Original article

Giant appendiceal mucoceles of cystadenomas type: CT and MRI evaluation. Report of three cases and review of literature

I. Tsitouridis, G. Kouklakis, A. Kalambakas, Ch. Papastergiou, M. Emannouilidou, F. Goutsaridou, Ch. Tsantiridis

SUMMARY

Mucocele is a general descriptive term, denoting an accumulation of mucin in a dilated lumen. Mucocele of the appendix is a frequent entity when it has a small size hut is very rare when there is grant enlargement of the appendix. We evaluated 3 cases with giant mucocele of the appendix of cystadenomas type, with calcification of their wall, US, CT and MRI findings, the diagnostic approach and the differential diagnosis with a literature review were presented.

Key words: Mucinous cystadenoma, Mucocele, Appendix, CT, MRI

INTRODUCTION

Appendiceal mucocele is a general term applied to the abnormal accumulation of mucus into the lumen of the appendix, regardless of the underlying disease. The incidence in appendectomy specimens is about between 0,2% to 0,3[1-3]. Most of the appendiceal mucocele histologically classifies into four groups, retention mucoceles focal or diffuse hyperplasia without atypia, mucinous cystadenoma and its malignant cystadenocarcinoma with glandular invasion into the stroma[4,5].

The preoperative diagnosis of mucinous cystadenoma is very important, because some of them are considered to have malignant potential. We analyzed morphologic characteristics of three cases of giant mucoceles cystadenoma type at US, CT and MRI.

Department of Radiology and Gastroenterology, Papageorgiou General Hospital, Thessaloniki, Department of Gastroenterology, 424 General Hospital, Thessaloniki

CASE REPORTS

CASE 1:

The first patient was a woman, 72 years old, complaining of for mild pain at the right iliac fossa experienced during the last two years. An abdominal x-ray film revealed two calcified lesions, one in the right iliac fossa and another in the lesser pelvis centrally located and corresponding to a well known calcified leiomyoma of the uterus (Figure 1A). On CT examination of the lower abdomen the first lesion was found to be a large well-circumscribed cystic mass to calcified wall in close proximity with the cecum and extending to the topographic position of the right ovary (Figure 1B, C). MRI examination clearly picked up the cystic origin of the lesion with low signal intensity on T1-weighted images and high signal intensity on T2-weighted images and the relationship with the cecum (Figure 1D). The patient had an operation which revealed a giant mucocele of the appendix of cystadenoma type, with calcified wall and intraluminal mucinous material.

CASE 2:

The second patient was a 65 year old man who complained mild pain in the right lower abdomen experienced during the last four years. Ultrasound examination revealed a cystic lesion with an echogenic wall and intraluminal low intensity echoes (Figure 2A). On CT, the lesion revealed low density, partialy calcified wall, and was in close proximity to the cecum. The latter relationship was clearly obvious after the orally administration of contrast medium (Figure 2B). On MRI examination and in T2-weighted and T1-weighted images the lesion revealed low signal intensity in the wall and a watery intraluminal content with high signal intensity at T2-weighted images (Figure 2C). At surgery, a large mucocele of the appendix of cystadenoma type was removed.

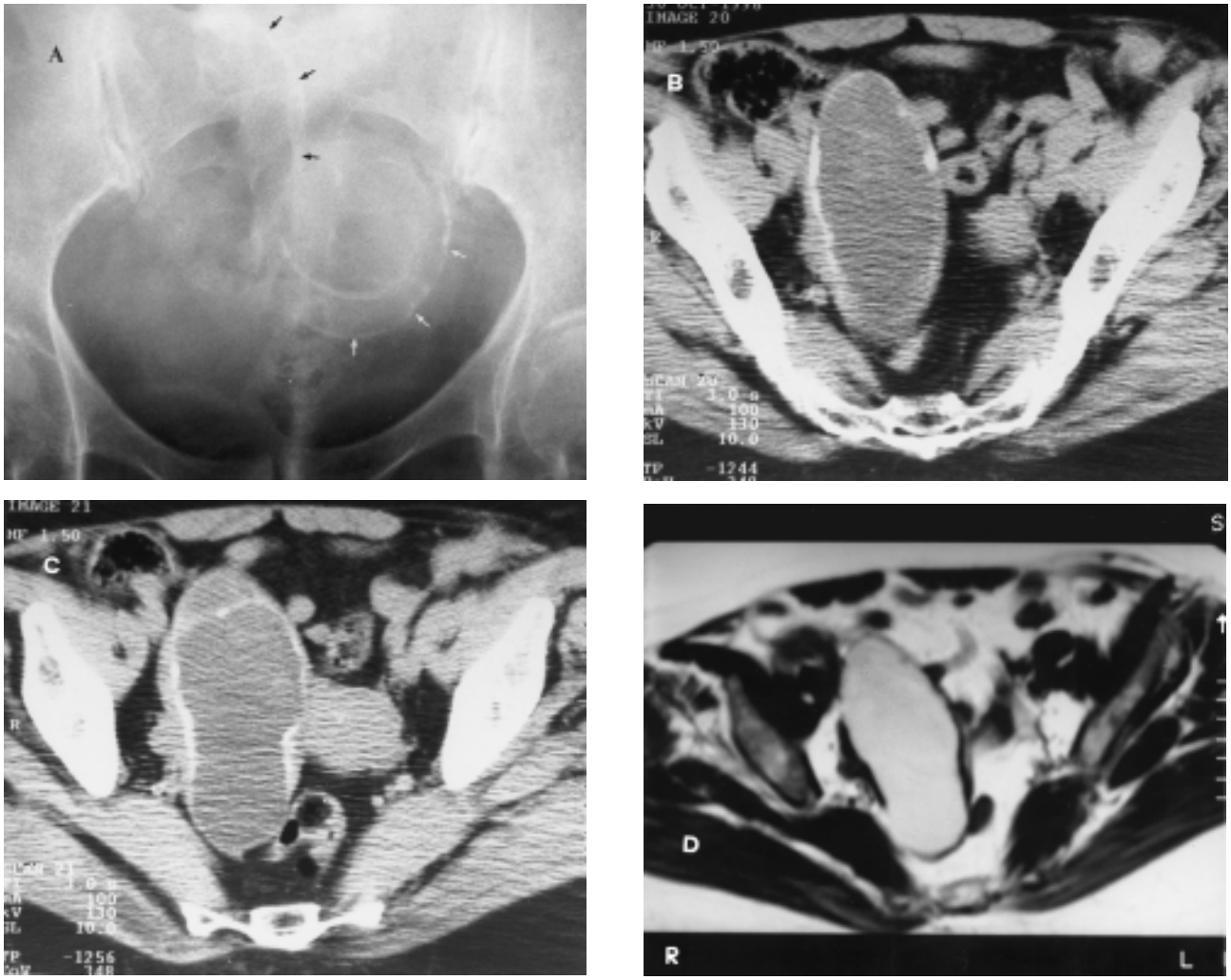

Figure 1. A. Plain x - ray of the lower abdomen shows two calcified lesions; a leiomyoma on the left (white arrows) and an ovoid linear calcification on the right (black arrows). **B, C.** Axial CT scans revealed the cystic lesion with rim calcification and its relationship with the cecum. **D.** Axial MR T2-weighted image shows the mucocele with high signal intensity and the rim calcification with low signal intensity.

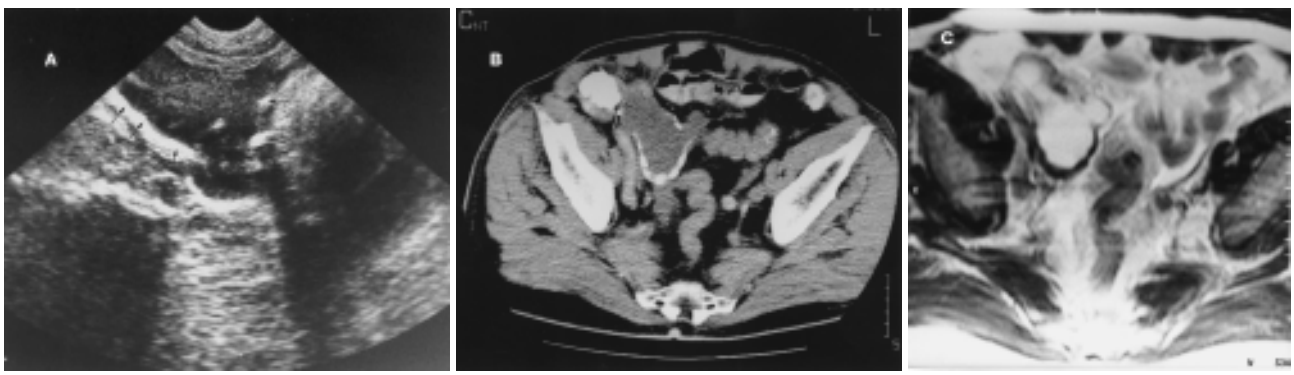

Figure 2. A. Ultrasonogram of the pelvis which reveals a hypoechogenic mucocele of the appendix with low intensity echoes and linear or nodular calcifications of the wall (arrows). **B.** Axial CT scan of the pelvis which clearly reveals the appendiceal mucocele, with linear and nodular calcifications of the wall and in close proximity to the cecum (white arrow). **C.** Axial MR T2-weighted image which shows the mucocele with high signal intensity and the calcification of the wall.

CASE 3:

The third patient was a 56 year old man, who was asymptomatic and had only a CT examination as a staging procedure for urinary bladder cancer. In the right side of the pelvis there was a cystic lesion with small nodular calcification of the wall and in close proximity to the cecum (Figure 3). At surgery the lesion proved to be an appendiceal mucocele of cystadenoma type.

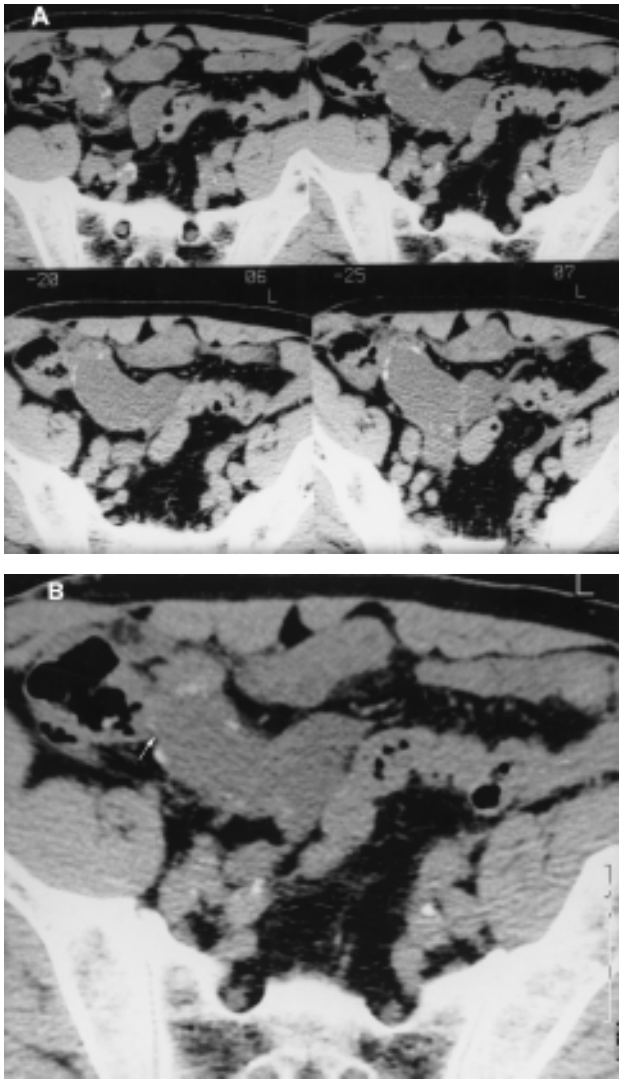

Figure 3. A. Axial CT scans with 5 mm slice thickness which clearly reveals a large appendiceal mucocele attached to the cecum, with some spotty calcifications of the wall. **B.** Axial CT scan (the second of the above scans) which shows the relationship of the cystic lesion to the cecum (arrow).

DISCUSSION

The term mucocele is a descriptive one, which is used in multiple sites and means the dilatation of a lumen and filling with mucus, without any comment on the underlying disease. Mucocele of the appendix is a very rare entity and the incidence in appendectomies and autopsy series is about 0.2% to 0.3%[1-3].

The macroscopical appearance of an appendiceal mucocele depends on the degree of dilatation and filling of the lumen, which are responsible for the various morphological appearances of mucocele, from an almost normal appendix to a giant appendix over 10cm, with differing viscosity of content, from watery to gelatinous one[6,7].

In our three cases, the appendix was over 10cm in length and all had gelatinous appearance of the mucus.

Most of the appendiceal mucocele are histologically classified according to their lining epithelium into three groups[4-6]: a) simple or retention mucocele resulting from the obstruction of the appendiceal outflow, with a normal epithelium and luminal dilatation up to 1 cm, b)mucocele with focal or diffuse hypeplasia of the epithelium without atypia and with mild dilatation of the appendiceal lumen, c) mucinous cystadenoma and cystadenocarcinoma. Mucinous cystadenomas are the most common mucoceles and, in most cases exhibit, epithelial villous adenomas changes with some atypia, and also with marked dilatation of the appendiceal lumen of up to 6cm.

Mucinous cystadenomas are associated with micro or massive perforations of the appendix in 20% of cases[4].

The mucinous material into the peritoneal cavity is usually acellular without epithelial cells and the spreading of mucin from the ruptured mucinous cystadenomas is usually around the appendix and only occasionally is found free in small amounts in the peritoneal cavity[8]. Mucinous implants in the peritoneal cavity regardless of the underlying disease are designated as pseudomyxoma peritonei, although it is thought by many authors that only mucinous cystadenocarcinoma is responsible for this complication[9,10].

Malignant mucinous cystadenocarcinoma is usually indistinguishable on gross appearance from benign cystadenoma. Microscopically, they also have many features in common. The two most reliable findings for the identification of cystadenocarcinomas, are the glandular stromal invasion, and/or the presence of epithelial cells in the extra-appendiceal mucinous implants[4].

When pseudomyxoma peritonei is found in a patient with cystadenocarcinoma of the appendix, the prognosis

is very poor[11].

Appendiceal mucocele is usually asymptomatic. The most common symptoms in symptomatic patients are acute or chronic mild pain in the right lower abdomen and vague abdominal distress[12]. Rarely intermittent pain and bleeding due to intussusception of the mucocele into the cecum may also occur[13,14].

Our patients were either asymptomatic or had mild chronic pain in the right lower abdomen.

Acute or chronic abdominal right lower quadrant pain occurs in about two thirds of patients with appendiceal cystadenomas[2]. Approximately 23-50% of patients are asymptomatic with the lesion being discovered incidentally during surgery, radiologic evaluation or endoscopic procedures[2,15].

About 15% of patients also present with a palpable abdominal mass, due to its large size[2,4,5].

Preoperative diagnosis is very important because cystadenomas of the appendix carry the risk of rupture with spillage of mucus and malignant transformation.

The imaging features and diagnostic criteria of the appendiceal cystadenomas are the calcifications of the wall, the imaging behavior of the mucus and the topographic relationship to the cecum.

Dystrophic calcification of the wall due the chronic inflammatory process may be curvilinear or punctate and this is better appreciated on CT scans than sonography which shows a highly echogenic wall with or without distal shadowing. The presence or absence of distal shadowing depends on the density of the calcified foci. Occasionally the wall is totally calcified, similar to the condition of porcelain gallbladder.

In our cases, curvilinear calcification clearly detected in the one case and punctuate calcifications in two.

Intraluminal mucin could be in liquid or gelatinous and viscous phase and also may coexist with macroaggregates of proteins, debris or cellular material providing a number of different acoustic interfaces[9,15]. In the large majority of the cases there is excellent through-transmission and posterior enhancement[14-16].

CT may show a cystic mass with or without septations. CT attenuation values should range from near water density to soft-tissue density[15].

Enhancing nodules in the wall of the mucocele at CT have been reported that suggest mucinous cystadenocarcinoma, although the number of reported cases was only 2 and further work with a larger number of cases is needed to established this finding as the differential point between cystadenoma and cystadenocarcinoma of the appendix[20].

MR imaging subsequently showed intraluminal mucin as a cystic mass of low intensity on T1-weighted images and high signal intensity on T2-weighted images[21].

The relationship of the appendiceal mucocele with the cecum is very important for the diagnosis. This becomes more apparent when the cecum is filled with oral contrast material and thin sections with CT were obtained[20]. Sometimes when the obstruction of the appendiceal lumen is distal to its orifice then the mucocele is not in close proximity to the cecum[15]. In comparison, CT is more helpful than MRI in the evaluation of the area of the cecum and appendix.

The differential diagnosis of an appendiceal mucocele includes mainly hydrosalpinx, ovarian cystic lesions, secondary duplication cyst, mesenteric and omental cysts, lymphocele, mesenteric and retroperitoneal hematoma or tumors and abdominal or retroperitoneal abscesses[19].

In conclusion, appendiceal mucocele has a spectrum of radiological findings classifable into 3 groups (calcifications, imaging of the mucin content and relationship with the cecum), which, combined together, differentate this condition from mimicking diseases.

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