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Case Report

Segmental type of gallbladder adenomyomatosis – Case report and literature review

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ABSTRACT

Introduction: Adenomyomatosis of the gallbladder is a benign mural disorder characterized by a thickened wall, proliferation and distention of Rokitansky–Aschoff sinuses surrounded by proliferated fibromuscular tissue.
Aim: Overview of radiological imaging methods used to evaluate the segmental type of adenomyomatosis of the gallbladder.
Case report: The patient was admitted to the Emergency Department with typical symptoms of hepatic colic. Ultrasonography did not allow the exclusion of gallbladder cancer and diagnosis required clarification in Multidetector CT and MR with MR cholangiopancreatography. The surgical pathological specimen revealed segmental form of adenomyomatosis with cholelithiasis and chronic inflammation.
Results and discussion: Adenomyomatosis is not considered a pre-cancerous condition, but elevated intraluminal pressure, gallstones and chronic inflammation are risk factors for gallbladder cancer. The most common imaging methods used to diagnose adenomyomatosis of gallbladder are US and MRI with MRCP.
Conclusions: The segmental type of gallbladder adenomyomatosis with a tendency of cholelithiasis, and higher risk of gallbladder malignancies, is a direct recommendation for cholecystectomy. Despite improvements of diagnostic imaging methods differentiation of segmental adenomyomatosis from early gallbladder cancer still remains challenging.

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1. Introduction

Adenomyomatosis of the gallbladder (GBA) is a relatively common disorder that has been reported in up to 8.7% of cholecystectomy specimens, diagnosed usually in fifth to sixth decade of life. Considered as a benign disorder it may be asymptomatic and detected incidentally. The GBA pathogenesis is unknown. Usually it is characterized by excessive proliferation of the gallbladder mucosa that creates invaginations through the thickened muscular layer, known as Rokitansky–Aschoff sinuses (RAS). Based on the location and the spreading area different types of GBA have been described: the segmental type, localized fundal type, localized annular type surrounding midportion of gallbladder and the last diffuse type involving entire gallbladder.[1,2] The most common segmental type is composed of annular stricture dividing the gallbladder into to the ''fundal compartment'' with a thickened wall and the ''neck compartment'' with normal size gallbladder wall. In diagnostic imaging, GBA manifests as diffused wall thickening or a local mass with intramural cysts, diverticula and increments required to be distinguished from gallbladder carcinoma. The differential diagnosis also includes: adenomatous, hyperplastic and cholesterol polyps, xanthogranulomatous cholecystitis, rarely mesenchymal neoplasms, gallbladder metastases or true diverticulum of gallbladder fundus.[3] Identification of the RAS is the key point in diagnosing GBA on the basis of different imaging examinations.

2. Aim

Discussing the role of radiological imaging in evaluation and differential diagnosis of segmental type of gallbladder adenomyomatosis.

3. Case report

A 60-year-old patient was admitted to the Hospital Emergency Department with pain localized in the upper right abdominal quadrant diffused to the right shoulder, caused by dietetic error. Symptoms developed over a period of four days. The laboratory tests which indicated a slight inflammation were showing CRP – 6.25 mg/L and WBC within normal limits – 7000 mm^3. A similar abdominal pain incident was noticed a year ago, but then the pain subsided after taking painkillers and spasmolytic drugs. Ultrasound (US) examination showed a thickened gallbladder wall, the fundal part of which resembled diverticulum with calcified deposits (Fig. 1). Also single small intramural foci of increased echogenicity were visible. US examination was determined to be inconclusive and did not give the accurate diagnose. Multidetector CT (MDCT) examination confirmed fundal wall thickening, poorly calcified deposits and no other organ damage (Fig. 2). Magnetic resonance (MR) imaging with MR cholangiopancreatography (MRCP) examination showed intramural cysts and areas of focal signal loss corresponding to calculi (Fig. 3). After three days from admission of the patient to the hospital

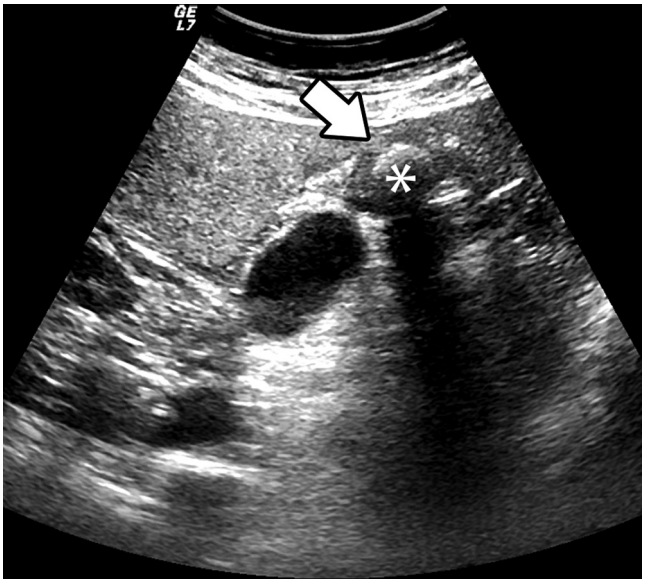

Fig. 1 – US examination in presentation B. Gallstone (asterisk) and thickened gallbladder wall (white arrow).

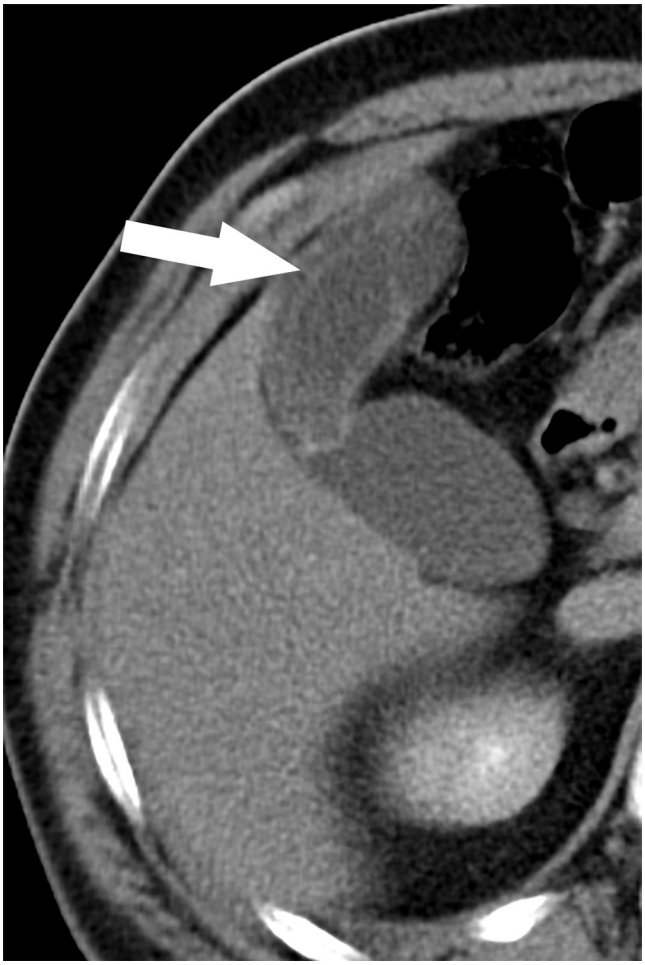

Fig. 2 – Axial CT scan after i.v. administration of iodine contrast and thickened gallbladder wall (white arrow).

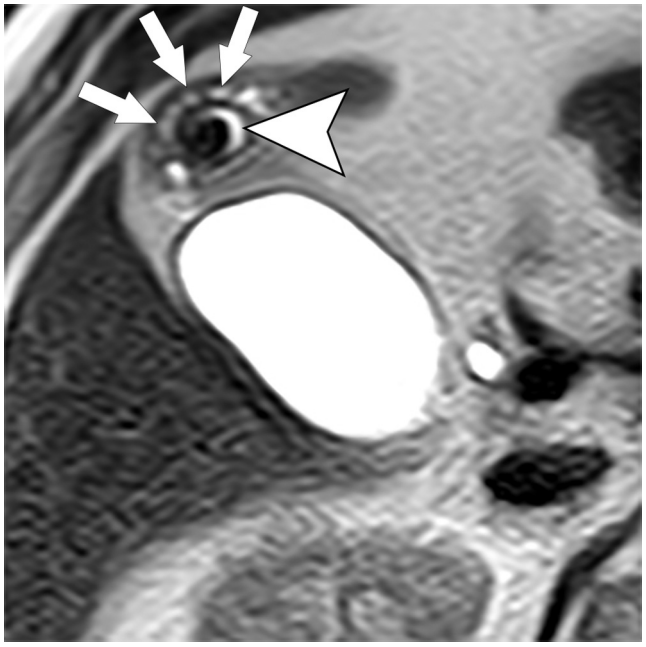

Fig. 3 – Axial T2-weighted image. Thickened gallbladder wall with a row of intramural cysts – ''The pearl necklace sign'' (white arrows) and gallstone (white arrow head).

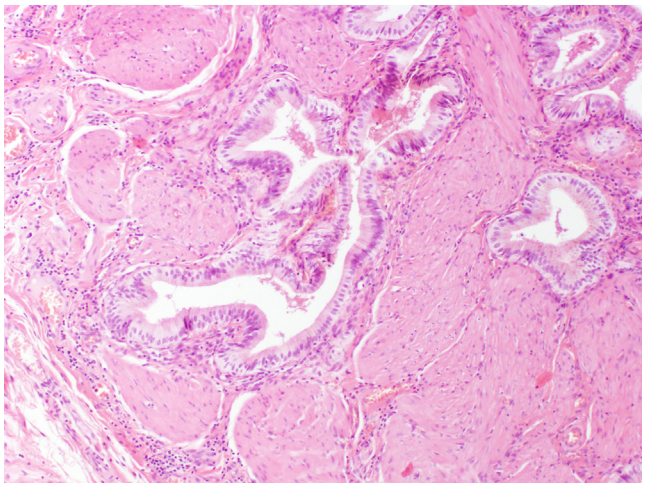

Fig. 5 – Photomicrograph of gallbladder wall (H&E stain). Cystic formations lined with glandular epithelium surrounded by hyperplastic smooth muscle fibers.

laparoscopic cholecystectomy was performed. The histologic and pathologic examinations revealed the segmental type of angiomyomatosis, some features of a chronic inflammatory response and focal low-grade glandular epithelial dysplasia (Figs. 4 and 5).

4. Results and discussion

Segmental GBA has a higher cholecystolithiasis prevalence (88.9%) due to a lithogenic environment caused by biliary stasis in the fundal compartment. Fundal or diffuse adenomyomatosis types are unrelated to cholecystolithiasis.[4] Although adenomyomatosis is not considered a pre-cancerous condition, patients with an elevated intraluminal pressure and with

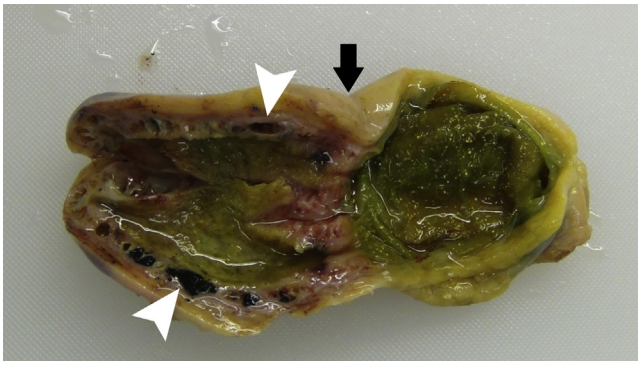

Fig. 4 – Surgical pathology specimen. Enlarged RAS (white arrow heads) and annular stricture (black arrow). Gallstone has been removed.

segmental type adenomyomatosis, gallstones and chronic inflammation are considered to be at risk for gallbladder cancer. In a case of gallbladder cancer associated with adenomyomatosis tumor arises predominantly at the fundal compartment of segmental-type adenomyomatosis. On the other hand localized adenomyomatosis may mimic gallbladder malignancy.[5]

Ultrasonography remains the primary examination method for the imaging of gallbladder diseases. A new solution, as high-resolution ultrasound (HRUS), allows for more accurate evaluation of the gallbladder wall compared with conventional US and increased incidence of GBA during the last few years. Symmetrical wall thickening, intramural anechoic cystic spaces, intramural echogenic foci, comet-like echoes, and twinkling artifacts are significantly associated with adenomyomatosis. However, the irregular thickening of the outer gallbladder wall, irregularity and thickening of the innermost wall layer which indicates the mucosa, loss of multilayer pattern of the gallbladder wall, focal discontinuity and intralesional vascularity are extensively associated with cancer occurrence.[6] For visualization of Color Doppler twinkling artifacts (CDTAs) 1.8 MHz color Doppler frequency is more highly recommended.[7]

MR imaging clearly demonstrates gallbladder wall thickening and reveals RAS as intramural lesions that are hyperintense on T2-weighted images, hypointense on T1-weighted images, and nonenhancing.[8] MRCP frequently depicts RAS within the thickened gallbladder wall as a row of high signal spots (''the pearl necklace sign''). The sign is highly specific (92%) for adenomyomatosis of the gallbladder and it is not seen in gallbladder carcinoma. Concomitant gallstones and biliary sludge are identified as areas of low signal intensity.

Post-gadolinium T1 images may be helpful because of their better demonstration of RAS within thickened gallbladder wall. The usefulness of an early enhancement pattern of gallbladder wall neoplasm is controversial.[9] The clinical application of diffusion-weighted MR imaging (DWI) has been

slowly expanding to gallbladder disorders. According to recent study, the mean ADC value is significantly lower in gallbladder cancer than in benign gallbladder diseases.[10]

CT has limited value in the detection and differentiation of adenomyomatosis and early gallbladder cancer. MDCT provides very high spatial but still insufficient contrast resolution. From a practical point of view adenomyomatosis can be recognized when thickened gallbladder wall contains small cystic-appearing spaces of water density. A CT "rosary sign" has been described, formed by enhancing epithelium within intramural diverticula surrounded by the relatively unenhanced hypertrophied muscular layer of gallbladder. Sometimes small calcified foci are seen within gallbladder wall.[1,11,12]

FDG PET is useful in differentiating benign from malignant lesions within the gallbladder. Limitations of the method are chronic inflammatory conditions related to adenomyomatosis with increased glucose metabolism which can lead to false-positive FDG PET findings.[13] In these cases FDG PET is recommended after reduction of the serum level of C-reactive protein.[14]

According to treatment algorithm developed by Pellino et al. symptomatic patients with GBA should undergo surgery. In asymptomatic patients management depends on the different GBA types. Pellino recommends cholecystectomy to all patients with segmental or diffuse GBA types because of risk of coexisting malignancies and difficult interpretation in diagnostic imaging and "wait-and-see" strategy for localized and fundal types.[15] Selection of the appropriate surgical technique in case of difficulties in the differentiation of GBA and early cancer is still being discussed. Preoperative suspicion of gallbladder cancer is generally considered to be a contraindication to laparoscopic cholecystectomy. On the other hand avoiding open surgery is a great benefit in those patients who are finally confirmed to have benign lesions. The key is the operation planning on the basis of combination of expertise diagnostic imaging techniques (transabdominal, endoscopic or laparoscopic US, CT, MR) and intraoperative pathology consultation. In a case of benign lesion or gallbladder cancer in stage pT1a laparoscopic cholecystectomy alone is sufficient. In order to avoid potential tumor spread into the peritoneal cavity during the extraction of resected gallbladder the procedure requires extra instrumentation like a protective bag. For pT1b and pT2 frozen biopsy-proven cancer, portal lymphadenectomy is strictly recommended. Laparoscopic locoregional lymphadenectomy requires more experience than open surgical lymphadenectomy. Patients with pT3 require open abdominal surgery.[16]

5. Conclusions

The exclusive criteria of the segmental type of GBA are unique morphology, a tendency to cholelithiasis, usually symptomatic process of progression, and a higher risk of gallbladder malignancies. The most commonly used and the most effective diagnostic imaging methods for identification, analysis and observation of GBA are US and MR. However the clinico-radiological differentiation of segmental adenomyomatosis from early gallbladder cancer still remains challenging.

Conflict of interest

None declared.

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