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Case Report

A Case of Presacral Teratoid Cyst in an Adult Patient

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Abstract

Presacral teratoid cyst (PTC) is a congenital structural abnormality located in the pararectal area and containing tissues from different germ layers. Cases of diagnosis and treatment of PTC in adults are extremely rare and there is little information about the treatment tactics and prognosis. We describe a case of PTC in a 28-year-old woman, with a description of the diagnostic process, preoperative and postoperative management, as well as a brief review of the literature on this topic. A reconstructive surgery was performed on the perineum, including: parasacral access, excision of the presacral cyst together with removal of the coccyx and plastic of the opening. The postoperative observation of the patient was without complications.

Keywords

presacral teratoid cyst, parasacral access, presacral area

INTRODUCTION

The presacral region is a complex area, limited in front by the posterior wall of the rectum, posterior by the anterior surface of the sacrum and inferior by the retro-sacral fascia (Waldeyer). This space is limited laterally by the intrapelvic fascia (lateral ligament), the ureter and the iliac vessels.

Presacral teratoid cyst (PTC) - is a congenital structural abnormality located in the pararectal tissue and contains tissue from different germ layers (ectoderm, mesoderm, and endoderm). Most often, it is diagnosed in childhood, with incidence of 1 in 35,000-40,000 and 4-10:1 ratio of women to men.[1,2] However, the diagnosis of these abnormalities and treating them in adults is an extremely rare occurrence. Bull and colleagues in 1999 conducted an analysis of literature data on the subject and indicated that only 85 cases of PTC in adults were reported.[3] The experience of large medical centres over a 40 year period often includes about 70 cases of PTC.[4] Such a rare occurrence is due to the fact that in 50-70% of cases, PTCs are detected in the first few days of life and in less than 10% after 2 years.[4] As a rule, PTC is benign and can become malignant only in 1-2% of cases.[2] Diagnosis of the disease is mainly due to imagistics. The main method of treatment is the complete removal of PTC together with the coccyx.[5] At the same time, despite successful surgical treatment and a good prognosis, the chance of relapse is about 10-37%.[6,7] In this article, we present a case of examination, treatment and postoperative observation in the dynamics of a patient with PTC.

CASE REPORT

A 28-year-old patient was admitted to the hospital with complaints of constipation and pain in the coccyx area. Symptoms appeared two months before hospitalization. The laboratory analyses were normal, and the tumor markers CA 125, CA 19-9, CEA were negative. Colonoscopy revea-

led a polyp in the rectum and gastroesophagoduodenoscopy revealed cardia insufficiency, chronic gastroduodenitis, a forming polyp in the stomach, and duodeno-gastric reflux. Ultrasound of the abdominal and pelvis organs revealed an 11.5 × 10.2-cm anechoic round shape formation with clear even contours (below the uterus and ovaries). An MRI scan was performed to clarify the diagnosis ***(Figs 1a, 1b)****. In the MRI, a round formation was found in the pelvis with a diameter of 11 cm which corresponded to PTC in terms of localization and structure.*

According to the chosen treatment tactics, we performed reconstructive surgery on the perineum, including: parasacral access, excision of the presacral cyst together with removal of the coccyx and plastic of the fistulous opening with a full-layer rectal wall flap.

The patient was operated under combined endotracheal anesthesia. A skin incision was made in the area of the intergluteal fold up to 15 cm in length. The coccyx is the region was removed. The incision was then continued parasacral. The tendon of the left gluteal muscle in the area of attachment to the left lateral surface of the sacrum was dissected. The sacral space was opened, and an excision of the presacral teratoma was performed accompanied by the reconstruction of the pelvic floor ***(Fig. 2)****. Drainages were established in the area of the*

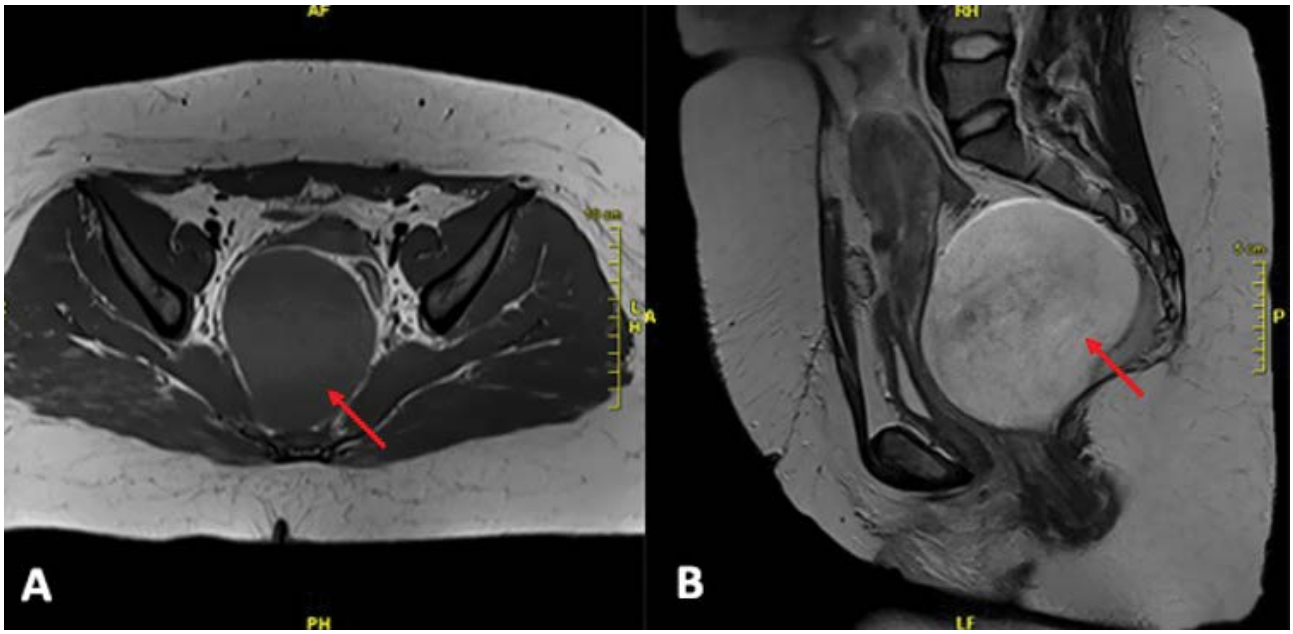

Figure 1. A. MRI in the axial plane, voluminous structure in the pelvis. **B.** MRI in the sagittal plane, voluminous structure in the pelvis (indicated by an arrow).

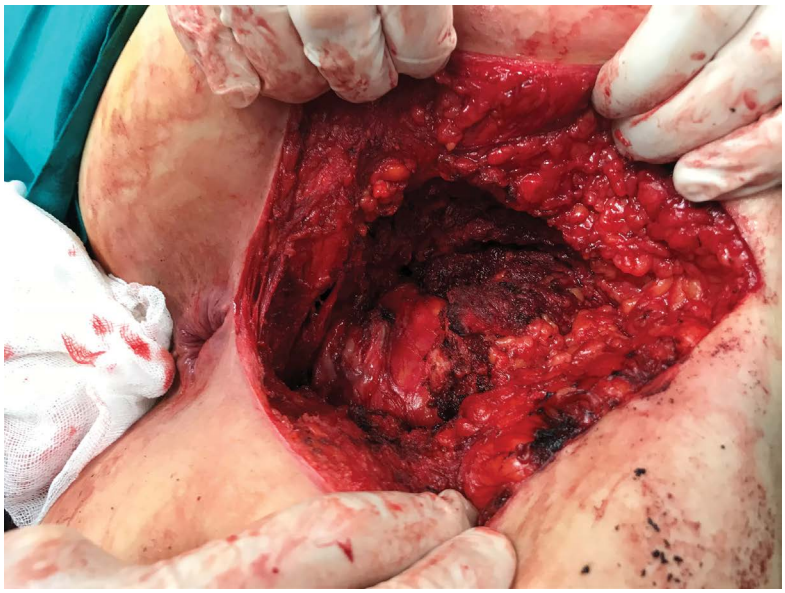

Figure 2. Intraoperative image after the removal of PTC.

removed presacral formation, inferred through contraperture in the left gluteal region. The operation ended with layered suturing and aseptic dressings.

At macroscopic appearance, the teratoma was a soft rounded structure with a diameter of 11 cm ***(Fig. 3)****. On a section, it was soft uniform and pale yellow. At microscopic appearance, it was an encapsulated ganglioneuroma with petrificates (mature monomorphic spindle-shaped Schwann cells in the collagen stroma, interspersed with ganglion cells of various sizes, some of them contained lipofuscin).*

The postoperative period was uneventful. The patient was discharged 7 days after hospitalization. Postoperative MRI data after 1 month are presented in ***Figs 4A, 4B****.*

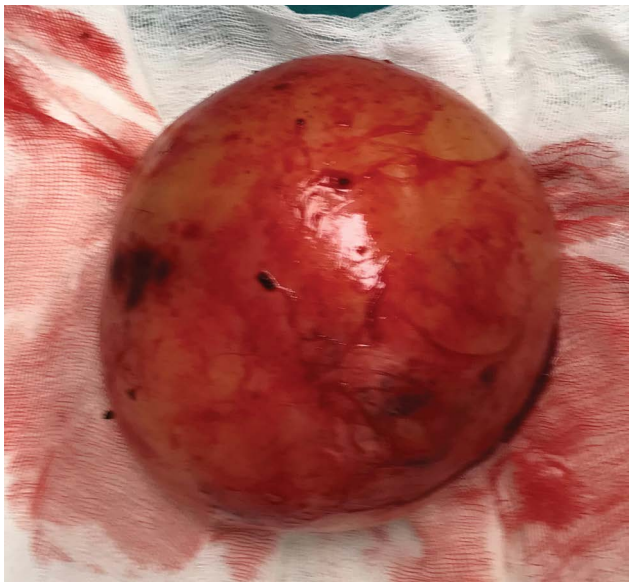

Figure 3. Macroscopic PTC specimen.

DISCUSSIONS

Teratomas are benign tumors consisting of tissues that do not correspond to their anatomical location. They are formed in the embryonic period from pluripotent cells and have various degrees of maturity.[8]

They also have the potential for malignancy. Thus, at the age of 2 years, approximately 7-10% of the PTCs can be malignant, while after 2 years the percentage of malignant cases increases to 50-67%.[6] The potential for malignancy increases with age, which should be taken into account when planning surgical intervention and postoperative management of the patient.

Patients with PTC may be asymptomatic. In case of symptoms, most often patients complain of pain in the lower back and/or pelvis, constipation, urinary retention, paresthesia and weakness of the lower extremities.[9,10] Other symptoms include menstrual irregularities, nausea, vomiting, and swelling of the limbs.[11] Most often, the symptoms are due to the large size of the PTC and its pressure on nearby organs (rectum, bladder, uterus). Extremely rarely, symptoms are due to malignancy and invasion of the tumor into adjacent anatomical structures.[11]

For the diagnosis of PTC, the most effective and significant methods are CT and MRI. Both methods have their advantages and disadvantages. Thus, CT allows a better assessment of the degree of calcification of the formation and the integrity of nearby bone structures. MRI allows the clinicians to determine more accurately the topographic relationship of the tumor to nearby structures, and provides better detail of the anatomy of the area.[11] Assessment of the degree of calcification is an important marker. Uneven calcification occurs in 75% of benign PTCs and in about 12.5% of malignant ones.[12,13] If malignancy is suspected,

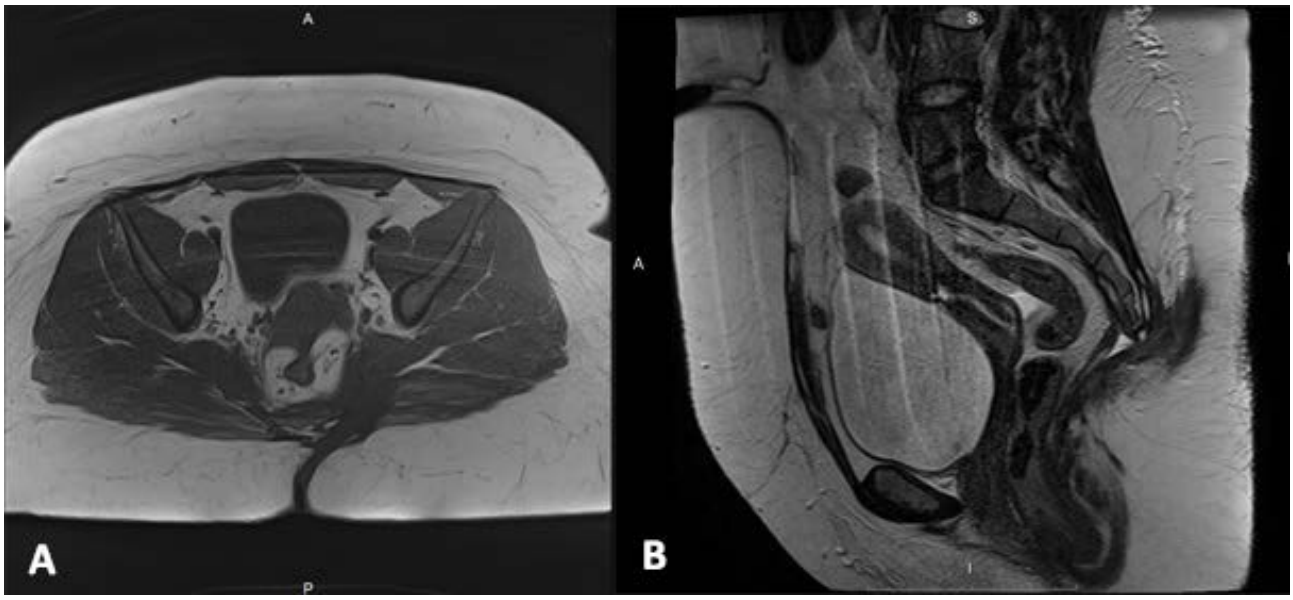

Figure 4. A. Postoperative MRI of the pelvis in the axial plane. **B.** Postoperative MRI of the pelvis in the sagittal plane. The cavity after removed PTC.

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an assessment of blood levels of AFP, βhCG, CEA, Ca-125, CA19-9, and CA 15-3 is useful.[14]

Altman and co-authors proposed a classification of PTC, thus dividing PTC into 4 types. Type I - mainly tumors with an external component and minimal coccyx involvement. Type II - external tumors, but with a significant spread within the pelvis. Type III - these also have an external component, but mainly pelvic localization and intraperitoneal distribution. Type IV - presacral location without external component.[6]

Depending on the localization and prevalence of the process, abdominal, sacral, combined and laparoscopic approaches to the tumor can be applied.[15] If the PTC is distributed within the abdominal cavity, abdominal access is usually preferred. Abdominal access is also indicated when the tumor is not lower than S4 and the nerve structures are not affected. Ligation of *a. sacralis media* may be necessary to control intraoperative bleeding. If it is impossible to ligate the artery, alternatively, an aortic clamp can be applied over its bifurcation. These techniques require laparotomy. The advantage of laparoscopic access is that it is minimally invasive and has reduced number of postoperative complications. For large tumors that are located above and below S4, combined access is often used. For structures that were preoperatively regarded as benign and were below S4, with the involvement of the coccyx, parasacral access is indicated. Removal of the coccyx is a necessary procedure to ensure adequate access, as well as to prevent relapse of teratomas.[16]

Differential diagnosis is carried out with tumors, cysts, developmental abnormalities and abscesses, taking into account the clinical picture, age, gender and imagistics. The final diagnosis in this case is always made on the basis of histological examination. Another important syndrome worth keeping in mind is Currarino syndrome (anomaly), which is an autosomal dominant genetic disease of the HLXB9 gene, located at the 7q36 locus. The three most characteristic clinical signs of this syndrome are presacral tumors, an underdeveloped coccyx (agenesis), and anomalies in the development of the anus and rectum.[17]

In the present case, the patient was diagnosed with PTC (type IV according to Altman classification), which was removed using parasacral access, with good clinical and cosmetic results and without subsequent complications.

CONCLUSIONS

Cases of PTC in adults are rarely found in the literature. Treatment of patients with this pathology can be associated with serious difficulties, depending on the involvement of anatomical structures in the pathological process. The latter may require participation of surgeons of different specializations. In the discussed case, a 28-year-old patient with type 4 PTC underwent surgery with presacral access with removal of the coccyx, teratoma with a good cosmetic result. The management of this case may require the involvement of several specialists such as colorectal surgeons, vertebrologists, and neurosurgeons.

Ethics

Written informed consent was obtained from the patient for publication of the present study.

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Случай пресакральной тератоидной кисты у взрослого пациента

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Резюме

Пресакральная тератоидная киста (ПТК) – это врождённая структурная аномалия, расположенная в парасакральной области и её тканях из различных зародышевых листков. Случаи диагностики и лечения ПТК среди взрослых крайне редки, мало информации о подходах к лечению и прогнозах. Мы описываем случай ПТК у 28-летней женщины с описанием диагностического процесса, предоперационного и послеоперационного ведения, а также краткий обзор литературы по этому вопросу. Выполнена реконструктивная операция промежности, включающая: парасакральный доступ, иссечение пресакральной кисты с удалением копчика и пластикой отверстия. Послеоперационное наблюдение за пациентом прошло без осложнений.

Ключевые слова

пресакральная тератоидная киста, парасакральный доступ, пресакральная область