Mucocele of the appendix mimicking an adnexal mass: a case report

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Summary

Mucocele of the appendix is a rare entity usually mimicking an adnexal tumour. There is no specific imaging or screening method to determine the diagnosis with certainty preoperatively. Appendiceal malignancy can be the underlying cause, although it is not common. We present a case of an appendiceal mucocele mimicking an ovarian tumour by both clinical and imaging (TVS and MRI) methods. This pathological condition should be considered by all the gynaecologists in the differential diagnosis of a right-sided pelvic mass.

Key words: Adnexal mass; Appendix; Cystadenoma; Mucocele.

Introduction

Mucocele of the appendix is a rare entity, infrequently diagnosed prior to surgery or autopsy. It is characterised by cystic dilatation of the appendiceal lumen due to abnormal accumulation of mucus. Its incidence ranges between 0.2% and 0.3% of all appendicectomy specimens [1], with a higher frequency in females (4:1) over the age of 50 years. Mucocele of the appendix is often asymptomatic. Due to the rarity of this entity and the lack of specific symptoms this condition is often not considered when women complaining of lower quadrant pain present to the gynaecologist.

We present a case of a 63-year-old woman with a right-sided pelvic mass misdiagnosed to be of ovarian origin.

Case Report

A 63-year-old postmenopausal woman was referred to our department with the diagnosis of an asymptomatic right-sided pelvic mass. On admission a mobile, cystic, painless mass, about 7 x 3 cm in size was palpated in the lower right abdomen.

Transvaginal ultrasound (TVS) scan revealed a multilocular oval cyst measuring 7.5 x 3.5 x 3.0 cm, that was thought to be of ovarian origin. The uterus and left adnexa were normal. Magnetic resonance imaging (MRI) examination revealed a cystic oval mass, 7.9 x 2.8 x 3.2 cm in size, in the right lower abdomen with clear margins, fluid content and rim enhancement (Figure 1).

Laboratory tests, including AFP, CA 15-3, CA 19-9 and CA 125 were unremarkable, while CEA was slightly elevated measuring 11.2 ng/ml (normal values < 5 ng/ml).

An exploratory laparotomy was performed. The uterus and both adnexa were macroscopically normal. A mass 8 x 3 x 3 cm in size was found arising from the appendix. The mass was mobile, in close relation to the right ovary, and without any adhesions in the area (Figure 2).

An appendicectomy was performed. The appendiceal tumour was removed intact. Frozen section revealed mucocele of the appendix with no evidence of malignancy. The final pathologic diagnosis was mucocele caused by mucinous cystadenoma with slight cellular dysplasia. A colonoscopy was performed three months later and no abnormality was found.

Discussion

Appendiceal mucocele is a general term applied for dilatation of the lumen of the appendix due to mucinous secretions, regardless of the underlying disease.The pathogenesis could be neoplastic or non-neoplastic. Simple (non-neoplastic) mucoceles are usually the result of obstruction of the appendiceal outflow by postinflammatory fibrosis or fecolith while the epithelium may be normal or just hyperplastic. Mucinous cystadenoma is the most common subgroup (63-84%), its pathogenesis is neoplastic, but histological examination does not reveal any malignant cells [2]. Finally, mucinous cystadenocarcinoma, applicable for 11-20% of all cases, is the malignant form of mucocele. Intraperitoneal spread of the neoplastic cells and formation of the adhesive, semi-solid mucin results in a condition called pseudomyxoma peritonei [2].

A correct preoperative diagnosis of appendiceal mucocele is extremely difficult and rarely made. Usually it is considered as an adnexal mass. There is no specific imaging or screening method that could diagnose this condition with certainty preoperatively [3, 4], except for a slight elevation of serum CEA which should always alert the physician as CEA rises in mucinous tumors [5]. CEA was also elevated in our case, but it was estimated wrongfully to be of ovarian origin. Diagnosis is confirmed only during surgery, as also occurred in our case. Rare complications include intestinal obstruction, intestinal bleeding and pseudomyxoma peritonei [6].

The differential diagnosis mainly includes cystic tumours of the right adnexa as well as mesenteric and omental cysts, lymphocele, mesenteric and retroperi-

Revised manuscript accepted for publication October 13, 2008

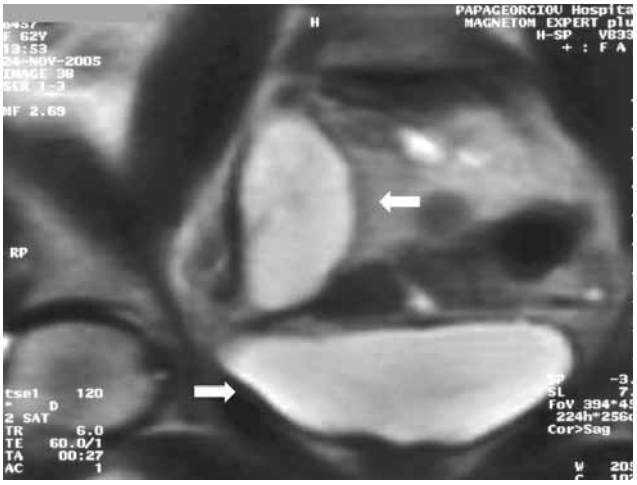

Figure 1. — MRI examination showing the cystic mass (top arrow: cystic mass; bottom arrow: urinary bladder).

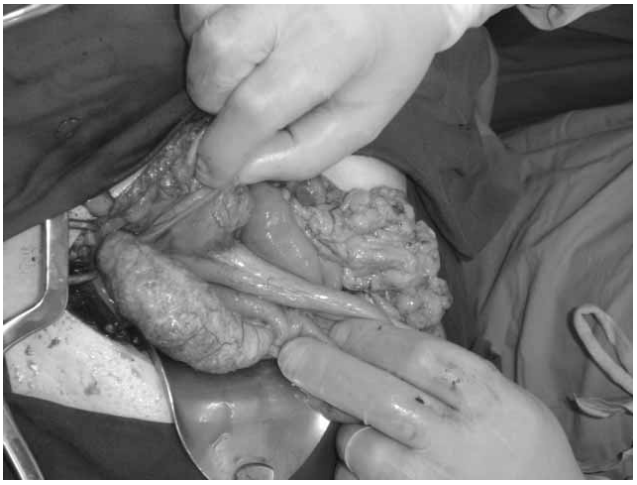

Figure 2. — Appendiceal tumour during the operation.

toneal haematoma or tumours and abdominal or retroperitoneal abscess [4].

Simple appendicectomy is curative in uncomplicated unruptured cases. When surgical exploration reveals a ruptured mucocele, then removal of all gross implants should follow the primary resection. Surgical excision of mucocele can be done either by laparotomy or laparoscopy. The endoscopic approach provides the advantages of good exposure and exploration of the abdominal cavity as well as rapid recovery postoperatively [7, 8].

The prognosis of patients with simple or benign mucoceles is excellent with a 5-year survival rate of 91-100%. Patients with cystadenocarcinomas, however, have a markedly diminished 5-year survival rate of 25%, mainly due to the complications of pseudomyxoma peritonei [6]. Postoperative evaluation by a gastroenterologist is always suggested, as cases with adenocarcinoma of the appendix have been missed by the pathologist or pseudomyxoma peritonei has been reported without intraoperative rupture of the mucocele [9]. In our case a colonoscopy was performed three months after the operation and no obvious pathology was detected.

Conclusion

Appendiceal mucoceles are extremely rare. They should always be considered in the differential diagnosis in women presenting with a right-sided pelvic mass, especially when clinical features are not indicative of gynaecological pathology. A correct preoperative diagnosis, though quite difficult, could be helpful in avoiding iatrogenic rupture as well as to plan surgery.

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