E-QUID: ANSWER / Gastrointestinal imaging

Appendiceal mucinous cystadenoma

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Observation

A 62-year-old male came to the emergency department presenting abdominal pain in the right iliac fossa together with fever and inflammatory markers. The interview revealed neither transit problems nor alteration in his general condition. The only feature in his history was sigmoid diverticulitis, diagnosed 10 years previously.

A CT scan following injection of iodinated contrast agent in the portal phase brought to light sigmoid diverticulitis complicated by pylephlebitis of the inferior mesenteric vein. It also showed a pelvic mass (Fig. 1a, b and c).

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* Here is the answer to the case ‘‘Pain in the right iliac fossa: An aetiology that should not be underdiagnosed’’ previously published. As a reminder we publish again the entire case with the response following.

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What is your diagnosis?

From the observations, what diagnosis would you choose from the following proposals:
- Meckel’s diverticulitis;
- non-Hodgkin’s lymphoma of the appendix;
- appendiceal mucocele;
- actinomycosis of the appendix;
- appendiceal abscess.

Discussion

An appendiceal mucocele is a rare condition, observed in 0.2 to 0.6% of appendectomy specimens [1]. The mean age for its occurrence, predominantly in women is 50 to 60 years [2]. It poses the dual problem of its possible malignancy and the risk of gelatinous disease of the peritoneum (peritoneal pseudomyxoma) in the event of perforation, which occurs in 10 to 15% of the cases.

There are many etiologies for an appendiceal mucocele, which can be malignant or benign [2]. The mucocele may be a simple retention cyst caused by the accumulation of mucus in the appendix, secondary to its obstruction by a coprolith or proximal to an inflammatory stenosis or tumor. In this case, the mucocele usually measures less than 2 cm in diameter. Other etiologies are mucus-secreting tumors, including villous hyperplasia, mucinous cystadenoma and mucinous cystadenocarcinoma. Histological examination is essential [3].

In mucosal hyperplasia (villous adenomas), the appendix is normal or slightly dilated with a thinned mucosa. Histologically, the lesions are limited to the mucosa and arranged in thin papillary structures without atypia or mitosis. In mucinous cystadenomas, the appendix is dilated.
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by the mucus and the lumen is lined with a single-layered mucosecreting epithelium. Papillary forms can exist but the epithelium is usually flat. Various degrees of cell dysplasia may be observed. The mucinous cystadenocarcinoma is the most feared etiology. Macroscopically, the lesions are no different from those of mucinous cystadenomas but there is a high degree of cell atypia and mitosis, invasion of the muscle by neoplastic cells and the presence of neoplastic cells in the intraperitoneal mucus effusion.

Where there is a mucocele, the diagnosis from an AXR is indicated by arcuate calcifications at the location of the appendix that are present in half of the cases [3].

Ultrasound shows a distended appendix with a cystic, anechoic content or sometimes containing a finely echogenic sediment in layers and moving with changes of position [4]. Ultrasound may also show an associated mucoid effusion and in women, an ovarian mass should be sought (an associated mucinous tumor of the ovary). The most helpful imaging examination is an abdominopelvic CT scan. It shows a formation of liquid density (10–30 HU) connected to the ceacum, with walls which are sometimes finely calcified and enhanced by the contrast agent, and more or less regular depending on the etiology [4]. The infiltration of periappendiceal fat is non-specific and may be of inflammatory or neoplastic origin. A CT scan also allows complications to be diagnosed: inflammation, invagination, torsion, compression of the ureter and finally, the most to be feared, pseudomyxoma [5]. With MRI, a fusiform dilatation of the appendix is seen, hypointense with T1-weighting and hyperintense with T2-weighting. Its walls are enhanced by gadolinium [1].

Peritoneal pseudomyxoma is a peritoneal or retroperitoneal accumulation of gelatinous substance secondary to the rupture of a mucinous appendiceal lesion [4]. This gelatinous substance may be acellular and have a very good prognosis or include free mucus-secreting epithelial cells of tumoral origin. Peritoneal pseudomyxoma, thus, sometimes combines mucinous ascites and peritoneal implants, in which case the prognosis is very much poorer, synonymous with recurrent peritoneal involvement still known as gelatinous disease of the peritoneum.

Figure 2. Abdominopelvic CT scan in the portal phase: Fig. 1a and b: axial slices; Fig. 1c: sagittal reconstruction.

Figure 3. Macroscopic examination of the appendectomy specimen. Appendix distended by gelified mucus. Thin wall without vegetation. No perforation.
The treatment of appendiceal mucoceles is surgical for two reasons: their potential malignancy and the possibility of rupture in 5 to 15% of the cases with the risk of dissemination and peritoneal pseudomyxoma [2]. As a rule, the surgical approach is laparotomy, but laparoscopic surgery may possibly be chosen for simple, non-ruptured forms. Benign forms amount to standard appendectomy (McBurney incision or laparoscopy), avoiding cell dissemination. For malignant forms, concomitant ablation of the caecum can be envisaged from the outset. Clear signs of malignancy in the images and/or on extemporaneous study of ablated tissue may lead to a right hemicolectomy being performed [2]. If there is contamination with inoculation into the peritoneal cavity, additional treatment by intraperitoneal chemotherapy may be indicated [1].

The prognosis for benign forms of mucoceles (retention mucoceles, mucosal hyperplasia and mucinous cystadenum) is excellent following complete ablation, with survival at 5 years of almost 100%. For malignant forms, the survival rate correlates with the degree of extension of the tumor and varies between 30 and 80% [2].

Conclusion

Radiological diagnosis of a non-ruptured appendiceal mucocele is an essential element for providing the prognosis for the disease, allowing the surgeon to take the necessary precautions to avoid an intra-operative peritoneal rupture. Certain features, particularly appendiceal distension and parietal calcifications, should evoke a mucocele in an appendiceal condition, irrespective of the clinical picture. Histopathological examination of any appendectomy tissue is essential for determining subsequent therapeutic management.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

References