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Huge retrorectal cystic teratoma mimicking meningitis. Case report and review of the literature

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CASE STUDY

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ABSTRACT

Retrorectal primary mature cystic teratomas are extremely rare and can be challenging to diagnose and treat in adults. These lesions are frequently clinically unrecognized and misdiagnosed. We present a case of a 39-year-old male patient with mature cystic teratoma presenting with repeated episodes of meningitis. He presented to our emergency department with complaints of fever, headache and vomiting since 15 days. He was treated for initial diagnosis as tuberculosis meningitis by a specialist of infection disease with antibiotics after a CSF study which was culture negative. Since he had recurrent meningitis, radiological investigations revealed by suggested it to be a retrorectal mature cyst 20cm in diameter which may be related with the cerebrospinal fluid. He underwent a total cyst excision of the lesion and histopathology confirmed a mature cystic teratoma. We identified about 25 cases of recurrent meningitis associated with dermoid and epidermoid cysts in the Medline literature search. A high index of suspicion is an important factor in making an early diagnosis; rectal examination and radiologic evaluation are also valuable.

Key Words
Cystic teratoma, diagnosis, meningitis, sacrum, treatment

Implications for Practice:

1. What is known about this subject?
Retrorectal developmental cyst (tailgut cyst, dermoid cyst, and teratoma) is very rare disease. A doctor would have such an experience at least once in their lifetimes.

2. What new information is offered in this case study?
Retrorectal cystic teratomas symptoms are not characteristic so that sometimes this disease is still misdiagnosed as a perianal problem and also meningeal irritation signs as we encountered.

3. What are the implications for research, policy, or practice?
All patients presenting meningeal irritation signs without any cause, they should undergo abdominal and pelvis examination including proctologic examination.

Background

Retrorectal cystic lesions are extremely rare in adults and in most cases are congenital.[1] The differential diagnosis includes a wide variety of conditions that occur in the retrorectal space. Because of their localization, clinical manifestation of cystic teratomas are delayed, despite their congenital nature.[2] The symptoms of teratomas vary between individuals, but these lesions are mainly identified as a result of organ occupation and compression of the surrounding organs or infectious complications of the cyst.

The most important complications of these cysts are infection with a secondary fistula and malignant degeneration.[2]

We present an extremely uncommon case of mature cystic teratoma in adult presenting with repeated episodes of meningitis in the retrorectal space.

Case details

A 39-year-old male patient who presented with repeated episodes of meningitis was referred to our hospital. He presented to our emergency department with complaints of fever, headache and vomiting since 15 days. He had no complaints of abdominal pain, or changes in habitual normal bowel movement (diarrhoea or constipation) except abdominopelvic discomfort and temnesmus. Urination was normal. He explained that all sexual functions and physical activities were normal before this problem. Neurological examination showed that there was neck stiffness with positive Kernig and Brudzinski signs. On further enquiry, he described similar episodes twice in past. Cerebrospinal fluid (CSF) analysis showed lymphocytosis and elevated proteins. No organism could be identified in culture. He was treated for initial diagnosis as tuberculosis meningitis by a specialist of infection disease with antibiotics. Since he had recurrent meningitis, radiological investigations revealed by suggested it to be a retrorectal mature cyst 20cm in diameter which may be related with the cerebrospinal fluid. Pelvic MRI revealed that fat saturated T2-weighted (A) and T1-weighted (B) axial images show the relation between retrorectal cyst and spinal canal. It can also be seen “lipoma of the filum terminale” in spinal canal (Figure 1). It is known that a relatively common finding on imaging of the lumbar spine, and in most cases is an incidental finding of tethered cord syndrome. This syndrome commonly associated with sacral meningocele, So It needs to be considered for differential diagnosis. Anorectal malformation and sacral bony deformity resembling Currarino syndrome were not detected. Routine laboratory tests and serum tumour markers including CEA and CA19-9 were all within normal ranges. He underwent transsacral approach and laparotomy for total excision of the cystic tumour.

He was firstly operated by transsacral approach than through a lower midline abdominal incision. At the first step in a jackknife position, the connection between spinal canal and presacral (retrorectal) cystic lesion was carefully dissected and cut short by the brain surgery team. The posterior dural defect was closed by direct suturing. On laparotomy, cystic lesion in the abdominal pelvic space is formed posteriorly by the sacrum and coccyx and anteriorly by the rectum. The lesion displaced the rectum anteriorly. The pelvic peritoneal reflection forms the superior border, and the levator ani and coccygeus muscles form the inferior border. Retrorectal space was opened laterally, large silvery white epidermoid tumour was found completely occupying the retrorectal space. The cystic mass had a diameter of 20 cm and its outer border was smooth with no vessel or ligamentous connection to the surrounding organs, including the rectum, prostate and bladder. There was no invasion to the spinal cord, sacral plexus, nerve roots or adjacent tissues. In which it was gradually dissected from the adjacent tissues. The cyst was adherent to the sacral bone and was found to be entering the sacrum posteriorly from a defect in the sacrum which was removed gradually. The cystic lesion was found to contain thick greenish granulated yellow fluid.

Macroscopically, the cystic tumour was a multilocular cystic lesion containing keratin, sebaceous matter, hair, and follicles. Histopathology confirmed as a mature cystic teratoma (retrorectal cystic teratoma) (Figure 2). Postoperative period was uneventful and patient was fine with no added neurological deficit.

Discussion

Retrorectal cystic lesions are the most common solid neoplasm in neonates, with an estimated prevalence of one in 35,000–40,000 births.[1] It is estimated that about 50 per cent of these tumours are completely asymptomatic, being discovered accidentally during a rectal/ vaginal examination or abdominal-pelvic ultrasound made for another reason.[3,4] Size of a cystic teratoma is average 8cm (range 1 to 30cm). Our case is 20cm in diameter and is one of the biggest cyst in the literature.[3–5]

Biological behaviour of mature cystic teratomas does not predict. Hence, it is important to consider the disease as a differential diagnosis.[6,7] Our patient was admitted to hospital with meningeal irritation symptoms. He did not claim specific symptoms secondary of mass effect caused by the lesion volume such as constipation, rectal fullness, dysuria and pelvic pain. Actually, we expected these symptoms and interrogated, but there was not. The chronicity of disease may be the reason of this condition. In our case, the lesion discovered incidentally during magnetic resonance imaging for differential diagnosis of meningitis. We hypothesized that there could be a communication between cystic fluid and cerebrospinal fluid, so the cystic content passed through to the cerebrospinal fluid lead to chemical meningitis. At first admission, its acute clinical course and negative laboratory findings are

indistinguishable from those of bacterial meningitis. So, our colleagues have had to start standard antibiotic and steroid treatment for meningitis. But, they have to do advanced diagnostic studies because of the second presentation of the patient with similar symptoms. Magnetic resonance imaging studies (Figures 1 and 2) and intraoperative findings supported our hypothesis. Finally, our patient is still uneventful for 9 months straight after operation. To the best of our knowledge, there are about 25 case presentations similar with our case in the Medline literature.[8–10]

It is generally suggested that the patients with retrorectal mature cystic teratomas are suitable for surgical treatment because of a malignant potential.[2] The surgical approach selected should be based on various factors including tumour size, location, and invasion to the surrounding organs, and appropriate selection might be the key to successful operation. Zhou and Qui suggested that the treatment of choice for mature cystic teratoma is early surgical resection with complete excision of the coccyx, because microscopic nests of neoplastic cells are commonly found in or immediately adjacent to the coccyx.[4] Therefore, various surgical approaches have been reported, including trans-abdominal, trans-sacral, and a combined abdomino-sacral approach. In our case, an abdomino-sacral approach was adopted because of the suspicion that the cyst had related with the cerebrospinal fluid.

Conclusion

In conclusion, we described an extremely rare case of a patient with a retrorectal mature cystic teratoma mimicking meningitis. Special attention should be paid for the clarification on the differential diagnosis and therapeutic problems of cystic teratomas. A high index of suspicion is an important factor in making an early diagnosis; rectal examination and radiologic evaluation are also valuable. The preferred surgical approach should be early surgical resection with complete excision of the coccyx, because microscopic nests of neoplastic cells are commonly found in or immediately adjacent to the coccyx.

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PEER REVIEW

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CONFLICTS OF INTEREST

The authors declare that they have no competing interests.

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PATIENT CONSENT

The authors, *Dilek ON, Halıcı AH, Acar T, Gür EO, Özşay O, Çakalağaoğlu F, Altay S, Hacıyanlı M*, declare that:

1. They have obtained written, informed consent for the publication of the details relating to the patient(s) in this report.
2. All possible steps have been taken to safeguard the identity of the patient(s).

3. This submission is compliant with the requirements of local research ethics committees.

Figure 1: Pelvic MRI; Fat saturated T2-weighted (A) and T1-weighted (B) axial images show that there is relation between retrorectal cyst and spinal canal. It can also be seen "lipoma of the filum terminale" in spinal canal (arrows). Note that cystic lesion displaced the rectum anteriorly

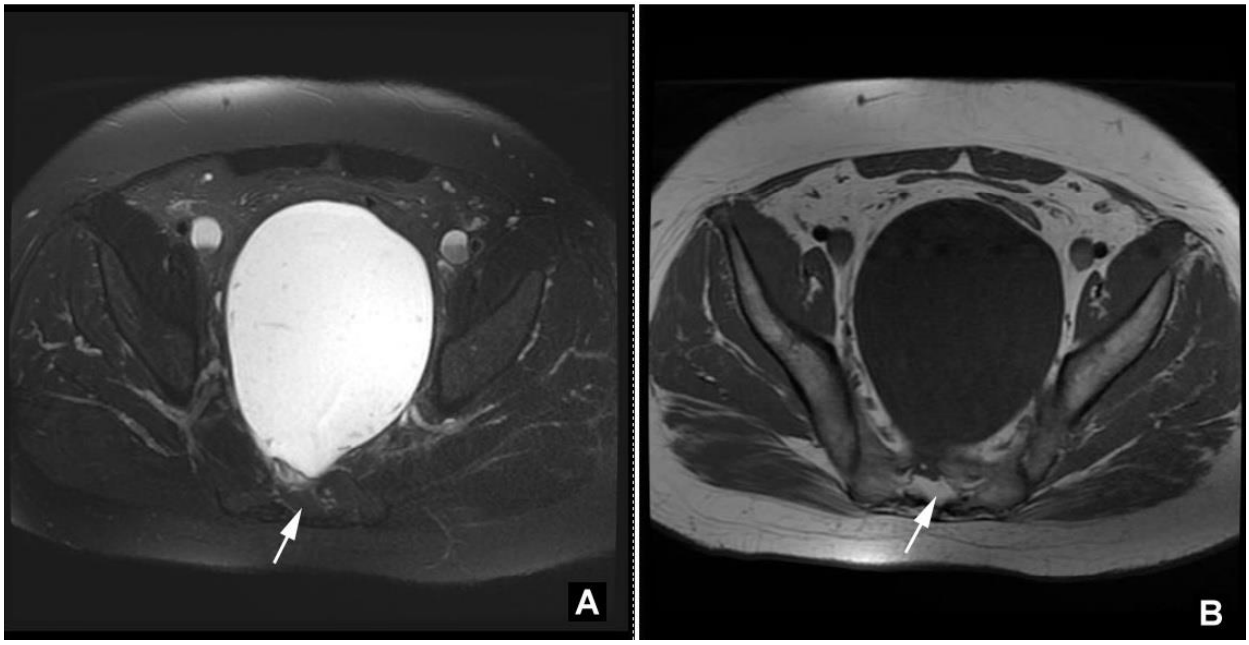

Figure 2: Photomicrograph shows stratified squamous epithelium lining the cyst containing necrotic debris (Original magnification, ×10; HE stain)