CASE REPORT

Ciliated foregut cyst of the gall bladder: the largest ever described

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SUMMARY

A 78-year-old man presented to the accident and emergency department with acute abdominal pain. A CT scan done to investigate the pain showed an intra-abdominal abscess medial to the caecum and an incidental exophytic lesion in the gall bladder. This was excised during surgery and sent for histological examination. Histology showed a ciliated foregut cyst of the gall bladder. A literature review revealed that this was the 13th case worldwide, as ciliated foregut cysts are usually found either above the diaphragm or in the liver. It was also the largest cyst described, with 45 mm diameter. So far none of these cysts found in the gall bladder have shown malignant transformation, though those found in the liver have been known to become malignant. Due to histological similarity, excision of ciliated foregut cysts of the gall bladder should be considered as there is still a risk of malignant transformation.

Figure 2 CT cross-section showing a cystic structure in the gall bladder.

BACKGROUND

Ciliated foregut cysts of the gall bladder are a rare entity. At this point in time, there is no consensus for treating these cysts, and none of those described were malignant. However, their histological similarity to ciliated foregut cysts of the liver, which may undergo malignant transformation, implies that a closer look at these cysts is required. If enough evidence is gathered, a decision may be made regarding the appropriate treatment.

CASE PRESENTATION

A 72-year-old man presented to the accident and emergency department with worsening abdominal

pain over 1 month, becoming more acutely painful over 1 week prior to admission, associated with nausea, decreased appetite and fever. His medical history included hypertension, hypothyroidism, benign prostatic hypertrophy and atrial fibrillation for which he was on warfarin. On examination, he was febrile with abdominal tenderness in the right iliac fossa with guarding.

INVESTIGATIONS

A decision was made to perform a CT scan, which showed a large intraperitoneal abscess medial to the caecum. A coincidental finding was an exophytic lesion was seen on the anteromedial aspect of the gall bladder wall for which an ultrasound was done

Figure 1 CT sagittal section showing cystic structure in the gall bladder.

Figure 3 Resected gall bladder.

CrossMark

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Figure 4 Cystic structure in gall bladder opened.

to further characterise this lesion and this was found to be a cyst with debris inside (figures 1 and 2).

TREATMENT

The patient underwent a laparotomy during which a right haemicolectomy was performed and an open cholecystectomy was done at the same time due to the above-mentioned cystic gall bladder lesion (figures 3 and 4). The postoperative course was uneventful, and the patient was discharged home in good condition.

OUTCOME AND FOLLOW-UP

Histology from the right haemicolectomy showed a perforated and inflamed diverticulum of the caecal region.

Sections from gall bladder showed mild chronic inflammation and several mucosal invagination (Rokitansky Aschoff sinuses). The macroscopically described cyst situated next to the gall bladder neck and cystic duct, measuring

Figure 5 At low power, normal gall bladder epithelium (top right) juxtaposed against the cyst lining (bottom left) (H&E $\times 10$).

Figure 6 A medium power view of the cyst wall showing a columnar epithelial lining, with very focal pseudostratification (H&E $\times 20$).

approximately 45 mm in greatest diameter, was composed of hypocellular, hyalinised fibrous tissue with few calcifications, lined predominantly by columnar ciliated epithelium (figures 5–7).

The epithelial lining of the cyst was cytokeratin (CK)7 positive and carcinoembryonic antigen, calretinin CK20 negative (figures 8 and 9). There was no overall evidence of dysplasia or malignancy.

There was no communication between the cyst and the gall bladder.

DISCUSSION

Ciliated foregut cysts are a rare entity and are usually found above the diaphragm, usually bronchial or oesophageal.¹ They are thought to be a remnant of the embryonic foregut.²

Figure 7 A high power view of cyst epithelium showing the columnar lining, pseudostratification and ciliary tufts (H&E $\times 40$).

Figure 8 Diffuse positive staining with CK7. Immunohistochemical CK7 stain $\times 40$. CK7, cytokeratin 7.

When they are found below the diaphragm, the distribution is mainly in the liver but data are limited as <100 cases have been described.³ When they are found in the liver, they are most commonly seen in the segments 4A and 4B, and more commonly found in men. There are also reports of such cysts found in the gall bladder.⁴ A review of the current literature found 12 such cases reported making this the 13th case.^{1 2 5–14}

The first described case in the literature was by Kakitsuba *et al*⁵. The term ‘ciliated foregut cyst of the gall bladder’ was first suggested by Nam *et al*¹. A ciliated foregut cyst may also arise in anatomically relevant areas as shown by a recent case in Al Betedini *et al*¹³ study reporting the first case of a ciliated foregut cyst in the triangle of Calot.

These cysts are usually unilocular and there is no communication with the gall bladder or bile ducts. The classic histological pattern comprises four distinct layers (inner ciliated epithelial lining, smooth muscle, loose connective tissue and fibrous capsule).¹⁵ Malignancy may also arise in these hepatic foregut cysts, mainly squamous cell carcinoma, unlike the rest of the gastrointestinal tract where adenocarcinoma is more prevalent.^{15–17} Malignant ciliated foregut cysts of the gall bladder have not yet been described.

Figure 9 Negative staining with CK20. Immunohistochemical CK20 stain $\times 4$. CK20, cytokeratin 20.

Epidemiology

Of the described cases so far, only two of the subjects have been male. These gall bladder ciliated foregut cysts are most commonly found in middle-aged females. Half of these cases were incidental findings, while the other half presented with pain, mainly in the right upper quadrant. Table 1 compares the characteristics of the ciliated foregut cysts of the gall bladder described in the literature.

Our case has the largest cyst described in the literature so far with 45 mm diameter. It is also unusual as it was found in an elderly man. Even though chronic cholecystitis was described in the histology, our patient was asymptomatic.

The incidental nature of the presentation of this foregut cyst makes it difficult to draw conclusions regarding treatment. This implies that finding such cysts incidentally on imaging or during surgery for other pathology may lead to uncertainties regarding treatment. So far none of the cysts described in the gall bladder have been found to be malignant, however malignancy has been described in ciliated hepatic foregut cysts (metaplasia and squamous carcinoma).¹⁸ Thus, excision should be considered in these cystic lesions due to the related nature of the pathology, especially if they anatomical position of the cyst is such that its excision would not cause harm to vital structures. A laparoscopic excision is an acceptable option in patients who do

Table 1 Comparison of ciliated foregut cysts of the gall bladder

Case	Year	Age/gender	Symptoms	US	Size (mm)	Location
Kakitsuba <i>et al</i> ⁵	1995	M/71	N/a	Anechoic	N/A	Corpus
Nam <i>et al</i> ¹	2000	F/36	No	Anechoic	15	Fundus
Hirono <i>et al</i> ⁷	2002	F/43	No	N/a	25	Collum
Muraoka <i>et al</i> ⁶	2003	F/37	No	Anechoic with echoic foci	24	Corpus
Bulut <i>et al</i> ⁹	2010	F/41	RUQ pain	N/A	35	Collum
Tuncyrek <i>et al</i> ¹⁰	2013	F/42	RUQ pain	Anechoic with echoic foci	7	Corpus
Giakoustidis <i>et al</i> ²	2014	F/29	Epigastric pain	Anechoic	30	Neck
Hwang <i>et al</i> ¹¹	2015	F/39	RUQ pain	N/A	35	Neck
Lee <i>et al</i> ¹²	2015	M/61	RUQ pain	Not seen	27	Corpus
Al Betedini <i>et al</i> ¹³	2016	F/33	RUQ pain	Not seen	15	Triangle of Calot
Cottreau <i>et al</i> ¹⁴	2016	57/M	RUQ pain, jaundice	N/A	17	Common hepatic duct
Present case	2016	M/72	No	N/A	45	Neck

F, female; M, male; N/A, not available; RUQ, right upper quadrant; US, ultrasound.

not present with other incidental pathologies requiring open surgery.

Learning points

- ▶ A ciliated foregut cyst of the gall bladder should be considered as a differential diagnosis of right upper quadrant pain.
- ▶ Malignancy cannot be excluded in such cysts and thus treatment is required.
- ▶ A laparoscopic excision should be considered as part of the treatment, especially if no other pathology is present that requires open surgery.

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Contributors AF did the original literature search, wrote up the article and contacted all contributors for their work. IB had the idea for the article and supervised the write up. PA supervised the write up. The patient was also under his care.

Competing interests None declared.

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