PSEUDOMYXOMA ON APPENDICULAR MUCOCELE

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Abstract

Mucocoele of the appendix is a descriptive term for mucinous distension of the appendiceal lumen (vermiform appendix) regardless of the underlying pathology. It refers to the progressive retrograde dilatation of the vermiform appendix with concomitant intraluminal accumulation of the mucoid substance. It is an uncommon pathology that occurs in both sexes. It especially poses the problem of differential diagnosis in particular in women because of the location of clinical symptoms in the right iliac fossa. The incidence is estimated between 0.2% and 0.4% of the appendectomied specimens. The estimated incidence is approximately 1/1 000 000/year. The disease onset is usually after the age of 40 years and more frequently affects females. The means of medical imaging are mainly ultrasound and scanner. On CT typical mucocoele appears as a cecal-based, rounded and well-defined mass, thin-walled, with fine parietal calcifications; CT density is variable, from fluid to tissue. A stercolith is sometimes visible at the base of the appendix. The wall of the mucocoele may be thickened, irregular, taking the contrast there may be peri-appendicular inflammation, which may be inflammatory or tumor, without specificity. The treatment of unbroken appendicular mucocoele is surgical, preferably by laparotomy than laparoscopy. The appendectomy is performed, without breaking the appendix, with complete resection of the meso-appendix, and sampling for cytology of the peritoneal fluid.

Introduction:

Case report:

Medical Observation:

55-year-old woman, admitted to the emergency room for abdominal pain in right iliac fossa, associated with vomiting and fever. This symptomatology evolving for one week, without notion of stopping stool. Physical examination reveals diffuse abdominal pain more pronounced in the right iliac fossa with localized defense.

Biological exams reveal:

Total WBCs (white blood cells count): 24x10³/µL

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C Reactive Protein 42 mg/l
High sedimentation rate 55mm/h
The computed tomography shows:( Figures 1,2,3,4)
1. large appendix in laterocecal position with with hypodense content and swollen pelvic,
2. Mass effect on bladder, compression on right pelvic ureter and contact with anterior uterus face
3. Peritoneal effusion
4. Infiltration of meso colic fat
5. Multiple lymphadenopathy meso colic, inter aortico cave, latero aortic

**Diagnosis:**
The very large masses connected to the cecum, in addition to nodular peritoneal effusion, peri-aortic lymphadenopathy are in favor of pseudomyxoma secondary to an appendicular mucocele.
Some differential diagnoses can be find about this symptomatology:

**Appendicular abscess:**
US shows collection of fluid (hypoechoic) in the appendicular region which may be well circumscribed and rounded or ill-defined and irregular in appearance.

The appendix can be visualized in the mass
On CT Fluid collection is observed in the appendicular region with or without air inside. Many times an appendicolith can be visualized.

**Adnexal lesion:**
(TOA tubo ovarian abscess…..) it can be confused with an adnexal lesion in the case of pelvic cecum; the key to the diagnosis then lies in the identification of a connection to the cecum, which must be carefully sought. The uterus and ovaries appeared to be normal

A tubo-ovarian abscess (TOA) is a complex infectious mass of the adnexa that forms as a sequela of pelvic inflammatory disease. Classically, a TOA manifests with an adnexal mass, fever, elevated white blood cell count, lower abdominal-pelvic pain, and/or vaginal discharge; however, presentations of this disease can be highly variable.

**Discussion:-**
Mucocele of the appendix is a descriptive term for mucinous distension of the appendiceal lumen (vermiform appendix) regardless of the underlying pathology. It refers to the progressive retrograde dilatation of the vermiform appendix with concomitant intraluminal accumulation of the mucoid substance. The incidence is estimated between 0.2 % and 0.4 % of the appendectomied specimens [1]

An appendix tumour is found every 100 appendectomies. It can also be incidental discovery during a routine radiological examination and presents as a mucocele of the appendix. This obstruction can either result from extrinsic compression (for example, by an endometriosis nodule) or be the result of an intrinsic obstacle. [1]
Pseudomyxoma peritonei is a macroscopic term used for jelly-like mucinous material within the peritoneum that accumulates as a result of a mucin-secreting

The appendix is by far the most common primary site for tumors producing pseudomyxoma, but rarely tumors of other organs.

The estimated incidence is approximately 1/1 000 000/year. The disease onset is usually after the age of 40 years and more frequently affects females. [2]

US shows a collection of the right iliac fossa with a distended appendix with an endoluminal collection. It can be associated with an infiltration and inguinal lymphadenopathy.Typical mucoceles of the appendix are hypoechoic masses. They may have a layered appearance.They may have a layered appearance (onion skin sign) They are well defined, cylindrical or lobulated in shape (pear shaped), with increased through transmission, and frequent thin curvilinear or punctate echogenic mural calcifications They are mobile but attached to the cecum.[3]
On CT typical mucocele appears as a cecal-based, rounded and well-defined mass, thin-walled, with fine parietal calcifications; CT density is variable, from fluid to tissue. A stercolith is sometimes visible at the base of the appendix. The wall of the mucocele may be thickened, irregular, taking the contrast there may be peri-appendicular inflammation, which may be inflammatory or tumor, without specificity. [3]

CT scan, pseudomyxoma mucinous ascites is hypodense, often discreetly more hyperdense than a simple transudate; it can be partitioned and contain fine curvilinear calcifications.

The radiological diagnosis of an unruptured appendicular mucocele is essential in the prognosis of the disease, allowing the surgeon to take the necessary precautions to avoid intraoperative peritoneal rupture; Its most serious complication is peritoneal rupture, which causes peritoneal pseudomyxoma which have the same treatment with mucocele appendicular.

The treatment of unbroken appendicular mucocele is surgical, preferably by laparotomy than laparoscopy. The appendectomy is performed, without breaking the appendix, with complete resection of the meso-appendix, and sampling for cytology of the peritoneal fluid.

Depending on the result of the cytology, the possible rupture of the mucocele, and the involvement of the meso-appendix, additional subsequent treatment may be performed: right hemicolecotmy, tumor cytoreduction, intraperitoneal chemohyperthermia. [3]

Figure 1: Axial IV contrast – enhanced CT throws the pelvis shows cecum (blue arrow) with large appendix (white arrow) and pelvic effusion (yellow arrow).
Figure 2: Coronal IV contrast-enhanced CT throws the pelvis shows lymphadenopathy (yellow arrow), cecum (white arrow), mesocolic fat infiltration (black arrow) and appendix (blue arrow).

Figure 3: Axial IV contrast-enhanced CT throws the pelvis shows a mucocele in large appendix (blue arrow) which is continuous with the cecum (white arrow).
Figure 4: Sagittal IV contrast-enhanced CT throws the pelvis shows an appendix, bladder, uterus.

Declaration of conflicts of interest:
The authors declare that there is no conflict of interest.

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