CASE REPORT

A case of xanthogranulomatous appendicitis in the female pelvis

Wai Heng Yung, MB BCh[1], Ching Hsia Caroline Siew, FRCSEd (Gen Surg)[2], Hwei Yee Lee, FRCPath[3], Hsien Min Low, FRCR[1]

[1]Department of Diagnostic Radiology, Tan Tock Seng Hospital, Singapore, [2]Department of General Surgery, Tan Tock Seng Hospital, Singapore, [3]Department of Pathology, Tan Tock Seng Hospital, Singapore

SUMMARY

Xanthogranulomatous inflammation is an uncommon form of chronic inflammatory process. Only a few isolated case reports of xanthogranulomatous appendicitis (XA) have been published. XA has nonspecific imaging findings and cannot be reliably differentiated on imaging from locally advanced malignancy. XA however follows a benign course and can potentially be treated with surgical resection.

INTRODUCTION

Xanthogranulomatous inflammation (XI) is an uncommon form of chronic inflammation. It can involve multiple organs and the most reported presentations include cholecystitis and pyelonephritis. Histologically, it is characterized by lipid-laden macrophages or histiocytes admixed with lymphocytes and plasma cells.

Xanthogranulomatous appendicitis (XA) has nonspecific imaging findings and may mimic a locally advanced or remnant malignancy as it cannot be reliably differentiated on imaging. XA however follows a benign course and can potentially be treated with surgical resection.

CASE REPORT

A 41-year-old Chinese female with no significant past medical history presented to the Tan Tock Seng Hospital, Singapore with a 2-week history of right iliac fossa pain associated with low grade fever. Subsequent pelvic ultrasound revealed a right pelvic mass. Because of the size, it was difficult to establish if this mass was of ovarian or uterine origin. As such, further evaluation with magnetic resonance imaging (MRI) of the pelvis was performed. This showed the mass to be a thick-walled, lobulated rim-enhancing cystic lesion. Thick, enhancing septations were also seen within the lesion. The fluid contents of the lesion demonstrated T2 hyperintensity and restricted diffusion. No discrete solid component was noted. The lesion was inseparable from the caecum.

Based on the findings of the MRI, the primary considerations were that of a neoplasm or inflammatory process arising from either the appendix or gynaecological organs. Computer tomography (CT) of the thorax, abdomen and pelvis was thus performed to exclude distant metastases. The CT scan also showed a complex cystic structure in the right iliac fossa, inseparable from the caecum and terminal ileum. No distant metastases were seen. Of note, the appendix and right ovary could not be identified on both CT and MRI. There was no inflammatory fat stranding surrounding the lesion. The differential diagnoses based on the imaging studies thus far remained as either an appendiceal or ovarian pathology, of which a neoplastic or inflammatory process was a possible cause.

The patient underwent a colonoscopy which did not reveal any colonic masses. However, the mucosa overlying the appendiceal orifice was noted to be abnormal. Diagnostic laparoscopy was thus performed which showed a mass posterior to the caecum. This mass was noted to involve the caecum and terminal ileum. The appendix could not be identified. The right ovary and fallopian tube were attached to the mass. The working diagnosis after the diagnostic laparoscopy was either an appendiceal abscess or neoplasm. Right hemicolectomy with en bloc right salpingo-oophorectomy was subsequently performed, one week after the initial pelvic ultrasound, in view of the inability to rule out an underlying neoplasm.

The appendix showed mucosal ulceration, and suppurative and xanthogranulomatous inflammation. The inflammation extended into the periappendiceal fat and into the right ovary and fallopian tube. No acid fast bacilli or fungal organisms were identified on special stains.

DISCUSSION

Xanthogranulomatous inflammation is a rare pathological finding that was first described in the kidney by Osterlind in 1944. It has also been reported in the gallbladder, kidneys, endometrium and female genital tract including the fallopian tube.[1] It usually presents as pyelonephritis and cholecystitis. Only a few isolated case reports of XA have been published so far.[2]

The pathophysiology of XI is not well understood. The proposed mechanisms include lymphatic obstruction, defective lipid transport as well as a immune response specific to *Escherichia* and *Proteus*.[3] On histology, XI is characterised by the development of foamy macrophages.

XA has variable imaging features. It can present as a heterogeneously enhancing mass or inflammatory process in

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Corresponding Author: Dr. Hsien Min Low
Email: Hsien_Min_LOW@ttsh.com.sg

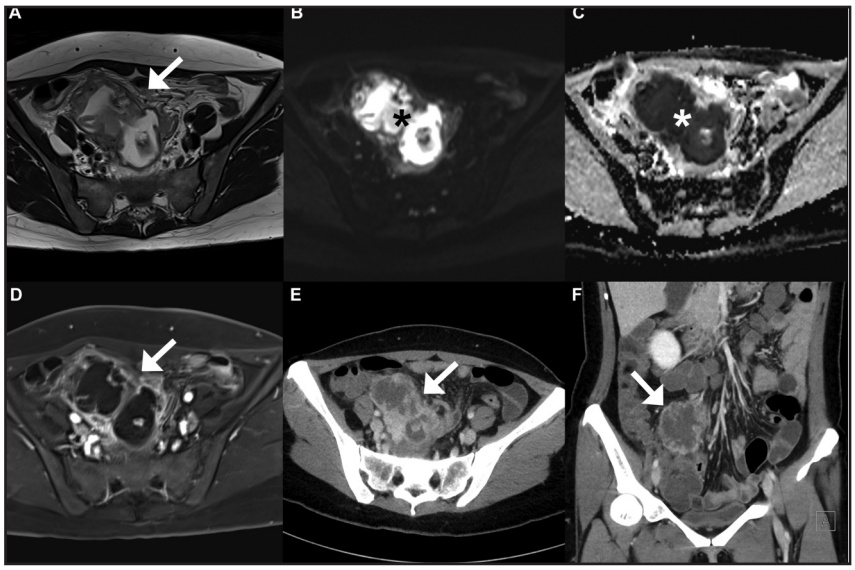

Fig. 1: MRI of the pelvis. (A) T2-weighted image reveals a heterogeneous solid cystic lesion in the right hemipelvis. (B-C) Diffusion weighted image demonstrates hyperintensity (black asterisk) in the contents of the lesion corresponding to T2 hyperintense areas. Abnormally low values on the apparent diffusion coefficient map (white asterisk) in the contents of the lesion confirm the presence of abnormal restricted diffusion. (D) Post-gadolinium T1-weighted image shows rim-enhancement of the complex cystic lesion. Of note, the appendix and right ovary could not be seen. Contrast-enhanced CT of the abdomen and pelvis in (E) axial and (F) coronal planes reveals a corresponding rim-enhancing complex cystic lesion (arrow). There is minimal surrounding inflammatory stranding. Of note, the appendix and right ovary could not be seen.

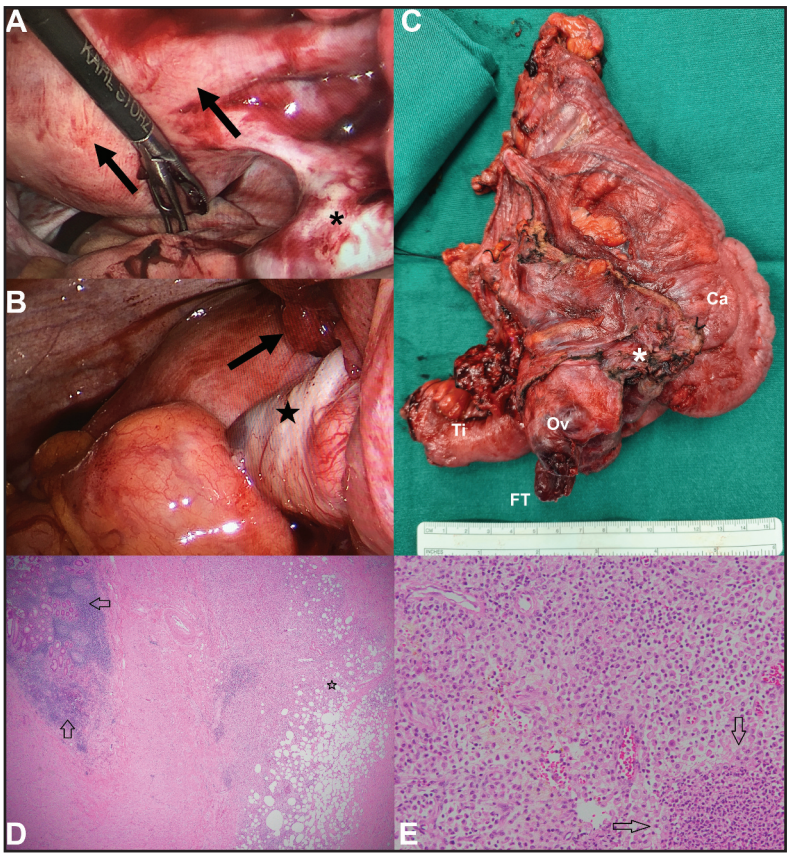

Fig. 2: Diagnostic laparoscopy showed an (A) inflammatory mass (black asterisk) posterior to the caecum (black arrows) which is being lifted. (B) The mass also involves the right fallopian tube (black arrow) and right ovary (black star). (C) Posterior surface of resection specimen demonstrates the appendiceal mass (white asterisk) densely adhered to the caecum (Ca), terminal ileum (Ti), right ovary (Ov) and fallopian tube (FT). (D) Low power view (H&E x20) shows the tip of the appendix on the left, with an area of xanthogranulomatous inflammation extending into the periappendiceal fat on the right. The open arrows show the appendiceal mucosa and the star marks the area of inflammation in the periappendiceal fat. (E) High power view (H&E x200) shows the xanthogranulomatous inflammation with sheets of foamy macrophages admixed with small lymphocytes and plasma cells. On the bottom right marked with open arrows is an area of suppurative inflammation, consisting of abundant neutrophils.

the right iliac fossa, as in our case. A few reports have also described XI being detected in patients who undergo a delayed, interval appendicectomy after an acute episode of appendicitis.[4] The described clinical course is similar to our case as the patient underwent definitive surgery approximately three weeks after onset of symptoms and one week after first presentation to the hospital.

Although this is a known entity, it is difficult to differentiate it from a locally advanced malignancy on imaging since the inflammatory process often extends to the surrounding structures or soft tissues, giving an appearance of an invasive malignancy.[5] Our case profoundly shows this difficulty in the female pelvis, where the main differential diagnoses encompass entities such as tubo-ovarian abscesses, perforated appendicitis, mucinous tumours of the appendix and ovary. These entities can potentially resemble one another on imaging.

Tubo-ovarian abscess

Tubo-ovarian abscess is a late complication of pelvic inflammatory disease (PID) which is most commonly seen in sexually active women. This can occur unilaterally or bilaterally as a result of inadequately treated PID. On imaging, tubo-ovarian abscesses appear as a complex, rim-enhancing fluid collection with surrounding inflammatory changes. The involved ovary is often obscured by the abscess and inflammation. Internal gas bubbles are specific sign of an abscess on imaging but are not common in tubo-ovarian abscesses.[6] A loss of definition of the uterine border and anterior displacement of thickened broad ligament may also help to distinguish a tubo-ovarian abscess from other causes of pelvic abscess.[7]

Perforated acute appendicitis

Perforated appendicitis can present in a variety of ways on imaging. A few features such as extra-luminal appendicolith, abscess formation, and discrete enhancement defect in the wall of the appendix are often described as specific features of perforated appendicitis.[8] However, perforated appendicitis can manifest as a complex-looking abscess in the right lower quadrant. At times, these processes can also obscure the adjacent right ovary which would make differentiation from ovarian pathologies difficult.

Mucinous tumours of the ovary and appendix

Mucinous tumours of the appendix and ovary often present clinically with vague, non-specific symptoms. The imaging features can vary depending on the stage of disease. Mucinous cystadenomas of the appendix can appear as an appendiceal mucocele with no solid component and can mimic the appearance of appendicitis.[9] Conversely, mucinous cystadenocarcinomas may present as a complex solid-cystic mass with enhancing solid components and septa. Mucinous cystadenocarcinomas have a higher chance of perforation resulting in diffuse ascites, peritoneal nodules and pseudomyxoma peritonei.[10]

Mucinous epithelial lesions of the ovary also present as a multiloculated, complex cystic lesion with thickened enhancing septa. The presence of solid components increases the suspicion of malignancy.[11] Given the similarity in imaging features between appendiceal cystadenocarcinomas and ovarian mucinous tumours, differentiating between the two entities can be difficult.

MRI is helpful to distinguish between abscesses and mucinous tumours. On MRI, the contents of abscesses typically show restricted diffusion due to the highly cellular and complex nature of purulent fluid. In contrast, the fluid contents of mucinous tumours demonstrate T2 hyperintensity, DWI hyperintensity and high ADC values (T2-shine through).[12]

The contents of the lesion in our case showed restricted diffusion which would allude to presence of an abscess. However, as there can be overlap in the imaging findings between an abscess and a necrotic or mucinous malignancy, diagnosis and curative treatment would still depend on surgical resection.[13]

Identification of a normal appendix or ovary on imaging is very important in narrowing down the differential diagnoses of solid-cystic lesions in the female pelvis. Unfortunately, solid-cystic lesions involving these organs can be large and extensive in the pelvis. As such, these structures may not be readily identified. In such scenarios, the definitive diagnosis can only be established intra-operatively or on histological analysis.

CONCLUSION

Xanthogranulomatous appendicitis is a rare disease that is difficult to diagnose on imaging alone. The imaging features of XA can resemble other inflammatory and malignant processes of the pelvis and is difficult to diagnose prospectively on imaging. Nonetheless, the condition should be considered in the differential diagnosis during the evaluation of pelvic masses and pathologies. The definitive diagnosis relies on surgical resection and histopathological examination of the mass.

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