Diagnostic challenges in acute appendicitis: atypical presentation and pitfalls

Poster No.: C-1995
Congress: ECR 2015
Type: Educational Exhibit
Authors: D. Mandich¹, E. Martinez Chamorro², G. Ayala², V. Rueda², M. Pont², S. Borruel Nacenta³; ¹Madrid, Ma/ES, ²Madrid/ES, ³ES
Keywords: Inflammation, Diagnostic procedure, Ultrasound, CT, Emergency, Abdomen
DOI: 10.1594/ecr2015/C-1995

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR's endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys' fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

To determine and review the cases of acute appendicitis that raised diagnostic doubts in ultrasonography and MDCT.

Background

Appendicitis is the most common cause of acute abdomen in young people. Currently, the diagnostic role of imaging tests like ultrasonography and multidetector computed tomography (MDCT) is essential, especially when history and symptoms are atypical.

Despite the advances and the overall high accuracy of both techniques, diagnostic difficulties are not over. Atypical appendix positions or non inflammatory appendiceal entities are potential pitfalls and mimickers of appendicitis causing abnormal clinical presentation and misdiagnosis.

We classified the cases that have caused doubts in the diagnosis of acute appendicitis in our tertiary hospital emergency radiology department in two situations:

1. Acute appendicitis with atypical presentation: retrocecal location, malrotation by situs inversus, appendix with inguinal hernia, tip appendicitis, stump appendicitis and intraluminal appendiceal air.

2. Appendicular abnormalities simulating acute appendicitis: appendicular dilatation, appendicular diverticulitis, appendicular inflammation by contiguity, tumors and mucocele.

Findings and procedure details

Acute appendicitis is a common clinical entity and is the main cause of surgical acute abdomen in young patients. Acute appendicitis shows a characteristic history and clinical presentation, which is the base of the diagnosis of this pathology. Nevertheless, this typical presentation, to which we are familiar with, is not as frequent as believed and for this reason, the importance of imaging tests has been growing during the last years.
Nowadays, both ultrasound and multidetector computed tomography (MDCT) play a fundamental role in the correct diagnosis of acute appendicitis.

Ultrasound and MDCT have a high sensitivity and specificity for the diagnosis of acute appendicitis contributing to its correct diagnosis, especially in cases where the history and symptomatology is not the usual one. A meta-analysis Curtis et al. showed that, in children and adults, both ultrasound and MDCT have a high specificity (approximately 93-95%) whereas the sensitivity of MDCT is higher compared with that of ultrasound (94% versus 83-88%, respectively).

Despite the progress in imaging, difficulties in the diagnosis of acute appendicitis remain because there are appendicular and non-appendicular entities that may mimic this clinical condition. On the other hand, atypical presentations of acute appendicitis may lead to erroneous or delayed diagnosis.

The typical MDCT findings in acute appendicitis are appendicular dilatation, increased wall thickness, signs of periappendicular inflammation and the presence of an appendicolith.

We have classified cases that may lead to doubts in the diagnosis of acute appendicitis into:

I Appendicitis with atypical presentation:

- **Atypical location**: A common cause of atypical presentation and false negatives are the variations in the location of the vermiform appendix. Such variations may account for one third of cases of abdominal pain localized outside of the right iliac fossa.

The location of the base of the appendix is relatively constant on the surface of the posteromedial wall of the cecum. On the contrary, the tip of the appendix is mobile (Fig. 1,2). Changes in position are also favored by the dependence of the appendix on the cecum, which is also a mobile structure, because the position, distention and rotation of the cecum are variable.

Most commonly the appendix is intraperitoneal and shows a descending tip, but the appendix may rarely be located in a retrocecal-retrocolic position. Retrocecal appendicitis, specifically the ascending variant, may lead to inflammatory changes in retroperitoneal places such as the pararenal or perirenal spaces (Fig. 3), or may lead to intraperitoneal changes such as subhepatic appendicitis (Fig 4). These variants may
cause atypical manifestations of acute appendicitis in up to half of the cases, with upper abdominal pain that imitates biliary or renal disorders.

Another variation is the pelvic location of the appendix. Pelvic appendicitis is complicated frequently than retrocecal appendicitis because in these cases the appendicular pedicle is less prone to torsion and compression. Clinically, it can present with urinary symptoms, rectal tenesmus or even pain in the lower limbs (Fig. 5).

Finally, acute appendicitis may present pain in the left iliac fossa because an atypically long right appendix, or an appendix actually located in the left iliac fossa in cases of situs inversus (Fig. 6) or intestinal malrotation. The latter includes a spectrum of congenital abnormalities in the intestine caused by the absence or incomplete rotation of the primitive intestine in relation to the superior mesenteric artery during fetal life (Fig. 7).

Finally, abnormal position, mobility or rotation of the cecum and colon may lead to atypical locations of acute appendicitis. One possibility is to find a redundant and hypermobile ascending colon caused by an inadequate attachment to the posterior peritoneum.

- Distal appendicitis: the obstruction of the vermiform appendix is frequently caused by an appendicolith which may be located at any segment of the appendix. Because of this, a small percentage of cases show an obstruction at the level of the tip with the consequent initial inflammation of only just a small segment distal to the obstruction at a relative distance from the base and, the typical findings, such as the inflammatory involvement of the cecum, could be absent and the more insidious clinic make diagnoses more difficult (Fig. 8).

- Stump Appendicitis: This is a late complication of surgical treatment of acute appendicitis that was described for the first time in 1945 by Rose. Stump appendicitis is defined as an inflammatory process of any residual tissue of the vermiform appendix remaining after an appendectomy. Its incidence and causes are not clear; however, it is considered that a decrease in the irrigation of the stump or an appendicolith harbored in its lumen may initiate the inflammatory process. It is also believed that anatomic variants and complicated appendectomies could be risk factors, while a short stump (0.5 cm) could decrease the incidence of the aforementioned complication. In a review, Kanona et al. analyzed 51 cases of stump appendicitis reported in the literature: the majority occurred in males (67%) in association with pain in the right iliac fossa. Nevertheless, an important percentage of patients had nonspecific abdominal pain as clinical presentation. Moreover, their analysis showed that the time interval from the surgery to the stump appendicitis varied from 9 weeks to 50 years, and that it occurred after both open and laparoscopic appendectomies. With respect to laparoscopic appendectomy, some authors consider that it may have increased cases of stump appendicitis because of
failure to invaginate the stump into the cecum during surgery, thereby causing a recurrent inflammation. The diagnosis of stump appendicitis is a challenge because of the clinical history of previous appendectomy, with the ensuing risk of a late diagnosis and eventually the appearance of complications. In the same study, Kanoma et al. demonstrated that only 47% of the cases had a pre-operative MDCT and that it was diagnosed in 27.5% of them. Inflammatory changes in the area, the presence of abscesses, pericecal fluid, thickening of the cecum wall and even an appendicolith are findings that suggest the diagnosis (Fig. 9).

Our group has recently reported four cases of stump appendicitis diagnosed before surgery. In two of them, the diagnosis was done by an ultrasound scan that showed dilatation of the appendix remnant as well as hyperechogenicity of the mesenteric fat and the presence of free liquid. In a third, case unspecific inflammatory changes were identified in the right iliac fossa. MDCT was particularly useful in both cases in which a perforated acute appendicitis was diagnosed, including the presence of up to three appendicoliths in one of them.

- Appendix with intraluminal air: In the literature, the significance of the presence of intraluminal air in the appendix continues to be controversial. Nevertheless, currently it is not considered that the presence of air excludes an acute appendicitis. It is quite frequent to find intraluminal air in normal studies, but it has also been a visible finding in patients with acute appendicitis, and, for this reason, it is always advisable to correlate this finding with other signs that may suggest an appendicitis in MDCT or ultrasound scans. Another issue to be taken into consideration is the morphology and distribution of intraluminal air. Intraluminal air may be present in two different patterns: one diffuse or tubular that is more frequent in normal appendices, whereas a second "dirty" pattern or appearing like bubbles may suggest acute appendicitis. Recently, Cabarrus et al. published a retrospective study reporting that, despite air being more frequent in normal appendices, intraluminal air was evident in 27% of the cases of appendicitis, although they found no differences in air patterns between normal and pathological appendices.

II Appendicular pathology resembling acute appendicitis

- Appendicular diverticulitis: This is an infrequent disorder whose incidence has been described as varying from 0.004 to 2.1% of cases of acute appendicitis in the literature. Being male, age over 30 year-old, and suffering cystic fibrosis have been described as risk factors. As opposed to acute appendicitis, the clinical presentation of appendicular diverticulitis is more insidious, presenting with pain in the lower hemiabdomen of up to two weeks of evolution and unspecific blood tests, frequently delaying the diagnosis and increasing the risk of perforation. With respect to imaging tests, ultrasound could reveal the presence of diverticula and thickening of the appendicular wall. Appendectomy is
indicated in both symptomatic and asymptomatic cases since the most of the latter will develop an inflammatory process (Fig. 10).

- **Mucocele**: Mucocele is a general term used to describe dilatation of the vermiform appendix with mucinous content in its interior that may be caused by several entities (Fig. 11). Probable causes can be simple obstruction causing accumulation of secretions (also termed appendicular ectasia) with an appendix that usually does not exceed two cm in diameter, or mucin secreting tumors including cystadenomas or even mucinous cystadenocarcinomas.

Mucoceles are infrequent with a prevalence of 0.2-0.4% of the appendectomies reported in the literature. Its presentation may vary, but most of the times it is asymptomatic (although cases presenting with pain in the right iliac fossa been described) and up to 50% of the cases have been described as incidental findings in laparotomies or in imaging scans. The lack of inflammatory symptoms as well as the lack of inflammatory signs in the adjacent structures in MDCT could help differentiating mucoceles from acute appendicitis. In summary, a dilated appendix in MDTC scan that contains liquid, some showing a calcified wall (Fig. 12) in association with little or no stranding of the adjacent fat, should lead us to consider the possibility of an appendicular mucocele.

- **Appendicular tumors**: It includes a heterogeneous group of neoplasias with variable prognosis, being the major groups identified epithelial neoplasms, carcinoids and metastases, the later particularly from tumors located in adjacent structures. Appendicular tumors are infrequent, with an incidence of up to 1% of the appendectomies, usually being findings in pathology in patients originally suspected as having acute appendicitis. Among the tumors, the most frequent is the carcinoid type, mainly in women and rather young patients. Appendicular tumors are mainly localized in the distal third, are usually smaller than 1 cm and are histologically well defined. Because the treatment approach is variable, including right hemicolectomy in the most aggressive cases, the pre-operative suspicion is ideal.

- **Endometriosis**: the presence of endometrial tissue outside the uterine cavity affects approximately 10% of women in fertile age, it been described in the literature practically and may affect virtually all abdominal organs. In the cecal appendix, a prevalence of approximately 0.08 % has been described, most cases being incidental findings in pathology, a half of them affecting the medial third of the appendix while the other half affects its distal portion.

Endometriosis may be a casual finding in asymptomatic patients or may be the cause of an invagination of the appendix or even of an acute appendicitis, either by the obstruction caused by an endometrioma or by the hemorrhage itself localized between
the serous membrane and muscular layers causing edema and obstruction. The typical form of endometriosis of the appendix consists of small nodules in the appendix wall, histologically localized in the muscular-serous membrane and not affecting the mucosa. In MDCT scan we may identify a dilated appendix, without effect on the surrounding fat in those cases where there is no concomitant acute appendicitis (Fig.13). The treatment is surgery associated with hormonal treatment.

Images for this section:

Fig. 1
Fig. 2: A) Coronal CT image, Appendix with inflammatory signs with cranial and medial direction towards umbilicus.
Fig. 3: Atypical locations of the appendix A) Sagittal CT image showing ascending retrocecal vermiform appendix. Note the distal appendix located in right anterior pararenal space surrounded by fat stranding.
Fig. 4: Atypical locations. A) Axial and B) Coronal CT sections. Ascending retrocecal intraperitoneal appendix with tip lying in a subhepatic location. Dilated appendix with mucosal hyperenhancement and adjacent fat stranding.
**Fig. 5:** Atypical locations. Patient with suspected appendicitis and ultrasound inconclusive. A) Axial CT section shows dilated fluid filled appendix with intraluminal air. Note the tip appendix in presacral location B) Another patient with abdominal pain. Distal portion of appendix is located anterior to the rectum in midline. There are two appendicoliths inside and free fluid surrounding.
Fig. 6: A) Axial CT image. Gallbladder (vb) located in left upper quadrant and tail of the pancreas (p) in right hemiabdomen. B) Axial CT image. Terminal ileum (i) and ascending colon (ca) are in the left iliac fossa, surrounded by fat stranding. C) Coronal CT image. The Liver (h) located in the left upper quadrant, the stomach and the spleen in the right hemiabdomen and terminal ileum (i) in the left iliac fossa confirms the diagnosis of Situs Inversus Totalis. Left sided appendicitis with appendicolith, with adjacent free liquid and periappendicular fat stranding.
**Fig. 7:** Atypical location. Patient with lower abdominal pain and leukocytosis, but without conclusive findings in ultrasound. A) Coronal CT image shows anomalous position of ascending colon which is located in the left abdomen and small bowel situated mainly in right abdomen in relation to intestinal malrotation. B) Axial section confirms the pelvic cecum and the dilated appendix located anterior to the cecum. There is adjacent fat stranding. C) CT image slightly caudal demonstrates a dilated fluid filled appendix with an enhancing appendiceal wall consistent with acute appendicitis.
Fig. 8: DISTAL appendicitis. Abdominal ultrasound was performed in a patient with right ilioc fossa pain. It was not possible to identify the appendix completely. (A) Coronal, (B) sagittal and (C) axial images showing normal appendix with focal inflammation of appendiceal tip with one appendicolith inside. Thickening of the abnormal appendiceal tip with inflammatory changes in the periappendiceal fat.
Fig. 9: Stump appendicitis A) Coronal and B) Axial CT images. Patient with history of appendicectomy who presented right lower quadrant pain. Note infiltration of periappendiceal fat (arrow). Surgery confirmed the diagnosis of stump appendicitis. C) and D) Ultrasound images of another patient with appendicectomy. Tubular structure with layers corresponding to the appendiceal stump with hyperechoic adjacent fat suggestive of inflammatory changes were observed.
Fig. 10: Appendiceal diverticulitis. A) Axial and B) Sagittal sections. Appendix with thickened, irregular and hyperenhancement wall, surrounding by inflammatory involvement. Surgery was performed due the clinical suspicion and imaging findings suggesting acute appendicitis. Pathological anatomy of the surgical specimen showed diverticulitis and diverticulosis of the cecal appendix.
Fig. 11: Mucocele. A) Coronal and B) Sagittal CT images. Note fluid-filled distended appendix, typical of a mucocele, without wall thickening and fat stranding. Patient referred lower abdominal pain and was operated with the preoperative diagnosis of appendicitis. The pathological study of the surgical piece was compatible with appendiceal hyperplasia.
Fig. 12: Mucocele. A) Axial and B) Sagittal CT images. A very distended fluid-filled appendix is noted, with partially calcified wall and hypodense content compatible with mucocele, without inflammatory changes.
**Fig. 13:** Appendiceal endometriosis. Patient with a history of endometriosis consulted for right iliac fossa pain. A) Coronal CT section shows output of vermiform appendix. B) The appendix was increased in size, up to 14 mm (yellow line), however there is not involvement of surrounding fat. C) Axial CT images from the same patient demonstrates a dilated segment of appendix (arrow). Pathology demonstrates endometriosis of appendix.
Conclusion

Even thought acute appendicitis is the most frequent cause of surgical acute abdomen in young patients, there are disorders that may mimic this entity making difficult its diagnosis. Such difficulties may end in overdiagnoses and subsequent negative appendectomies or, on the contrary, underdiagnoses with the risk of complications or a torpid evolution. Knowledge of the differential diagnoses as well as the role played by ultrasound, and especially MDCT, is essential for the correct management of these patients.

Personal information

danimandich@gmail.com

References


