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Malignant transformation of a tailgut cyst

INTRODUCTION: *Tailgut cyst are congenital cystic lesion arising from remnant of the embryological postnatal gut. Tailgut cyst are multinodular, uncapsulated and usually well-circumscribed. Presacral cysts are rare in adult and most of the lesions are benign. Malignant degeneration can occur, however is extremely rare.*
CASE REPORT: *We present the case of a 74 years old woman with slow increase in size and malignant degeneration of a tailgut cyst. Five years before, during the follow up after mastectomy for cancer, she manifested rise of CA 19-9 tumor marker and a presacral cystic collection on thoraco-abdominal CT. She was followed with CT and MRI that showed that the cyst, with a solid component of the wall, was growing larger.*
After a five-year evolution, the cyst was resected. The histological examination on the solid component demonstrated intestinal adenocarcinoma.
CONCLUSION: *MRI ant TC can play essential role in the preoperative detection and characterization for the differential diagnosis, treatment strategies and evaluate neoplastic degeneration. Due to the risk of malignancy surgical resection must be performed after the diagnosis. Surgical therapy is mandatory when the cyst grow larger and a solid component is present.*

KEY WORDS: Presacral cyst, Retrorectal tumors, Tailgut cyst

Introduction

Tailgut cysts or retrorectal cystic hamartomas are rare congenital presacral lesions that are believed to arise from the remnants of the embryological postanal gut.
The retrorectal space is bounded by the rectum anteriorly, the sacrum posteriorly, the peritoneal reflection superiorly, the levator musculature inferiorly, and the ureters on each side.
The true incidence in unknown as the literature contains only small series and case reports, with a predominance of female middle-aged patients.

The clinical diagnosis is often delayed by absent or non-specific symptoms [1]. Malignant degeneration in tailgut cyst is extremely rare.
We report a case of adenocarcinoma within a retrorectal cystic hamartoma
A unique feature of our case compared with previously reported is the observation of the evolution towards malignancy

Case Report

A 74-year-old woman presented to our department with an asymptomatic long standing retrorectal cystic mass. Eight years before she had been submitted to left upper quadrant mastectomy, axillary lymphadenectomy and radio-chemotherapy for infiltrant ductal adenocarcinoma. Durig the oncologic follow-up, five years before our observation, a rise of CA19-9 tumor marker was seen. A thoracoabdominal CT examination was performed, which showed a presacral water-like cystic mass, attrib-

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uted to a localized collection (Fig. 1). A diagnostic workup with RM imaging was advised. Instead a PET examination was performed which didn't show any active metabolic lesion. After one year the patient was submitted to a control CT examination which didn't show any change in shape, volume and texture of the mass. A RM examination was newly strongly advised. Finally,two years before our observation, the patient was submitted to a low-abdominal and pelvic RM. In the presacral space, between sacrum and posterior rectal wall, there was a polilobulated oval mass, delimitated by thickened wall, with axial diameters of 80x90 mm and craniocaudal extension of 90 mm. The collection was hyperintense in T1 weighted sequences, referred to hematic content. On the lateral left side there was a nodular thickening, hipointense in all sequences, with well defined boundaries, of 20 mm in major axe. The CA19-9 rise persisted, with normal values of CEA.

One year before our observation a TC scan revealed the cyst was larger, with axial diameters of 77x 92 mm and a craniocaudal extension of 118 mm. On the left side there was a lobulation with solid intraluminal tissue, of axial dimension od 22 mm (Fig. 2). The mass comprimed and dislodged the rectum without any infiltration.

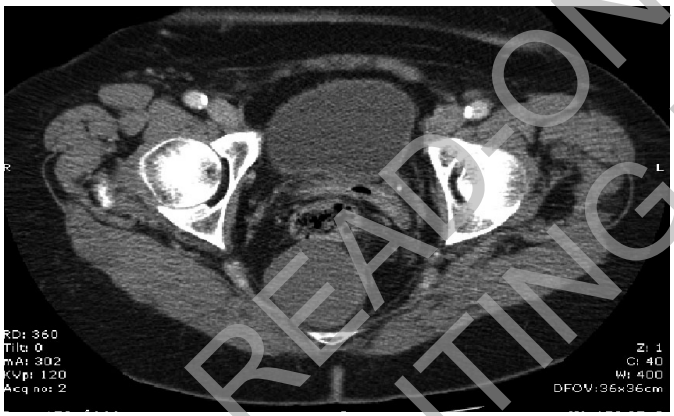

Fig. 1

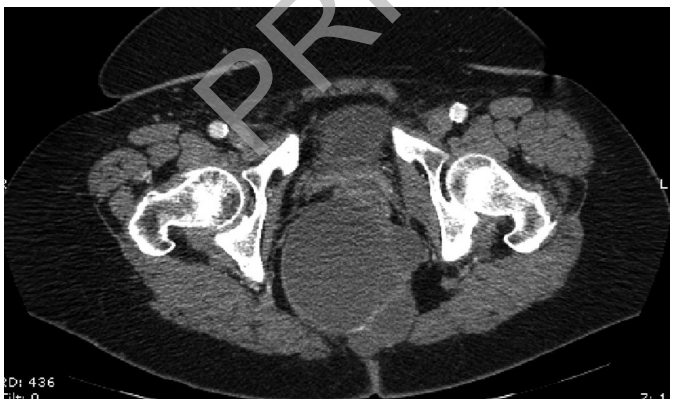

Fig. 2

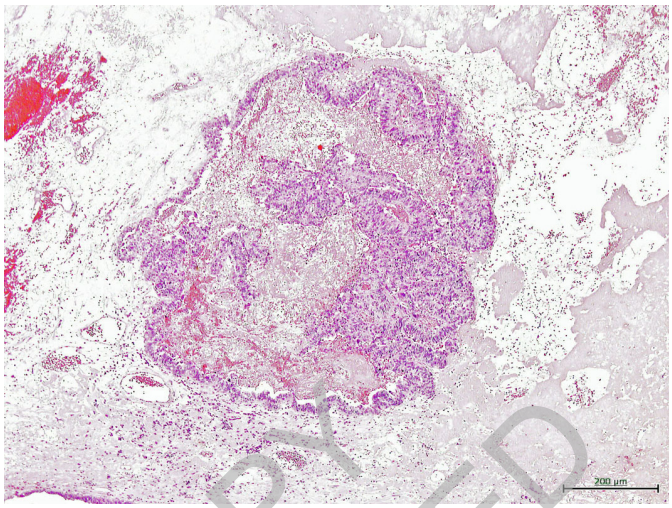

Fig. 3

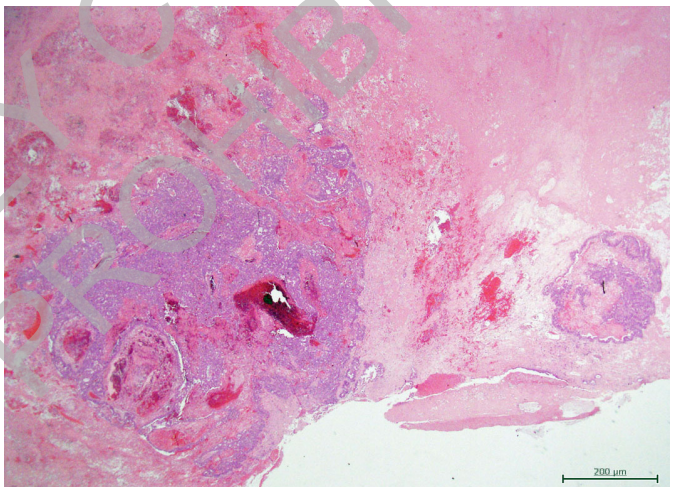

Fig. 4

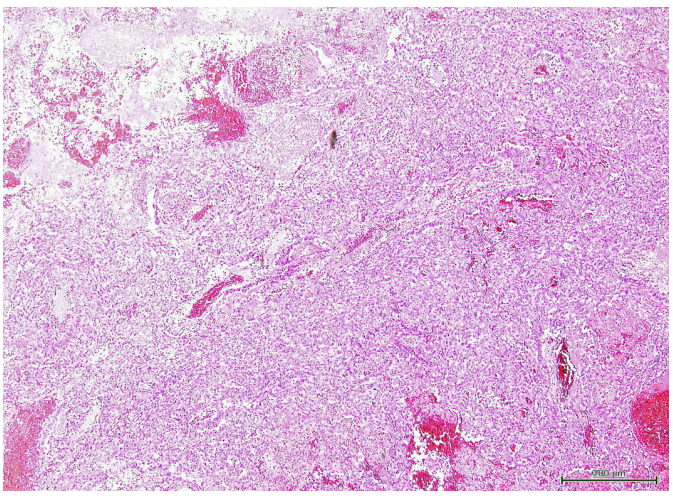

Fig. 5

After this a TC aspiration cytology was performed which didn't show any neoplastic cells. A surgical consult was advised which the patient refused.
After a 5 year story, with persistent CA 19-9 rise, the patient was taken in charge by our department. The patient was asymptomatic. On physical examination there was no abnormality. Digital rectal examination showed a non tender, extraluminal, well-defined presacral mass compressing the rectum. Sigmoidoscopy and colonoscopy revealed no mucosal abnormalities. Routine laboratory test results were within normal limits. Endorectal ultrasound revealed a polycystic pre-sacral mass, with hypoecogenic content , adherent but with a clear plane with the rectal wall.
The preoperative abdominal CT showed a presacral multiloculated cystic lesion, with hypodense content, axial diameters of 91x99 mm and craniocaudal extension of 132 mm. There was solid intraluminal nodular thickening of 25 mm.
At laparotomy the mass was strongly adherent to and not easily separated from the rectum and the sacrum. The cystic content (green, mucinous) was evacuated and the mass was excised. Inside a more solid area was palpable.
The patient's postoperative recovery was uncomplicated. The histologic examination of the thickening found a scarcely differentiated adenocarcinoma, with solid and focal micropapillary texture with necrotic and hemorrhagic areas (Figs. 3, 4, 5).
The diagnosis was of intestinal adenocarcinoma growed on amartomatous presacral cyst.

Discussion

The retrorectal (or presacral) space exist as a potential recess between the rectum and the sacrum; it is situated between the fascia propria that envelops the rectum and mesorectum and the presacral fascia. The presacral space ranges from the peritoneal reflection to the levators ani and coccygeous muscle, as its superior and inferior borders. The persistence in this space of the embryonic postanal gut remnant is very uncommon and results in the developement of a tailgut cyst.
In the literature various terms have been used to describe tailgut cyst: retrorectal cystic hamartomas, mucus-secreting cyst.
Most cases of tailgut cyst are discovered in adulthood and are more common described in middle-aged women. Nearly half the patients are asymptomatic [2], the remainder present with symptoms of rectal, buttock or perianal discomfort, particularly while sitting, as well as rectal bleeding, pain with defecation, lower back pain, constipation or obstructed defecation. Owing to the rarity of the condition and its nonspecific presentation, diagnosis is frequently late, during routine examinations, or in the course of investigation of other unrelated complaints. Delay in diagnosis may allow them to grow to a large size and this can further complicate the management [3]. They may become secondarily infected, and can present like pilonidal cysts, anorectal fistulas or abscesses [1]. Rectal examination usually reveals a well-circumscribed structure displacing the rectum posteriorly. Tailgut cyst should be distinguished from other cystic lesion which may occur in the presacral space: rectal duplication cyst, cystic teratoma and dermoid and epidermoid cyst.
Important differential diagnosis are excluded only pathologically, by considering the structure and epithelial lining of the cyst.
These lesions may have similar imaging features. Rough differentiation is possible by the fact that tailgut cysts are frequently multicystic or multilocular. CT and MRI have an essential role in the assessment of retrorectal masses. CT of the pelvis provides informations to guide surgical planning. MRI is the more useful to determine the internal composition and the characteristics of the wall [2,4]
Magnetic resonance imaging reveals a well-circumscribed usually multicystic lesion, that is hypointense on T1-weigted images and homogeneously hyperintense on T2-weighted images [5,6]. But the signal intensity of mucinous fluid is variable and depends on the protein concentration [6]. It adheres to the sacrum or rectum, but not communicate with the rectal lumen.
Malignant transformation is described in isolated case reports and includes adenocarcinoma, carcinoid and sarcoma. The risk of malignant transformation may be higher than previously thought [7].
The malignant degeneration within the cyst may be seen as irregular wall thickening with intermediate signal intensity on both T1 and T2-weighted MRI images [2,3,8].
On pelvic CT a malignant feature is the appearance of a solid component on the cystic wall [7]. Additional feature indication of malignancy include rapid increase in size [3]. Although diagnosis of malignancy has been reported with a needle biopsy, there is a potential for false-negative results [2] and also the biopsy carries the risk of spillage into the pelvic cavity and seeding of the biopsy tract. A biopsy is required only if it will alter management, and is therefore unnecessary if preoperative imaging provides information to allow appropriate surgical management [4,8].
Adenocarcinoma arising in a TGC has occasionally been associated with an elevated serum CEA and /or serum CA 19-9, however their elevation is not specific enough to permit a diagnosis of adenocarcinoma [2,9].
As the large majority of cystic lesions are benign, they may be managed with serial imaging, with surgical removal being reserved for those who develop radiological features suggestive of malignant transformation [10].
In our case a rapid increasing in size and a solid component in the cystic wall indicated surgical resection. The potential of malignant transformation means they should managed with caution. These lesions should be excised

early to reduce the risk of malignant change. Provided that the patient is fit for surgery presacral tailgut cysts should be resected when the lesion is identified [7,11].
Delaying surgery may increase the difficulty of removal.

Conclusion

Retrorectal cystic hamartoma is an uncommon entity that should be considered in the differential diagnosis of a retrorectal mass that have major cystic components.
MRI can play an essential role in the preoperative detection and characterization for the differential diagnosis and treatment strategies. Wall abnormalities, solid components or eterogeneity on MRI should raise the suspicion of malignancy.
Preoperative biopsy of cystic lesions is usually inaccurate. Malignant transformation, although unusual, may be focal, arising in a solid component of the cystic wall. Surgery is nearly always indicated.

Riassunto

Le tailgut cyst (amartomi cistici retrorettali) sono rare cisti congenite che originano nello spazio retrorettale o presacrale, generalmente benigne, multinodulari non capsulate, ben circoscritte e asintomatiche. La trasformazione maligna è un evento raro ma possibile. Presentiamo i casi di una donna di 74 anni, asintomatica. Cinque anni prima, durante il follow up oncologico per altra neoplasia, aveva manifestato aumento del CA 19-9 e quadro TC di raccolta presacrale. Le indagini eseguite in seguito (RM e TC) hanno dimostrato un progressivo e graduale aumento dimensionale della cisti retrorettale e la comparsa e l'incremento di una componente solida parietale. La paziente è stata sottoposta ad asportazione della cisti per via laparotomica. L'esame istologico ha mostrato la comparsa di aree di adenocarcinoma intestinale all'interno della componente solida della cisti. La possibilità della degenerazione maligna deve indurre ad una precoce asportazione chirurgica, a maggior ragione se è documentabile un incremento dimensionale o la comparsa di una componente solida.

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