one case, the patient developed initially pneumonia followed by septic shock. Secondary to the aggressive antibiotic therapy, *Clostridium difficile* infection occurred, but with favorable outcome. The second case, already mentioned above, consisted in septic shock occurrence after emergency surgery for pancreatitis and GI bleeding.

Renal failure appeared in only one patient and was successfully treated by dialysis.

The overall outcome was favorable, postoperative mortality being of only 3.44%, meaning only one patient, the one with septic shock secondary to acute postoperative pancreatitis.

### Table 3 – Postoperative complications

<table>
<thead>
<tr>
<th>Gender</th>
<th>Age [years]</th>
<th>Location</th>
<th>Surgical procedure</th>
<th>Complication</th>
<th>Management</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>F</td>
<td>63</td>
<td>Head</td>
<td>PD</td>
<td>Pancreatic fistula</td>
<td>Conservative</td>
<td>Favorable</td>
</tr>
<tr>
<td>F</td>
<td>26</td>
<td>B</td>
<td>CP</td>
<td>Distal stump pancreatitis</td>
<td>Conservative</td>
<td>Favorable</td>
</tr>
<tr>
<td>F</td>
<td>26</td>
<td>T</td>
<td>CGA</td>
<td>Upper GI bleeding and acute pancreatitis</td>
<td>SP and gastric resection</td>
<td>Septic shock – demise</td>
</tr>
<tr>
<td>F</td>
<td>58</td>
<td>T</td>
<td>SP</td>
<td>Left subphrenic hematomat</td>
<td>Surgical haemostasis</td>
<td>Favorable</td>
</tr>
<tr>
<td>F</td>
<td>61</td>
<td>T</td>
<td>SP</td>
<td>Acute renal failure</td>
<td>Dialysis</td>
<td>Favorable</td>
</tr>
<tr>
<td>F</td>
<td>68</td>
<td>T</td>
<td>SP (R)</td>
<td>Clostridium difficile infection, pneumonia with septic shock</td>
<td>Conservative (ICU)</td>
<td>Favorable</td>
</tr>
<tr>
<td>F</td>
<td>22</td>
<td>B and T</td>
<td>SP</td>
<td>Pancreatic fistula</td>
<td>Conservative</td>
<td>Favorable</td>
</tr>
<tr>
<td>F</td>
<td>44</td>
<td>B and T</td>
<td>SP</td>
<td>Pancreatic fistula</td>
<td>Conservative</td>
<td>Favorable</td>
</tr>
<tr>
<td>F</td>
<td>53</td>
<td>B and T</td>
<td>SP</td>
<td>Pancreatic fistula</td>
<td>Conservative</td>
<td>Favorable</td>
</tr>
<tr>
<td>F</td>
<td>58</td>
<td>B and T</td>
<td>SP</td>
<td>Pancreatic fistula and ascites</td>
<td>Conservative</td>
<td>Favorable</td>
</tr>
</tbody>
</table>

F: Female; B: Body; T: Tail; PD: Pancreatoduodenectomy; CP: Central pancreatectomy; CGA: Cysto-gastrostomy; SP: Spleno-pancreatectomy; R: Robotic; GI: Gastrointestinal; ICU: Intensive Care Unit.

A case that is worth mentioning is that of a 26-year-old female patient who was diagnosed in 1986 in another medical unit with a 6 cm pancreatic cyst, during a pregnancy follow-up. CT scan was suggestive of a pancreatic pseudocyst. After three years, the patient was reassessed in our Clinic. An initial US examination discovered some calcifications, misinterpreted as left kidney stones. However, the CT scan revealed that they belonged to a fluid collection of the tail of the pancreas of 9 cm diameter (Figure 6a).

Since the intraoperative aspect was also suggestive for a PP with pericystic chronic inflammation, thick wall, brown content on puncture, necrotic debris and a necrotic area found on the posterior gastric wall, a transgastric cystogastrostomy was performed. The patient developed, postoperatively, acute pancreatitis and intracystic bleeding.
requiring spleno-pancreatectomy and partial gastric resection. Inspection of the cyst revealed its multi-loculated appearance, wall papillary projections and a mucinous content.

The histological examination revealed a papillary architecture of the lining epithelium, with underlying ovarian like stroma in the cystic wall but with an epithelial low Ki-67 index (Figure 6b), pleading for a benign MCA.

Subsequently, the patient was operated again for recurrent upper GI bleeding, dehiscence of the gastric suture and peritonitis (total gastrectomy with esojunostomy was required). Septic shock and death occurred at 20 days following the primary operation.

Figure 6 – (a) CT aspect of a 9-cm distal pancreatic mucinous cyst; (b) Cyst epithelium with papillary architecture and low Ki-67 index (×200).

Topics

According to the WHO classification of pancreatic cysts (PCs), which divides the PC in neoplastic and non-neoplastic cysts, MCA is a type of mucinous pancreatic neoplastic epithelial cyst [7, 8]. MCA represent 10% to 45% of pancreatic cystic neoplasia, but given the fact that most of them are asymptomatic, it is difficult to obtain data about their actual incidence and prevalence [4]. They occur almost exclusively in perimenopausal women, the average age being 48 years. In this series, the median age was 47 years. There was a peak incidence between the decades three and six of age (76.47% of the patients).

MCA cases in male patients are exceptional; the F/M gender ratio is 20/1 to 13/1 [3, 9]. In the present series, there was a female predominance, with an F/M ratio of 14.5/1. The presence of estrogen and progesterone receptors within the ovarian-type stroma demonstrates maybe due to complex hormonal pathogenesis interactions its similarity to true ovarian stromal tissue [2].

MCAs are well-encapsulated spherical cysts, presenting septae and walls with rich vascular network. These cysts often exceed 10 cm in size [10]. They usually contain a viscous mucoid liquid, but there are also cases in which the cystic content is hematic, serous or purulent leading to confusion between MCA and a PP [3]. In the present series, we reported a few cases in which the cystic content was hematic and purulent (therefore these cysts were initially intraoperatively taken for PP). MCAs were originally referred to as “macrocystic adenomas” because of their uniloculated appearance but they usually have septae and sometimes a solid component [11]. MCAs develop mainly in the body and tail of the pancreas [3, 9, 12, 13]. In our series, MCAs were located mainly in the body and tail of the pancreas (80.6%). The median cyst size was 7 cm, five cysts having 10 cm to 15 cm.

The microscopic cystic epithelium of the walls is composed of columnar cells, among which there are mucin-producing goblet cells, and, in most cases, neuroendocrine cells. The latter are identified by immunohistochemical methods by using neuroendocrine markers such as chromogranin or synaptophysin. The cystic wall may present papillary projections with trabecular and thickened appearance. Nevertheless, due to possible existence of areas of malignant degeneration, microscopic examinations of solid areas are mandatory [3]. The ovarian type stroma is composed of columnar epithelium cells with oval nuclei arranged in bundles. In addition, cells of the corpus luteum and progesterone receptors can be identified in the stroma [3]. Given the presence of the ovarian stroma, one can assume that these cysts occur from residual ovarian tissue in the pancreas during the embryological development.

Various degrees of dysplasia may be present in the MCA epithelium. Dysplasia is more probable in large cysts mainly located in the head of the pancreas [3]. These data support the adenoma–dysplasia–carcinoma sequence in MCA, and therefore this type of cyst should be addressed as a premalignant lesion. In this study, we found low-grade dysplasia in three cases (we excluded the 17 patients with cystadenocarcinoma and high-grade dysplasia).

The presence of symptoms depends on cyst location and size; since most of them are located in the body or tail of the pancreas, no symptoms are expected to occur over a long period. Yeh et al. [10] stated that most MCAs with a long asymptomatic evolution, can reach over 10 cm in size at the time of diagnosis. This was also the case in one of our patients.
MCA symptoms are non-specific: nausea, vomiting, early fullness or intermittent abdominal pain [14]. In the present study, the majority of MCAs (90.3%) were symptomatic and only three were diagnosed incidentally.

In few cases, MCAs may be the cause of acute pancreatitis with subsequent pseudocyst formation, as has been reported in the literature [15–18].

The working group of Consensus Symposium within the “14th Meeting of the International Association of Pancreatology” in Fukuoka, Japan, in 2010, has developed in 2012 new guidelines based on “consensus”, rather than “evidence-based”, for the management of MCAs and intraductal papillary mucinous neoplasms (IPMNs) [19].

US examination is usually the first imaging step but preoperative diagnosis of MCAs is mainly based on CT examination. Multi-detector CT (MDCT) scan shows a typical appearance: mono/oligoloculated lesion with thick walls and septae that may have a regular or fine appearance and may have growths and peripheral nodules. In some cases, peripheral calcifications or linear calcifications within the cystic wall, as well as epithelial papillary growths or debris may be present [20]. Parietal calcifications make the differential diagnosis between an MCA and a PP difficult. These findings were present in two patients in our series (in one of them being misinterpreted as kidney stones on US). Unless there is a fistula, MCAs do not communicate with the pancreatic ductal system. Recent studies have shown, however, the existence of microscopic communications demonstrated by injecting dye into the pancreatic ducts [5].

CT examination may prove useful in detecting complicated MCAs. In three cases in our study, portal vein thrombosis with portal hypertension and collateral circulation was diagnosed preoperatively.

MRI and MRCP provide additional important information. MRI examination is especially useful in small MCAs in the differential diagnosis with other pancreatic cystic lesions. According to Bauer [21], MRCP detects [with greater accuracy than the invasive endoscopic retrograde cholangiopancreatography (ERCP)] even the smallest communication between the cyst and the pancreatic ductal system. Therefore, it is very helpful in differentiating MCAs from branch-duct-type IPMNs and the association of MRI and CT may increase diagnostic accuracy when results are equivocal [22].

Endoscopic ultrasound (EUS) is another powerful diagnostic tool, by providing detailed imaging and allowing fine-needle aspiration (FNA). Cyst fluid may be analyzed for tumor markers [carcinoembryonic antigen (CEA), CA125, CA19-9], amylase content, amylase isoenzymes, relative viscosity and cytology [23]. Tumor markers obtained by endoscopic FNA are more accurate than the serum markers and may differentiate MCAs from serous cysts.

CEA is the most important marker that can indicate with 75% accuracy the presence of a mucinous cyst, when it has a cut-off value of 192 ng/mL [24] but cannot differentiate between MCAs and IPMNs; also, it cannot make the difference between malignant and benign cysts [25, 26]. FNA is not required in all patients. MCAs in surgically fitted patients have a clear resection indication [19]. Therefore, FNA is only required in small cysts with uncertain diagnosis, in patients not fitted for surgery or elder patients with MCAs less than 4–5 cm in diameter (in which case surveillance is an option). In this series, cyst fluid tumor markers were not evaluated since EUS-guided FNA has not been available until recently.

Other authors considered Ki-67 expression as important differential diagnostic tool between benign and malignant intraductal papillary-mucinous tumors with impact on the pre-operative planning of treatment in individual cases [27].

Although high CEA, CA19-9, and CA125 serum levels can indicate the presence of a MCA, they have no relevance in diagnosing MCAs, since the sensitivity is of only 17% [28].

In the last years, new diagnostic tools have become available. It is worth mentioning the acoustic radiation force impulse (ARFI) US [29] or the imaging with confocal laser endomicroscopy [30].

Distinction between MCA and other pancreatic fluid collections (PPs, IPMNs and serous cystadenomas) is of paramount importance. PPs usually follow an episode of acute pancreatitis but there are reports in the literature describing MCA, associating acute pancreatitis and concurrent PP [15]. In our experience, large MCAs can be misdiagnosed as PP due to the necrotic content, thick wall and the inflammatory reaction of the nearby structures. When a clear diagnosis cannot be established preoperatively, fresh frozen pathological examination may be helpful, guiding the type of surgery.

Treatment of MCAs is continuously evolving. In selected cases (generally in patients not suited for surgery), after evacuation of cystic content via FNA under EUS guidance, ablation of cystic tissue may be achieved by injecting ethanol or Paclitaxel for 3–5 minutes [19, 31, 32]. On short term, the rate of success based on CT scans is between 33% and 79% [31, 33].

Surgical treatment of MCAs can be performed by an open or a minimal invasive approach [34]. According to the relatively recent consensus guidelines of the International Association of Pancreatology, surgical resection is recommended for patients with MCN any time possible. Laparoscopic resection, parenchyma-sparing resections and distal pancreatectomy with spleen preservation should be considered for MCNs of <4 cm without mural nodules [19].

In cases with uncertain preoperative diagnosis, fresh frozen examination is necessary. However, the risk of obtaining a false negative result, especially in large cysts, is increased due to the possible discontinuity of areas with malignancy [14]. Fresh frozen pathological examinations were performed in our study in three cases, showing a typical MCA appearance.

The type of surgical resection depends on the localization of the cyst, on its dimensions and its relation to adjacent anatomic structures. Since MCAs predominate in the body and tail of the pancreas, the most frequent procedure is SP. Cephalic MCAs require pancreateoduodenectomy (PD) and, in case of voluminous cysts, total pancreatectomy (TP) may be required. It is highly recommended that a proper approach be chosen.
in this type of pre-malignant lesion, with lymph node  
dissection in cases larger than 4 cm [19].

Surgical drainage procedures (for large painful cysts)  
may be performed in cases where major risk factors  
are involved (old age, severe comorbidities).

Although MCAs have a high risk of malignancy, most  
studies show that they are not aggressive tumors. Yamao  
et al. [5] found that the 3-, 5-, and 10-year survival rates  
were 97.6%, 96.6%, and 96.6%, respectively. The  
prevalence of invasive carcinoma is less than 15%, while  
cysts smaller than 4 cm without wall nodules do not have  
any signs of malignant degeneration. MCA resection has  
good results, recurrences are exceptional [19].

Conclusions

MCAs represent a rare pancreatic pathology with  
challenging diagnostic and therapeutic implications. Most  
of them are symptomatic but a small number may be  
discovered incidentally. MDCT scan, EUS and MRI/MRCP  
are useful in the differential diagnosis with other pancreatic  
fluid collections and in guiding the therapeutic approach.  
EUS with FNA is recommended in selected cases only.  
Oncological surgical resections are recommended, given  
the malignant potential of MCAs. Histopathological  
examination establishes the final diagnosis and the  
presence of focal dysplasia or malignancy. The most  
common postoperative complication is pancreatic fistula.

Conflict of interests

The authors declare that they have no conflict of interests.

References


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