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Unveiling Diffuse Gall Bladder Adenomyomatosis: A Rare Case Report With Insights Into A Distinctive Condition

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1. Abstract

Gallbladder adenomyoma / adenomyomatous nodule / adenomyomatosis is a benign condition characterized by mucosal invaginations within the gallbladder wall with or without proliferation of smooth muscle fibers. Gallbladder adenomyoma is often asymptomatic and discovered incidentally during imaging studies performed for other unrelated conditions. It can mimic malignancy on radiological findings, creating a diagnostic dilemma. The management and outcome of adenomyoma and malignant lesions of gall bladder are completely different. We present here a rare case of 52-year-old female diagnosed as Gall bladder adenomyoma in histopathological examination highlighting the insights in to this benign distinctive condition. Therefore, it is essential to completely understand the pathognomonic and histopathological features of gallbladder adenomyoma in order to accurately diagnose this condition.

2. Keywords:
Adenomyoma, Gall bladder, Benign, Adenomyomatosis

3. Introduction

Gallbladder adenomyoma/ adenomyomatous nodule is a relatively rare but benign condition characterized by mucosal invaginations within the gallbladder wall with proliferation of smooth muscle fibers [1,2]. Diverticular abnormalities called Aschoff-Rokitansky (RA) sinuses are common in chronic cholecystitis. Some authors believe that adenomyomas are exaggerated phenomenon of Aschoff-Rokitansky sinuses [1,2]. RA sinuses may sometimes exhibit reactive epithelial atypia, which can be extremely severe and misinterpreted for Invasive cancer. Conversely, Cholelithiasis and Cholecystitis are frequently associated with gallbladder carcinomas, which can have a deceptively innocuous appearance. [1] Prevalence of adenomyoma in cholecystectomy specimens is estimated between 1% to 9% and the incidence increases after the age of 50 [3]. Studies show different incidences for men and women, but some publications also claim similar incidences [3]. Regardless of whether these adenomyomatous nodules in gallbladder are a sign of persistent damage or just an amplification of Aschoff-Rokitansky sinuses or a developmental defect, the etiology and pathogenesis of this condition are still not well understood [2,4]. This condition has unique histological features, understanding its nature and differentiating it from other gallbladder lesions especially adenocarcinoma, is crucial for its diagnosis and proper management. This case report explores the key aspects of gallbladder adenomyoma, providing a comprehensive overview of the various terminologies used, its clinical significance, diagnostic histopathological features and appropriate management strategies.

4. Case Report

A 52-year-old female came to the OPD with complaints of intermittently recurring upper abdominal pain for one year. She also complained of vomiting for 2 weeks. Clinical diagnosis was given as cholecystitis with cholelithiasis. USG Abdomen revealed calculous cholecystitis and Grade 1 fatty liver. MRCP revealed diffuse circumferential gallbladder wall thickening involving the body of the gallbladder with multiple cystic spaces showing restricted diffusion and Cholelithiasis. Cholecystectomy was done and sent for Histopathological examination. Gross examination revealed Cholecystectomy specimen measuring 10 x 5.5 x 2 cm. External

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surface shows irregularly dilated and distended gall bladder. Cut Surface shows thickened gall bladder wall with multiple small cystic spaces. A large cystic space is seen near the fundus (Fig I). 3 grey yellow stones are noted. Gall bladder mucosa is velvety and bile stained.

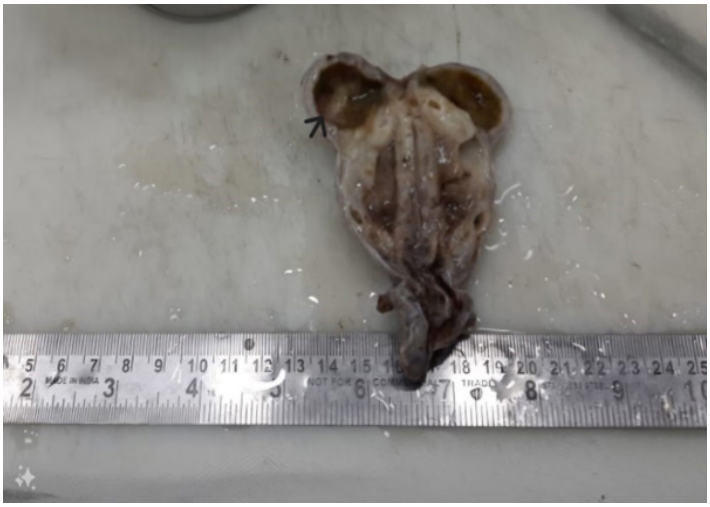


Fig I: Gross specimen shows markedly thickened gall bladder wall with multiple dilated cystic spaces. Note the large adenomyoma (near the fundus) (black arrow).

Multiple representative bits were taken. Microscopic examination revealed features of chronic calculous cholecystitis with hyperplastic mucosa (Fig II), hyperplastic muscle bundles with entrapped diffuse, dilated and irregular glandular structures lined by columnar epithelium ((Fig III, IV, V). Tiny islands of pyloric gland metaplasia is also noted (Fig VI). Final Impression was given as Adenomyoma (diffuse adenomyomatosis) of gall bladder with Chronic Calculous Cholecystitis.

Fig II: Low power view (10x) showing hyperplastic mucosa with epithelial lined glands in the subserosa and muscle layer (H & E stain).

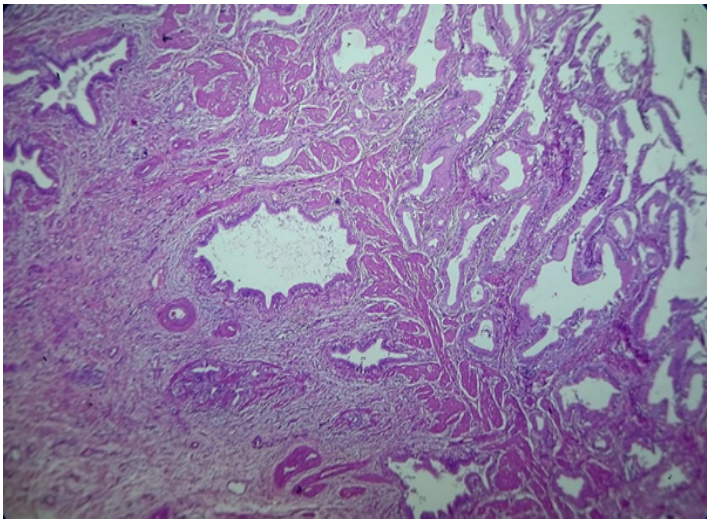


Fig III: Low power view (10x) of fundic adenomyoma shows multiple irregular, dilated and branching (black arrow) glands with prominent smooth muscle extending deeply into the gall bladder wall. (H & E stain).

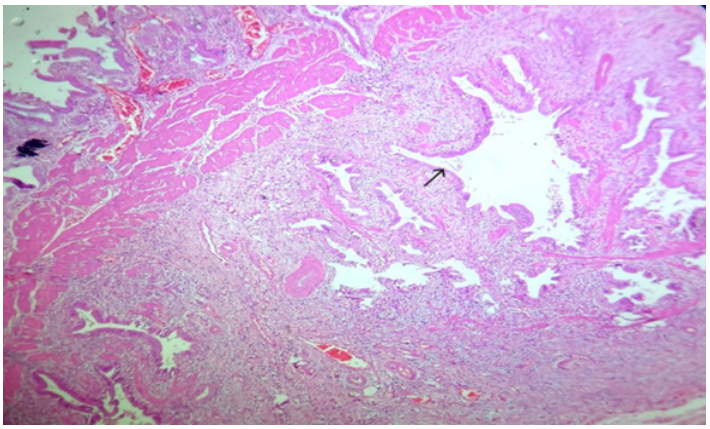


Fig IV: Low power view (10x) shows Hyperplastic muscle with chronic inflammation and haphazardly arranged glandular structures. (H & E stain).

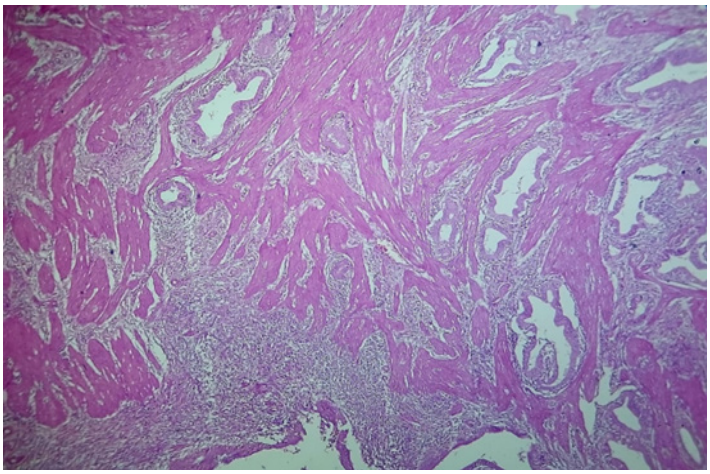
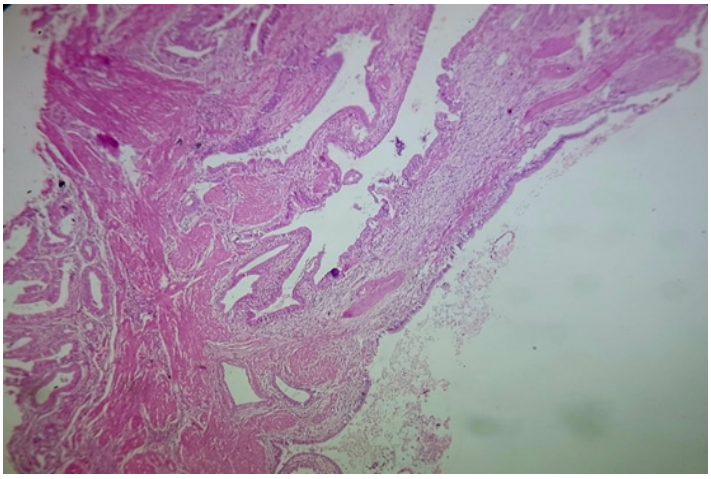


Fig V: Low power view (10x) shows Diffuse adenomyomatosis extensively involving gall bladder wall. (H & E stain).



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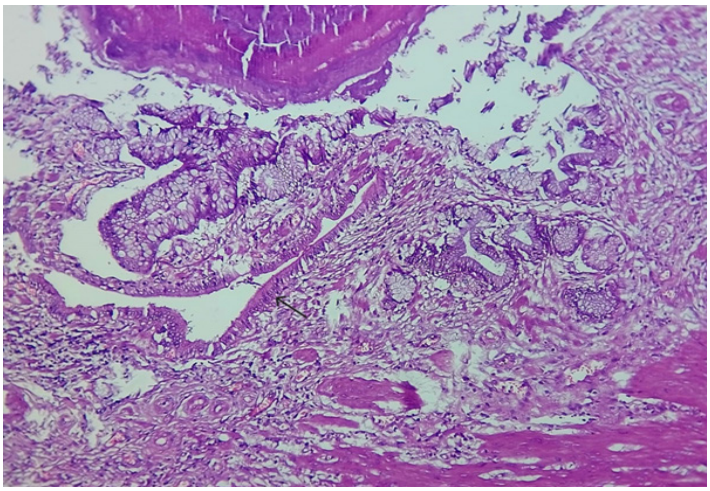
5. Discussion

Adenomyoma or Adenomyomatous nodules is characterized by cluster of cystically dilated glands in the gallbladder wall that are occasionally combined with prominent muscles. Studies show that the occurrence in males and women varies [3,5]. Most frequently, they are seen in the fundic area [2, 6]. They create a tiny isolated mass or a band of trabeculated thickening of the gallbladder wall [2]. The more widespread, extremely rare variant of this condition is referred to as "adenomyomatosis" as in our case which is a very rare occurrence (Fig VI).

Fig VI: High power view (40x) shows Microlith and pyloric gland metaplasia in the adenomyoma. Note the benign epithelium lining the glands without cyto-nuclear atypia. (Black arrow). (H & E stain).



5.1. Terminology and Controversies:

Various terms (hyperplastic adenomyosis, adenofibromyoma, proliferative glandular sickness) were used to refer to this proliferative condition until the 1960s till Judas simplified the phrase and gave it the label gallbladder adenomyomatosis [3]. While some believe it to be an exaggerated version of RA sinuses, in many cases, there is no communication with the surface mucosa, no additional sinuses are present. Furthermore, even though the term suggests a myoid process, the muscular component is sometimes absent or very little. But, our case has myoid component and associated Chronic Cholecystitis and Cholelithiasis. Some experts argue to call them adenomyomatous nodules since they are not real neoplasms [7]. Some authors suggest that Adenomyomatous hyperplasia, adenomyoma (when localized) and adenomyomatosis (when diffuse) are impressive but rather erroneous terms that have been used to denigrate exaggerated cases of gallbladder diverticulosis. The term, Adenomyomatous hyperplasia does not indicate true epithelial hyperplasia [2], but it may harbor dysplasia within it [4]. Our case also showed epithelial hyperplasia focally but no dysplasia is seen in our instance.

5.2. Etiology and Pathogenesis

The exact cause of gallbladder adenomyoma remains largely unknown. However, several theories suggest that smooth muscle hyperplasia and mucosal invaginations are believed to be adaptive responses resulting from chronic irritation or inflammation of the gallbladder wall [1]. It was initially recognized as a precancerous lesion; but recent studies consider it as a benign alteration [7]. Correlation with acquired wall motility as a consequence of increased endoluminal pressure due to cholelithiasis has also been postulated and further research is needed to fully understand the pathogenesis of this condition [4]. Gallstones and persistent cholecystitis have been linked in our case, corroborating these findings.

5.3. Clinical Presentation

Gallbladder adenomyoma is often asymptomatic and discovered incidentally during imaging studies [8]. When symptoms do appear, they are usually nonspecific, as in our case and may include abdominal discomfort [6,9]. Furthermore, adenomyoma is typically found during histological evaluation of the samples of individuals undergoing surgery for symptomatic cholelithiasis. Thus, clinical presentation of this condition might also be vague and misleading, as Teelucksingh S. et al. explain, and only resume in a persistent discomfort [9].

5.4. Imaging studies

Ultrasound (US) (High resolution) is the preferred imaging modality for preoperative diagnosis 20. There are three known morphological types [3]:

1. Segmental: they divide the gallbladder body into two parts in the form of rings. (as seen in our instance)

2. Fundal: present with Local thickening.

3. Diffuse type: diffuse irregularly thickened gallbladder wall. In our case, both the diffuse (a very unusual appearance) and segmental forms (found in the fundus) are present. In cases where the diagnosis is uncertain, MRI (cholangio-MRI) is necessary [3]. The "pearl necklace" sign shown in MRCP was regarded as pathognomonic for gallbladder adenomyomatosis [7] and its sensitivity is 80%, in a research done by Haradome et al [10].

5.5. Histopathology

Grossly, adenomyomatous nodules might result in tiny cysts or trabeculations that resembles a sieve [1]. This can present as polypoid lesion as well [4,11]. A sharply confined lesion that can resemble an adenocarcinoma both radiographically and grossly is seen in localized type, which can affect any part but is typically seen in the fundus [6]. Often, cysts in the GB wall can also arise from the Luschka ducts and tubulocystic variety of biliary adenocarcinoma, microscopic examination is definitely required in order to rule out these gross differential diagnoses. The lesion in microscopy consists of cystically dilated glands, within hypertrophic smooth muscle (Fig III, IV); the epithelium may show metaplastic or reactive changes, similar to our case and these changes are seen in the entire wall of the gall bladder (Fig VI). (diffuse adenomyomatosis)

5.6. Differentiation from Neoplastic process

In addition to these lesions simulating adenocarcinomas due to their pseudo invasive appearance, in situ or invasive carcinoma may be associated with them [1,5]. GA is not regarded as a premalignant lesion [3,5] as it lacks intrinsic malignant potential [12] and has no extrahepatic spread. Many

researches have observed that, carcinomas may occasionally be limited only to the adenomyomatous lesions [2]. In certain situations, GA may mimic intraductal papillary mucinous neoplasm if they exhibit widespread dysplastic changes and papillary structures [13].

5.7. Perineural invasion

It should be mentioned that glandular components of adenomyomatous nodules impinge on the nerves and resemble Intraneural and Perineural invasion [14]. Thus the gallbladder is now one more organ in which this phenomenon can occur in the absence of malignancy [7].

5.8. Management

Cholecystectomy is reserved for symptomatic patients or inconclusive imaging findings [1,6]. Histopathological examination still remains the gold standard method for its precise diagnosis as in many situations imaging studies alone cannot rule out an associated malignancy [6]. Frozen section should be done during surgery in order to rule out gallbladder cancer (GC) in doubtful cases [15].

6. Conclusion

Gallbladder adenomyoma/adenomyomatosis is a benign condition that is usually misdiagnosed both clinically and radiologically. Its radiological results and clinical presentation are deceptive. For proper patient care, it is crucial to distinguish between an adenomyoma and an early gall bladder cancer. Our rare case report presents this unique entity's insights with decision making algorithm and emphasizes the need for histopathological evaluation. However, Continued research and advancements in imaging technologies will definitely enhance the understanding and management of this unique gallbladder condition.

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