Presacral epidermoid cyst.
A case report

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Francesco Negro, Massimo Mercuri, Vincenzo Ricciardi, Massimo Massari, Carmelo Destito, Samuele Mafucci, Tiziana Cozza

Catholic University of the Sacred Heart, Rome; Department of Surgical Sciences (Director: Prof. A. Picciocchi); Institute of Clinical Surgery (Director: Prof. M. Castagneto); Complex Operative Unit, 1st General Surgery (Director: Prof. A. Wiel Marin).

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INTRODUCTION: *Most of the primitive cystic neoplasms arising in the retrorectal presacral space are congenital. Presacral epidermoid cysts are classified as a type of developmental cysts, resulting from an error during the embryogenesis. They have a slowly-progressive growth that only lately can cause clinically remarkable symptoms.*

Most patients are middle-aged women. They are often asymptomatic. The discovery is almost always casual. It happens during controls like US, CT, MRI, gynecological visit. The surgical treatment must be effected for a correct diagnosis of nature of the lesion and to remove the mass. In fact, infective complications may occur and the increasing volume of the cyst can give clinical symptoms. Posterior approach is indicated for low or mid presacral space tumors; it is the most commonly followed route. The abdominal approach may be adequate for large developmental cysts.

CASE REPORT*: A case of an incidentally found large presacral epidermoid cyst in a young woman, which was excised through an abdominal approach is reported.*

KEY WORDS: Developmental cysts, Presacral epidermoid cysts, Retrorectal tumors.

Introduction

Many types of tumors can develop in the presacral space, since it is composed of different kind of embryonic tissues. Neoplasms can be solid or cystic (40.8% of cases) [1] and are usually classified as either congenital or acquired (inflammatory, neurogenic, osseous, or miscellaneous) according to the tumor's tissue of origin. Congenital lesions represent more than 50 per cent of tumors. Developmental cysts, which result from an error during the embryogenesis, constitute the majority of cases [2]. Presacral epidermoid cysts are such type of lesions. Different surgical approaches are described for treating presacral tumors. A case of a large presacral epidermoid cyst, excised through an exclusive abdominal approach, is reported.

Case report

A 49-year-old woman was admitted to our institution for the incidental finding of a cystic mass in the presacral space, discovered during an ultrasound examination performed for gynecologic issues. In fact, she was suffering from uterine fibromatosis.

Past medical history did not evidence any notable diseases unless an uterine fibromatosis.

On digital examination, a painless elastic soft mass was palpable on the posterior wall of the rectum.

Ultrasound evaluation through the suprapubic and transrectal way demonstrated a cystic retrorectal mass, 85 mm in diameter, with decreased echogenicity and slight posterior enhancement.

Computed tomography (CT) scanning showed a large cystic mass with a thin wall and fluid density (Fig. 1). On Magnetic Resonance Imaging (MRI) the mass showed a heterogeneous low signal intensity on the T1-weighted image and a high signal intensity on the T2-weighted image; keratinous material appeared as multiple small hypointense foci (Fig. 2).

An abdominal surgical approach was preferred on account of the dimension of the cyst. It permitted to smoothly excise the large mass that dislocated the rectum anteriorly and was quite fixed to the coccyx.

The gross specimen showed a thin-walled cyst, filled with a large amount of greenish dense fluid. On microscopy there was evidence of a fibrous capsule, lined by stratified squamous epithelium containing keratinous material. The patient had an uneventful recovery.

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For correspondence: Francesco Negro, MD, Via della Pineta Sacchetti 175, 00168 Rome, Italy.

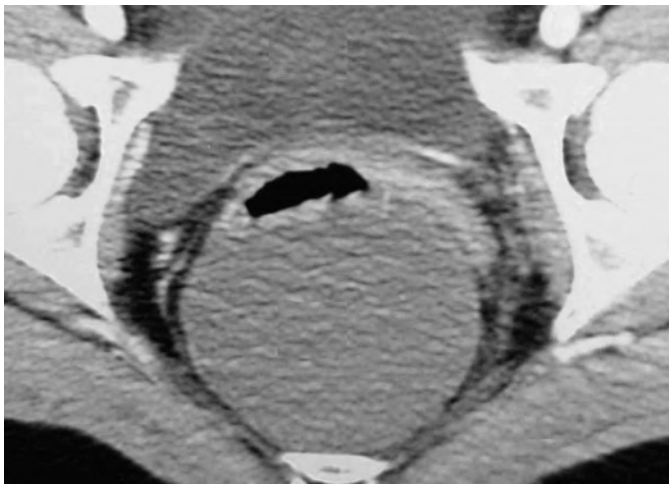

Fig. 1: Computed tomography scanning. An 85 mm cystic retrorectal mass is evidenced with a thin wall and fluid density.

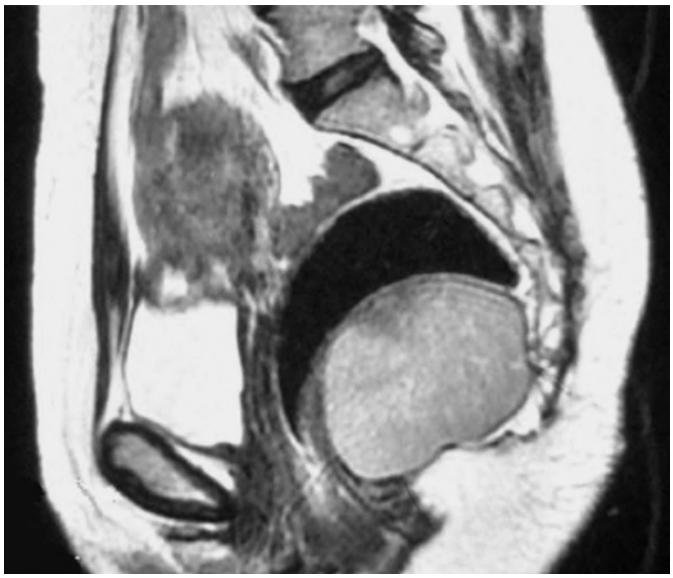

Fig. 2: Magnetic Resonance Imaging (sagittal section). The cystic mass and its relation to the surrounding organs are well defined.

Discussion

Tumors arising in the presacral space are rare: their incidence has been estimated as 1 in 40, 000 admissions [3]. Congenital lesions represent the most frequent type [1,2,4]. According to the different embryonic tissue of origin, they can be classified as developmental cysts, chordomas, anterior sacral meningoceles, adrenal rest tumors, duplications of rectum.

Developmental cysts are considered as the result of an error during the embryogenesis [5]. They include epidermoid cysts, dermoid cysts, cystic hamartomas (mucus-secreting cysts), teratomas, and teratocarcinomas. Epidermoid cysts contain squamous epithelium without skin appendages.

Acquired presacral tumors comprise inflammatory lesions (perirectal abscesses, complicated diverticulitis or Crohn's disease, foreign body granulomas), neurologic tumors (neurilemomas, neurofibromas, neurofibrosarcomas, ependymomas, and ganglioneuromas), osseous lesions (osteomas, osteosarcomas, Ewing's sarcomas, simple bone cysts, and aneurismal bone cysts), miscellaneous tumors (metastatic carcinomas, soft tissue sarcomas, carcinoid tumors).

Presacral cysts are prevalent in women. Percentage varied among 77, 86, and 94% in three large series [1,6,7]. Despite the congenital nature, their manifestation is delayed up to the middle age. Most patients are asymptomatic [6,8-10], because of the localization of the cysts and their slowly-progressive growth. When present, pain is usually described as a vague rectal discomfort and may indicate an infective complication (infected cyst or fistulization) or malignancy [6,11].

The diagnosis can be made by digital rectal examination [1]: this remains the most effective way to identify the mass and to get information for the operative approach.

On proctosigmoidoscopy rectum presents a normal mucosa overlaying a smooth protrusion.

Ultrasound evaluation through the suprapubic and transrectal way demonstrates the cyst as a retrorectal mass with decreased echogenicity and slight posterior enhancement [12].

The CT findings show a cystic mass with a thin wall and fluid density [12-14]. Usually there is a homogeneous low attenuation but keratinous material can show a heterogeneous hypodensity.

MR images may contribute to better define the spatial relation of the mass to the surrounding organs and differentiate developmental cysts from other cystic tumors as tailgut cysts [15, 16]. On the T1-weighted MR image the mass show a heterogeneous low signal intensity [12-14]. On the T2-weighted MR image there is a high signal intensity; keratinous material can be seen as multiple small hypointense foci [12].

In selected cases, barium enema, arteriography, or myelography may be requested.

It is not recommended to perform a preoperative biopsy because of the risk of infecting sterile cysts and seeding malignant cells [17].

Complete excision of presacral tumors is considered necessary for the possibility of malignancy or infection [2,18,19].

Four surgical approaches are described: abdominal, posterior (with or without sacral resection), combined abdominosacral, and transrectal. Abdominal approach is preferred for high presacral space lesions; it is used for benign cystic tumors [14]. Posterior approach is indicated for low or mid presacral space tumors; it is the most commonly followed route (in more than 80% of cases in large series) [1,2,4]. The combined abdominosacral approach is recommended for larger masses or malignant neoplasms infiltrating the sacrum [11, 20, 21]. The transrectal

approach is suggested for small cystic lesions in the low presacral space, without fixity to the sacrum [6,7,10].
Our experience confirms that the only abdominal approach may provide an adequate surgical treatment of large epidermoid cysts when the possibility of malignancy has been ruled out.

Riassunto

Molte delle neoplasie primitive che sorgono nello spazio retrorettale presacrale sono congenite.
Le cisti epedermidoidi presacrali sono un tipo di cisti crescexti risultanti da un errore embriogenetico. Esse hanno una lenta e progressiva crescita che può causare sintomi rilevabili clinicamente solo in ritardo. Molti pazienti sono donne di età media che spesso sono asintomatiche.
La scoperta delle cisti è quasi sempre casuale e avviene durante controlli quali la ultrasuoni TC, ecografia, RM o visita ginecologica.
Il trattamento chirurgico deve essere effettuato dopo una diagnosi corretta sulla natura della lesione e si deve rimuovere la massa. Sono infatti possibili complicazioni infettive e il crescente volume della cisti può dare sintomi clinici.
Per i tumori del basso e medio spazio presacrale è indicato l'approccio posteriore: e questa è la via comunemente usata. L'approccio addominale è preferito per le lesioni presacrali alte e di grandi dimensioni.
CASO CLINICO: Gli Autori descrivono un caso di una voluminosa cisti epidermoide presacrale, riscontrata occasionalmente, che è stata escissa attraverso un approccio addominale.

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