

# Adenomyomatosis of the gallbladder in an adolescent, a very rare condition

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## ABSTRACT

Adenomyomatosis of the gallbladder is an acquired, degenerative disease characterized by epithelial proliferation with hypertrophy of the muscularis layer with forming of sinus tracts, termed Rokitansky-Aschoff sinuses. Adenomyomatosis is diagnosed mainly by ultrasonography. The pathogenesis, pathology, and indications for surgery in this condition are not well understood. It is an extremely rare condition in children. We present a case of a 17-year boy with adenomyomatosis of the gallbladder successfully managed with laparoscopic cholecystectomy,

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## INTRODUCTION

Adenomyomatosis of the gallbladder (ADMG) is an acquired, degenerative disease characterized by localized or diffuse epithelial proliferation with hypertrophy of the muscularis layer and invagination of the mucosa through the muscularis, forming intramural diverticula termed Rokitansky-Aschoff sinuses.<sup>1,2</sup> ADMG is diagnosed mainly by ultrasonography (US).<sup>2,3</sup> The pathogenesis, pathology, and indications for surgery in this condition are not well understood, especially in children.<sup>2</sup> It is found in up to

5 % of cholecystectomy specimens<sup>4</sup> in adults, nevertheless it is an extremely rare condition in children.<sup>5</sup> We present a case of adenomyomatosis of the gallbladder successfully managed with laparoscopic cholecystectomy.

## CASE REPORT

A 17-year old boy looked for medical attention at a paediatric centre due to syncope, loss of body weight, and lumbar pain. US showed a mass located in hilar area of the liver; a computed tomography was performed, revealing a homogenous, hypodense, well-demarcated lesion 60 x 90 x 92 mm. As differential diagnosis enlarged gallbladder, dilated common bile duct, lymphoma, tumor of other origin or intramural hematoma were considered. The patient was referred to our hospital for further investigation.

In clinical history there was recurrent abdominal pain. The boy was hospitalized few times - at the age of 3 (diarrhoea), at the age of 10 (only unspecific abdominal pain), and at the age of 11 (gastritis antralis was diagnosed). The boy was treated in conservative manner. Despite the fact the boy was hospitalized few times, ultrasonography was performed only once, at the age of 7, showing "shadow" in the liver, with no further investigation and treatment. No other hospitalizations, symptoms and essential diseases were noted.

In our ward only moderate tenderness of the abdomen was observed. There were no significant findings in laboratory tests.

US showed an enlarged gallbladder of 94 x 62 mm with wall thickening to 30 mm and hyperaemia. The lumen was hourglass shaped. Next to its neck a hyperechogenic 5 mm structure appeared without acoustic shadowing, suggesting a polyp or uncalcified stone (*Figure 1*).

A cholangio-MRI revealed a gallbladder with dimensions 90 x 60 mm, containing two "cystic structures" of 23 x 35 mm and 26 x 25 mm connected by a narrow canal 16 mm long. Protrusions at the gallbladder fundus were apparent, interpreted as Rokitansky-Aschoff sinuses, and thickening of the gallbladder up to 30 mm (*Figure 2*). Contrast showed enhancement.

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The boy was qualified for laparoscopic surgery. Intraoperatively, a significantly enlarged gallbladder with a wall thickened up to 30 mm, mainly in the area of the fundus and the main body, was found. The posterior wall of the gallbladder displayed many adhesions to the liver. The gallbladder was excised.

Postoperative drainage of the peritoneal cavity was removed after two days. There were no intra- or postoperative complications. On histopathologic examination adenomyomatosis was confirmed with hyperplastic chronic inflammation and exacerbation, focally phlegmonous with focal adenomyoma.

## DISCUSSION

Gallbladder disease is still relatively rare in children and most of the pathologies consist of inflammation of the gallbladder and cholelithiasis.<sup>1,6</sup> In recent years there has been an increase in the incidence of gallbladder disease (including gallstones), mainly due to diagnostic improvements and the rise of childhood obesity.<sup>6,7</sup> ADMG was first described in 1960 by Jutras, and since then has increasingly been reported in adults,<sup>1,6,8</sup> nevertheless, in children it remains extremely rare.<sup>1,3,9</sup>

ADMG is divided into three types: generalized, localized, and segmental. Generalized adenomyomatosis is a diffuse thickening of the gallbladder wall with intramural cystic spaces.<sup>3</sup> Sometimes the gallbladder in generalized ADMG has a honeycomb appearance.<sup>3</sup> According to Eroglu<sup>3</sup> the localized type is the most prevalent,

usually with a single nodule (adenomyoma) in the fundus, which projects into the lumen.<sup>3,5</sup> The segmental type causes annular thickening of the wall, which can result in focal strictures. If the stricture occurs in the centre of the gallbladder, the organ may have an “hourglass” appearance.<sup>3</sup> In our case the wall of the gallbladder was thickened up to 30 mm (the normal gallbladder wall is not thicker than 1 mm<sup>8</sup>) with an hourglass appearance in MRI.

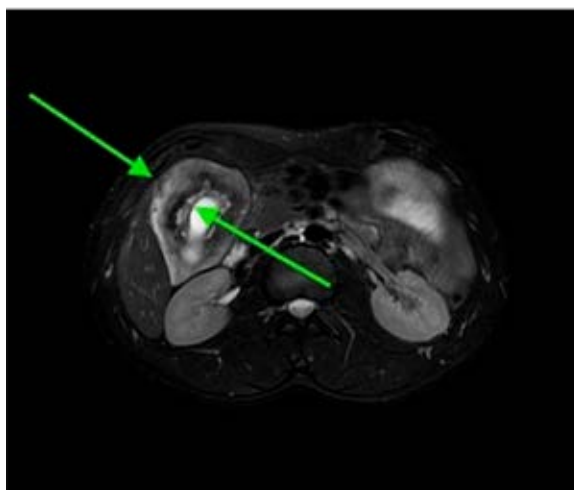
To the best of our knowledge, only ten cases of this disease have been described in children. The localized type was presented in two reports<sup>2,6</sup> (20 %) as well as the segmental type<sup>10,11</sup> (20 %). Although the localized type is most common, in the small group of children observed, the diffuse type was found in most cases, with five described by different authors<sup>1,3,5,8,9</sup> (50 %). Our case of the segmental type would be the third in the available literature. Alapati and Braswell<sup>12</sup> also presented a patient with spontaneous resolution of ADMG but without an indication of type.

Usually adult patients are asymptomatic, or symptoms are moderate. If symptoms occur, they are similar to cholelithiasis, usually self-limiting. Abdominal pain is mostly unspecific, usually localized in the upper right quadrant and epigastrium. Other symptoms such as vomiting, nausea, dyspepsia, or intolerance to fats are also observed.<sup>4</sup> In children unspecific and recurrent abdominal pain was mainly reported,<sup>1,3,6,9-11</sup> acute abdominal pain with vomiting and fever was less frequent.<sup>5,8</sup> In two

FIGURE 1. Hourglass shape of gallbladder (thick arrow showing annular thickening in the centre) with hyperechogenic structure (thin arrow) without acoustic shadow next to its neck



FIGURE 2. MRI showing thickening of the gallbladder wall up to 30 mm (thickened wall between arrows)



asymptomatic patients diagnosis was made incidentally by ultrasound imaging, one with Beckwith-Wiedemann syndrome,<sup>2</sup> and the other was a neonate.<sup>12</sup> In our patient the symptoms were highly unspecific. Despite the fact that most children in this series were symptomatic, according to Akcam<sup>1</sup> most patients with ADMG might not be diagnosed before adulthood because it shows no symptoms before the development of stones or inflammation.<sup>1</sup>

Laboratory tests are often normal, and there is no serological marker to detect ADMG.<sup>4</sup>

Ultrasonography seems to be the most accurate tool for routine diagnosis. The sonographic presence of intramural diverticula (Rokitansky-Aschoff's sinuses), seen as anechogenic (bile filled) or echogenic (biliary sludge or gallstone filled) foci within the thickened wall, with or without acoustic shadows, is typical for ADMG.<sup>6,8</sup> US is reported to display accuracy as high as 66 % in diagnosing ADMG, while MRI scores are 93 %.<sup>4</sup> However, US is a non-invasive examination and easily repeated, which makes it the ideal tool for follow-up,<sup>4</sup> as well as for little children, as it does not require general anaesthesia. In some cases additional studies are made, but according to the literature, diagnosis of ADMG in children was generally established with US and MRI (Table 1). Parolini et al. claim that cholangio-MRI should be performed before surgery as major variations and anomalies in the biliary tree must be identified to prevent severe lesions to the common bile duct. Furthermore, cholangio-MRI can detect stones in the choledocus.<sup>5</sup>

In our case US performed in our centre was enough to determine the diagnosis, however MRI also contributed to the diagnostic process.

Gallbladder carcinoma is often responsible for wall thickening, which is often irregular.<sup>4</sup> A well-differentiated gallbladder carcinoma with mucin production can have cystic components that may mimic adenomyomatosis,<sup>13</sup> thus distinguishing between gallbladder carcinoma and ADMG may be difficult.<sup>4</sup> Furthermore, stones and chronic inflammation secondary to ADMG may lead to dysplasia and cancer.<sup>2</sup>

In the ten published cases of this disease most children underwent surgery. Different authors agree that symptomatic patients should be operated on.<sup>4-6,8</sup> The question is whether asymptomatic patients should undergo cholecystectomy. Only Zarate et al.,<sup>2</sup> and Alapati and Braswell<sup>12</sup> chose to observe patients. Alapati and Braswell suggested spontaneous resolution

of ADMG in an infant, nevertheless no one has yet described such clinical cures in children, which raises the question of whether the authors actually observed ADMG. The rest of the patients in the series were symptomatic, making a course of action obvious.

Parolini et al. claim that conservative treatment and ultrasonographic monitoring should be reserved only for patients with clear contraindications to surgery.<sup>5</sup>

According to Cetinkursun when surgical treatment is chosen, the type of operation should be selected based on the patient's clinical situation and the surgeon's experience;<sup>8</sup> however Alberti et al. claim that laparoscopic cholecystectomy should be the treatment of choice.<sup>6</sup>

To the best of our knowledge, no one has previously described carcinoma combined with ADMG in a child. Nevertheless, there could be difficulties in distinguishing between carcinoma and ADMG. Thus, we retain our position that asymptomatic patients should undergo surgery to avoid neoplastic changes, however the question arises, when is the best time to perform a cholecystectomy? The symptoms of ADMG are mostly right upper quadrant abdominal pain, secondary to gallstones and inflammation,<sup>1</sup> which can make the operation more difficult and provide another reason to qualify an asymptomatic patient for surgery before symptoms occur. It is also an advantage that histopathologic examination gives a definitive diagnosis. Yet many questions remain before a clear route is found for proceeding with every case of ADMG in children. ■

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TABLE 1. Additional studies

Ref.	Ultrasonography	Magnetic Resonance	Others
Alberti, et al. <sup>6</sup>	Small and shrunken gallbladder with echogenic nodule next to the neck without acoustic shadow.		Technetium 99m HIDA liver scan + percutaneous transhepatic cholecystocholangiography
Cetinkursun, et al. <sup>8</sup>	Small and multiseptated gallbladder with thickened wall.	MRCP- a small gallbladder with lobulated mucosa and multiple heterogenous hyperintense cysts but no stone in the gallbladder.	Abdominopelvic computerized tomography. Normal parenchymal structure of abdominal organs and no primary or satellite cyst to confirm dissemination of hydatid disease.
Zani et al. <sup>10</sup>	No data		
Akçam et al. <sup>1</sup>	Thickening of the gallbladder wall, demarcated echogenic areas parallel to the wall of the gallbladder.	MRCP- grapelike cluster of shapes in the gallbladder, a long cystic duct.	
Zarate, et al. <sup>2</sup>	The gallbladder wall with echogenic foci.		
Alapati and Braswell <sup>12</sup>	A speckled appearance of the gallbladder with numerous echogenic foci causing "ring down artifact".		Followup ultrasound 8 months later, demonstrated resolution of the adenomyomatosis.
Parolini, et al. <sup>5</sup>	Diffuse thickening of the gallbladder, with multiple anechogenic nodular areas mainly localized in the fundus and in the body.	MRI -thickening of the gallbladder and the presence of multiple endoluminal irregular filling-defects with enhancement using a contrast dye. A tortuous cystic duct with an increased caliber (5 mm) was also evident.	
Eroglu, et al. <sup>3</sup>	Thickening of the wall of the gallbladder neck and infundibulum, including multiple cystic areas exhibiting demarcated echogenic areas parallel to the wall of the gallbladder.		
Agrusti, et al. <sup>11</sup>	Infundibulum with normal wall, central portion with wall thickening and microgallstones within the muscular layer, gallbladder fundus with a mild concentric wall thickening and small gallstones		
Eda, et al. <sup>9</sup>	No data.	MRCP - thickening of the gallbladder wall, multiple heterogeneous hyperintense cysts of the gallbladder fundus, and no dilatation of the bile duct.	Endoscopic retrograde cholangiopancreatography - pancreaticobiliary maljunction.
Our case	Enlarged gallbladder with wall thickening to 30 mm and hyperemia. Hourglass shape. 5 mm hyperechogenic structure without acoustic shadow next to neck.	MRCP - enlarged gallbladder, containing two "cystic structures" connected by a narrow canal. Rokitansky-Aschoff sinuses. Thickening of the gallbladder up to 30 mm.	Enhancement after intravenous contrast.

MRCP: Magnetic Resonance Cholangiopancreatography.

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