Appendicular calculi and carcinoid tumor of the vermiform appendix: a case report

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Abstract

Two uncommon distinct pathology occurring in the same patient without any symptom related to the organ is fascinating and interesting. In this case transabdominal hysterectomy was done in a woman aged 38 years for dysfunctional uterine bleeding. In addition appendisectomy was done as the appendix was found inflamed. Gross examination revealed two calculi at the tip and histopathology confirmed carcinoid tumor of the appendix.

Key words: vermiform appendix; calculi; carcinoid tumor.

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Introduction

Weisfing was the first to make a correct preoperative diagnosis of appendicular calculus radiologically.1 In 1947 Felson and Bernhard² estimated that just over 100 cases had been reported; this number had increased to 120 by 19513, and to between 130 and 155 by 1957.4,5 Regarding neoplastic diseases, carcinoid tumors(CT) are the most common neoplasm of the vermiform appendix.6 The overall incidence of carcinoid tumors has been estimated to 1 to 2 cases per 1000 appendisectomies in surgical specimens.7 CTs are discovered usually during the course of another procedure.8 In this case there are two stones situated at the tip and CT is situated in the mid portion in the same vermiform appendix.

Case report

A 38 year old woman, was admitted into a private hospital with the history of dysfunctional uterine bleeding (DUB) and lower abdominal pain for 6 months. On examination she was anemic with low grade fever. The estimated Hb level was 6.2 gm/dL. Abdominal and transvaginal sonography(TVS) was performed which revealed mild bulky uterus with an uterine fibroid. Considering the patient's clinical condition surgery was performed. Total abdominal hysterectomy (TAH) with appendisectomy was done. The appendix was found inflamed with distension of the tip, which was solid and firm on palpation. Gross examination has shown vermiform appendix measuring 9.0x1.0x1.0 cm, cut section

revealed 2 stones (the larger one measures 1.0x0.8x0.8 cm) at the tip (Figure 1) and a mass measures 1.0x1.0x1.0 cm at the mid point of its length. The histological examination revealed a typical CT of the appendix (Figure 2) and tumor-free margin in all specimens. The tumor was consisted of small island of uniform cell with scant, pink granular cytoplasm and a round to oval stippled nucleus. The muscular layer is infiltrated by the tumor cell nests. The mitotic activity was insignificant. The patient's postoperative course was uncomplicated and she was discharged on the 4th postoperative day. Diagnosis of appendicular calculus is not particularly difficult when the condition is borne in mind. A straight X-ray film of the abdomen in every patient with undiagnosed abdominal symptoms reveals a characteristically lamellated calculus, usually in the right iliac fossa. It is readily distinguished from the mottled appearance of calcified mesenteric nodes; ureteric calculi lie in the axis of the ureter, and gallstones are not usually so low. In case of gallstones and ureteric calculi, cholecystography and intravenous pyelography may be very useful adjuncts respectively.

Discussion

The simultaneous presence of appendicular calculi and CT in the same appendix is a rare pathological entity. The great majority of patients with appendicular calculi may present as acute abdominal emergencies. Since the calculus will in most cases have lain in the appendix for months or years, gradually

increasing in size in a confined space, it is surprising that a history of previous attacks of colicky abdominal pain or of symptoms suggestive of 'grumbling appendix' is seldom present. Eventually the typical picture of acute obstructive appendicitis develops and prompt surgical intervention becomes necessary. Early surgical treatment is therefore indicated when an appendicular calculus is revealed, by intention, by chance, or by radiological examination of the abdomen.⁹

In a study carried out by Forbes GB in 1966,9 two thousand appendices were examined for evidence of calculous disease. One thousand eight hundred were specimens removed surgically and 29 of them harbored stonyhard calculi. The incidence of calculous disease in 1,000 consecutive appendisectomy specimens was 08%. An additional calculus was observed radiologically in a case of appendicular actinomycosis. All but five of the 30 calculus-containing appendices were acutely inflamed and 50% of these were gangrenous or perforated. Others have reported fistula formation as a complication of calculous disease.

From the reports in the literature it can be clearly seen that stone in the appendix is very prone to cause serious illness, often with perforation and generalized peritonitis. Some patients harboring a calculus in the appendix will be seen in a non-acute phase, sometimes with vague symptoms. Felson and Bernhard record the case history of a 39-year-old male who complained of low back pain. In the preceding year he had three attacks of epigastric pain, and one attack in the right lower quadrant. In this case the patient had history of low back pain.

In case of CT, the clinical presentation is similar to acute appendicitis, but the CT can be an incidental finding during surgical procedures. In this case CT was diagnosed incidentally during TAH. It was confirmed on histological examination of the removed appendix.

In 60 cases reviewed by Felson and Bernhard² most patients were between the ages of 10 and 30, and there was a male preponderance of nearly 4:1. In our case, the patient is a female and much older than the patients in Felson's study.

In the 75% of cases the tumor is localized at the tip of the appendix, in 20% and 5% cases, it affects the mid portion and the base respectively.10 The tumor's median diameter is 6 mm. 8,10 In our case, the diameter of the tumor was 1 cm and it was situated at the mid portion. Generally, carcinoid tumors located at the tip of the appendix and measuring less than 10 mm and usually mimics the clinical presentation of acute appendicitis, while tumors measuring more than 20 mm and located at the base of the appendix may present with clinical signs of peritonitis. 11,12 The site and the size rather than the depth, are used for the assessment of the tumor.8 The prognosis is directly related to the tumor size. Localized disease has an excellent prognosis. The prognosis of patients with metastatic CT is very poor. If the tumor is smaller than 2 cm and has perforated the serosa, the treatment of choice is appendisectomy, whatever the location. Tumors measuring 2 cm or more in diameter may have widespread metastases upon detection. Symptoms of the carcinoid syndrome as flushing, diarrhea, cardiac disease have been rarely reported and usually associated with liver or retroperitoneal metastases. 13,14 In our case, CT was localized and the patient had no symptoms related to carcinoid syndrome.

Most interesting and surprising is that the patient in our case harboring appendicular calculi and carcinoid tumor simultaneously.

Conclusion

Our case highlights the rarity of the location of these two uncommon pathology with an emphasis on the diagnosis of these entities to generate clinical awareness, early diagnosis and proper management, thus decreasing significantly morbidity and mortality.



Figure 1: Gross appearance of the appendix with calculi.

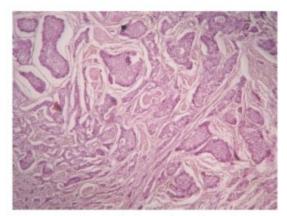


Figure 2: Histological section of carcinoid tumor of the appendix showing island of uniform cell (H&E stain x 400).

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