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Sacrococcygeal teratoma – case report and review of the literature

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Summary

Background: Primary presacral tumour is extremely rare among adults. Such lesions are often found incidentally, because they may not show any clinical symptoms. Moreover, differentiation between cystic changes in ovary and presacral region is extremely difficult; it might even be a diagnostic challenge. Imaging studies should include transvaginal and transrectal ultrasound, computed tomography and magnetic resonance imaging, which are useful in identifying the exact location and suggest character of the tumour. Such diagnostics are helpful in determining the optimal surgical procedure.

Case Report: We present a case of a 26-year-old patient with abdominal pain. Imaging studies were performed; they revealed a presacral cyst 5 cm in diameter. Laparoscopic removal of the cyst was performed. Histopathological examination of the lesion confirmed a mature teratoma. The postoperative period was uneventful.

Conclusions: Due to the rarity of reported changes no firm guidelines exist. In the literature, depending on the anatomical conditions, descriptions of various surgical procedures are reported. The presented case confirms that surgical treatment can be performed by laparoscopy.

key words: **sacrococcygeal teratoma • presacral teratoma • gynaecological laparoscopy**

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Background

Most teratomas occur in ovaries and testes of adolescents and originate from germ-cells. Teratomas can also be found in midline structures: sacrococcygeal, presacral, retroperitoneum, postanal or even anterior mediastinum and pineal gland [1]. Teratomas of non-gonadal location originate from embryonic cells. Various theories try to explain the origin of teratomas. The pathogenesis is generally thought to be a growth of primitive totipotential cells that have differentiated and matured. According to the germ cell theory the totipotential cell is a "wandering germ cell" left behind during migration of embryonic germ cells from yolk sac to the gonad. According to the most popular theory the source of totipotential cells is a residual cell from the primitive streak and Hansen node of very early embryonic development. Normally, this primitive structure degenerates, however remains may persist in the sacrococcygeal region and develop into teratomas. There is also the theory that teratomas might be the result of twinning attempts [2,3].

Familial form of presacral teratomas, inherited in an autosomal dominant pattern, has been reported. They differ from usual teratomas because of nearly equal sex prevalence, a low prevalence of malignancy and a sacral defect [3].

Primary presacral teratomas are extremely rare in adults. Teratomas of such localization appear in neonates and young adults. Sacrococcygeal teratomas are most commonly found in neonates, infants and children younger than four years. They appear with incidence of approximately 1:35,000–40,000 live births and a female to male ratio 4:1 [4]. Most changes can be diagnosed prenatally, 50–70% are found during first few days of life, less than 10% are diagnosed beyond the age of two years [2]. Malignant transformation has been found in approximately 1% of teratoma patients comprising squamous cell carcinoma, adenocarcinoma, sarcoma and other malignancies [4]. In adults presacral teratomas occur at a rate between 1 in 40,000 and 63,000 with female preponderance 3:1 [5]. Multiple masses of tumour are extremely unusual, only two cases have been reported [2].

Tumours of sacrococcygeal location originate ventral or dorsal to the sacrum, they may grow posteroinferiorly into the gluteal area or anterosuperiorly into the lesser pelvis. Especially in the ventral location these tumours may grow to a large size as they develop into the retrorectal or presacral space [5].

Altman et al. classifies sacrococcygeal teratomas. They marked out 4 categories by location. Type I are predominantly external tumours with minimal presacral component. Type II presents externally but with significant intrapelvic extension. Type III is still apparent externally but predominantly a pelvic mass extends into the abdomen. Type IV is a presacral mass with no external presentation. Our case conform type IV of Altman's classification, although most common in adults is type III [6].

Case Report

A 26-year-old woman, nulliparous, reported with abdominal pain occurring on the right side for about 11 months. In August 2010 on the right side behind the uterus an ovarian cyst 32 mm in diameter was found. Pharmacological treatment

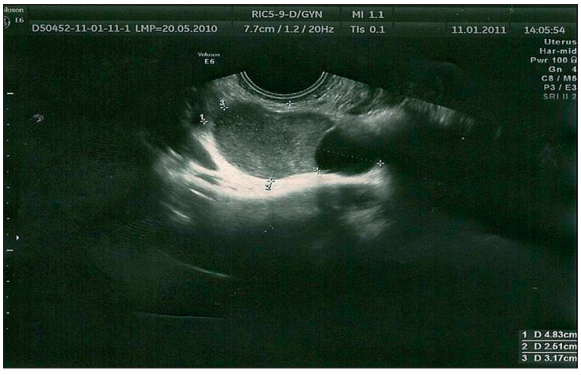

Figure 1. Transvaginal USG. Dual-chamber fluid space – a cyst – 48×25 mm Thick chamber of the lesion diameter 32 mm, fluid chamber 17 mm.

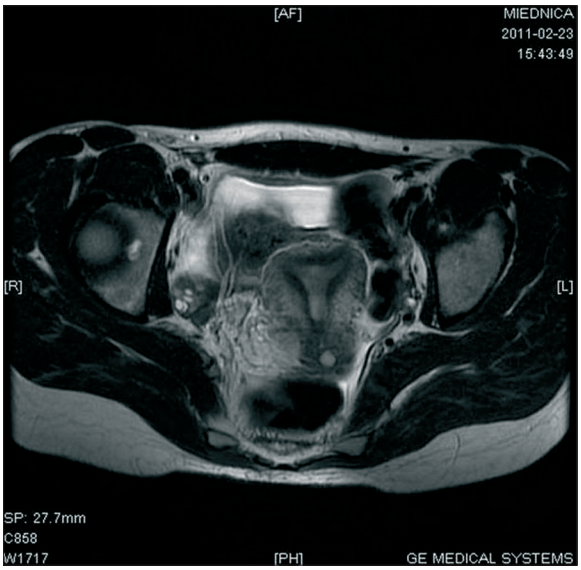

Figure 2. Magnetic Resonance Imaging – first scan. Axial MRI 3 serial scans – the uterus has been shown.

was recommended, patient received ethinyl estradiol with norgestimate. In January 2011 diagnostic laparoscopy was performed. A cystic lesion was revealed under bowel loops, 4–5 cm in diameter, lying retroperitoneal at the level of about 2 cm below the promontory on the right side of the midline.

In April 2011 the patient was admitted to the Department of Operative Gynaecology. Abdominal pain was still reported. The abdomen was soft with mild lower abdominal tenderness on the right during examination. The gynaecological examination revealed normal size uterus, anteflexion, shifted to the left. The adnexa of uterus were without palpable changes. On the right side at the level of second sacral vertebra a cyst, about 4–5 cm in diameter, immobile with tenderness during examination was palpated. Imaging diagnostics was performed: pelvic ultrasound (Figure 1), computed tomography and magnetic resonance imaging (Figures 2–4). Routine blood test and urine analysis findings were within normal limits. Antigen Ca-125 level was 13.9 U/ml. Laparoscopic removal of the lesion was suggested.

During the procedure the uterus of normal size, anteflexion, smooth and movable with macroscopically unchanged adnexa of uterus were shown. In the promontory region on the

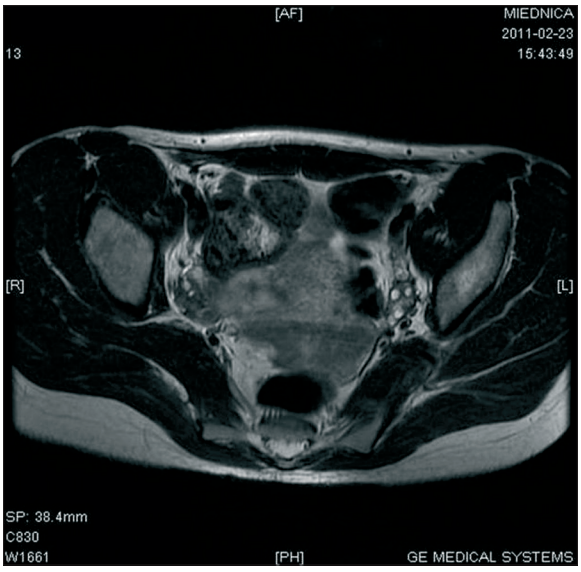

Figure 3. Magnetic Resonance Imaging – second scan. Axial MRI 3 serial scans – on the next scan the ovaries can be seen.

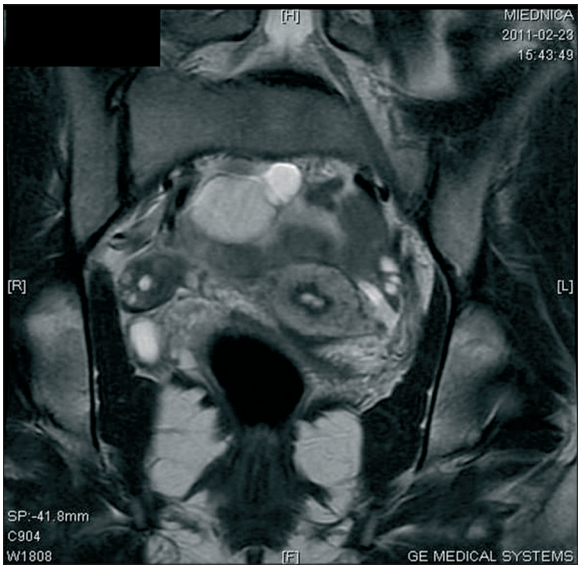

Figure 4. Magnetic Resonance Imaging – third scan. Axial MRI 3 serial scans – we see the lesion that has no topographic connection neither with the uterus nor with the adnexa of uterus.

right side retroperitoneal a lesion with a smooth surface, mobile, 4 cm in diameter was revealed (Figure 5). Peritoneum was cut over the lesion; a smooth cystic dual-chamber change was exposed (Figure 6). The contents of the cyst was aspirated, the cyst was enucleated and transferred to histopathological examination intraoperatively (Figure 7). A frozen section revealed a benign lesion. Result of the definitive histopathological examination stated a mature cyst (teratoma maturum). Operation took about 90 minutes. Blood loss was about 100 ml. The period after surgery was uneventful. The patient was discharged home on the 7th day of hospitalization. Abdominal pain was relieved.

Discussion

Sacrococcygeal teratoma might be asymptomatic in adults; usually it is detected by chance during imaging studies or

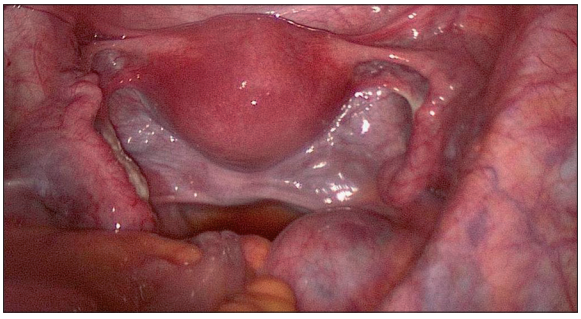

Figure 5. Laparoscopy. During laparoscopy the uterus and the adnexa of the uterus macroscopically unchanged and the presacral tumour localized retroperitoneally were seen.

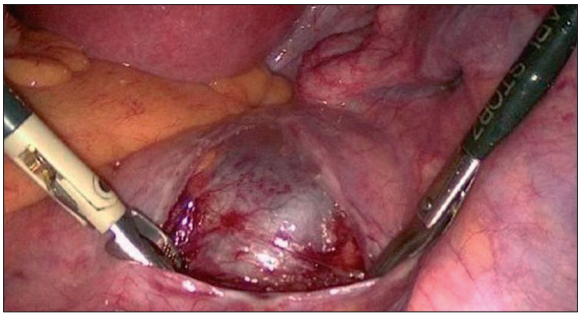

Figure 6. Laparoscopy. Peritoneum was cut over the lesion; a smooth cystic dual-chamber change was exposed.

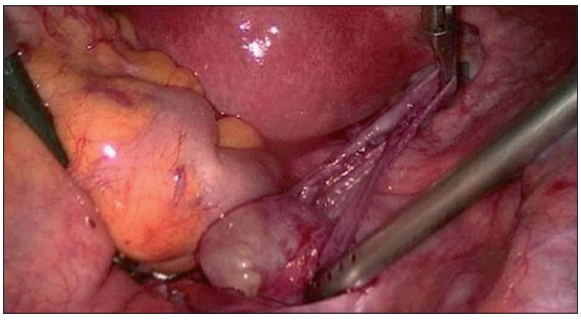

Figure 7. Laparoscopy. Chambers of the cyst were punctured and contents was aspirated. The cyst was enucleated and transferred to histopathological examination during operation.

clinical examination. Symptoms may be subtle and nonspecific as result of compression of adjacent structures: rectum, bladder or uterus [7]. Such symptoms are: lower back and pelvic pain, constipation, sensation of incomplete evacuation, narrowed stools or incontinence. Frequent urination, dysmenorrhoea, nausea, vomiting, oedema and lower extremity paraesthesia have been observed [2,5]. Cases of perinatal complications have been reported [8]. In cases of external growth of the tumour overlaying skin might be puckered and discoloured [9].

Differentiation between cystic changes in ovary and presacral region is extremely difficult; it might even be a diagnostic challenge. In this area we can distinguish the following types of changes: congenital, inflammatory, neurogenic, osseous and miscellaneous [10]. Conducting differential diagnosis should be considered: congenital abnormalities, cystic changes in the ovaries, meningocele, rectal duplication, tailgut cyst, neurogenic tumours, osseous lesions, renal

cysts, Wilms' tumour, soft tissue tumours such as lymphangioma, sarcoma, mucinous adenocarcinoma, cystic mesothelioma, the Mullerian ducts cyst, epidermal cyst, enlarged lymph nodes and metastatic tumours of the uterus or ovaries [1,2,5,11]. The most common tumours that develop in retroperitoneal space are the lymph nodes modified by metastases.

Differentiating presacral changes Currarino syndrome (Currarino anomaly CA) should also be taken into account. Syndrome is autosomal dominant disorder with mutation in homeobox gene HLXB9 located at the 7q36 locus. It is characterized by three main clinical features: presacral tumour, poorly developed sacrum (sacral agenesis), and malformations of the anus and rectum [12,13]. The most common symptom is chronic constipation.

In the case of accompanying inflammation the differential diagnosis is even more difficult and may be misdiagnosed as high lying pararectal abscess, fistulas, granulomas or even tuberculosis [5].

Imaging studies should include transvaginal and transrectal ultrasound, computed tomography and magnetic resonance imaging, which are useful in identifying the exact location and suggest character of the tumour. Such diagnostics is helpful for determining the optimal surgical procedure. Laboratory tests levels of AFP, βhCG, CEA, Ca-125, CA19-9 and CA 15-3 should be done in case of suspicion of malignant transformation [7]. Colonoscopy and sigmoidoscopy should also be considered.

During childhood, 85% of the presacral cysts are benign, but the risk of malignant transformation increases with age. It must be remembered in the case of adults. The definitive diagnosis is possible only after histopathological examination of the changes. Therefore the lesion should always be surgically removed. Drainage or biopsy is not recommended [10].

Due to the rarity of reported changes no firm guidelines exist [10,14,15]. In the literature, depending on the anatomical conditions, descriptions of various surgical procedures are reported. They include anterior abdominal approach, post anal and combined access, also laparoscopic-assisted approach. If removal of the coccyx and the part of the lower sacrum is necessary sacral nerves should be preserved [9]. In case of large tumours located in the abdominal cavity anterior abdominal approach was usually chosen and for changes with external extension posterior access. Reports of massive intraoperative haemorrhage associated with removal of the sacrococcygeal cysts forced changes in the surgical procedures [16]. Bentley was the first to apply ligation of the median sacral artery before attempted resection of the tumour. Then it was recommended to use of vascular clamp above the aortic bifurcation or encircling the distal aorta with a vascular tie. Although these techniques are logical and ensure safety, they require a laparotomy. When benefits of laparoscopy were realized, it was performed as assisting surgical procedure prior to the removal of the lesion from posterior sacrococcygeal approach. The median sacral artery was laparoscopically supplied, and then the tumour was removed via posterior approach, which significantly reduced the number of complications, such as intraoperative haemorrhage [16,17]. lt resulted in more common usage of laparoscopy and combination of different surgical approaches. Just a few descriptions of procedures performed only by laparoscopy are reported. The first description of the laparoscopic removal of the retroperitoneal cyst comes from 1995 [18]. Since then, we find in the literature 3 further cases of presacral teratomas treated by laparoscopic surgery [19].

Conclusions

Decision of laparoscopic surgery is certainly a challenge. The advantages offered by this approach should be considered, especially the facilitated and accurate visualization of pelvic tumour provides more security during removal of the lesion [19]. Faster recovery, a much lower mortality rate, less complications, such as bleeding, adhesions and postoperative pain and cosmetic appearance of the scar are observed. On the other hand, an adequate supply of equipment and operator's experience is necessary. Careful and sequential conducting of the operational procedures ensures the success of the surgery performed by laparoscopy. Teratomas can be easily separated and isolated from surrounding tissues; however, this procedure must be performed precisely by direct visualization. It's necessary to empty the patient's bowel prior to surgery and to inform about the possible risk of its partial removal. The mobilization of the uterus, vagina and rectum is required for removal of the lesion. Careful sharp dissection of the presacral space and tissue near the rectum is necessary to avoid inadvertent trauma or electrosurgical coagulation injury [19]. In our case, important adjacent structures were adequately protected; therefore there were no complications during surgery. In case of large cystic changes decompression of the lesion may be considered by aspirating the contents of the cyst, which facilitates the removal. Blood loss during removal of presacral changes can be significant, therefore with the utmost care tearing of presacral fascia must be avoided, careful dissection within the space between the rectal fascia and Waldeyer fascia helps to avoid puncture of large blood vessels and the ureter [19]. Preservation of the autonomous nerve supply to the bladder and rectum may be difficult. Therefore, postoperative complications (31%) that may be expected are bladder dysfunction (15%), incontinence for faeces (7%) and dysesthesia (7%), especially in the case of damage to pelvic splanchnic nerves [19]. Tumour removal using laparoscopic approach should be taken into consideration especially when a benign lesion is suspected. The risk of recurrence is extremely low with radical resection of the change.

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