—Case Reports—

A Complicated Case of Pregnancy Involving a Presacral Epidermoid Cyst

Daisuke Shigemi[1], Seiryu Kamoi[1], Akihisa Matsuda[2] and Toshiyuki Takeshita[3]

[1]Department of Obstetrics and Gynecology, Nippon Medical School Chiba Hokusoh Hospital, Chiba, Japan
[2]Department of Surgery, Nippon Medical School Chiba Hokusoh Hospital, Chiba, Japan
[3]Department of Obstetrics and Gynecology, Nippon Medical School, Tokyo, Japan

Although presacral developmental cysts, including epidermoid cysts, are relatively rare diseases, an intrapelvic mass found for the first time in early pregnancy should be followed-up with the possibility of presacral developmental cysts in mind to be alert to the signs of local infection and malignancy. We treated a pregnant patient with presacral cystic disease. During pregnancy, percutaneous fenestration was performed because the cyst caused severe compression symptoms and complicated bacterial infection. Laparoscopic total cyst excision was performed after cesarean section. There is no suggested criterion to make a decision for the delivery mode. The mass should be removed completely to reduce the risk of recurrence and malignant progression. (J Nippon Med Sch 2017; 84: 100–104)

Key words: epidermoid cyst, pregnancy, presacral developmental cyst

Introduction

Presacral or retrorectal developmental cysts are rare[1]. The presacral epidermoid cyst, which is a developmental cyst, arises from remnants of embryonic tissues and is detected most often in women of reproductive age. However, presacral epidermoid cysts, which are asymptomatic in 26% to 50% of patients[2], can easily be mistaken for other types of cyst. An intrapelvic mass first observed early in pregnancy should be followed up as a possible presacral or retrorectal developmental cyst, so that signs of local infection or malignancy might be detected if present. The best method of fetal delivery for safety of mother and fetus should also be considered in each case. We report a presacral epidermoid cyst, which could be diagnosed while a woman was pregnant.

Case Report

A 34-year-old, gravida 3, para 2 pregnant woman was referred to our outpatient department after a perinatal follow-up examination for the purpose of thorough examination on an intrapelvic cyst at 9 weeks' gestation. The medical history was uneventful, and family history was not relevant. The patient's first child had been delivered 4 years earlier via cesarean section (CS) because of arrest of labor; an intrapelvic cyst (greatest diameter, 5 cm) was detected with ultrasonography during the puerperium. Symptoms were not present, and the cyst was followed without surgical treatment. A second CS was performed uneventfully 1 year before the present admission. No intra-abdominal cysts, including ovarian cysts, were recognized during the second CS.

During the patient's admission to another hospital for the last pregnancy, the intrapelvic cyst was noted to have increased to 8 cm in diameter and magnetic resonance imaging (MRI) and diagnostic centesis were performed. The MRI examination revealed that the intrapelvic mass was near the forefront of the sacrum and similar to a simple cyst with a smooth wall, no solid part, and high intensity on T2-weighted images. Cytological examination did not reveal malignancy, and a diagnosis of the cyst could not be made at previous hospital.

At initial presentation to our hospital at 9 weeks' gestation, ultrasonography revealed a normal-sized fetus (crown-lump length was 14.8 mm) with a regular heartbeat and the presacral cyst (8×4 cm in diameter). At 10 weeks' gestation, the patient was admitted to our hospital because of acute dysuria, dyschezia, and mild tenderness of the left hip. Blood tests revealed a normal white blood cell (WBC) count and normal level of a C-reactive protein (CRP).

Correspondence to Daisuke Shigemi, MD, Department of Obstetrics and Gynecology, Nippon Medical School Chiba Hokusoh Hospital, 1715 Kamagari, Inzai, Chiba 270–1694, Japan
E-mail: dshigemi@nms.ac.jp
Journal Website (http://www2.nms.ac.jp/jnms/)

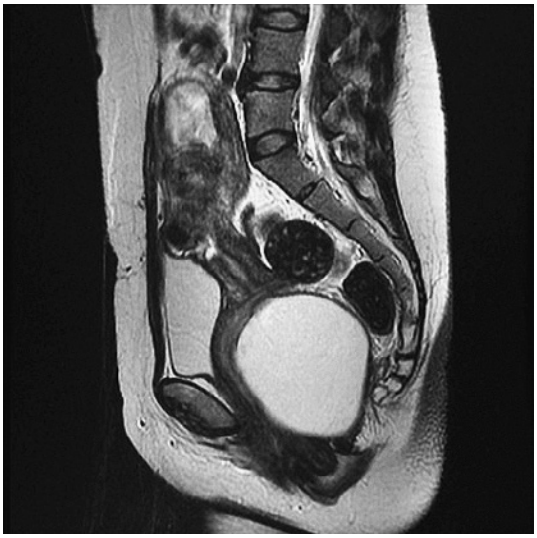

Fig. 1 Pelvic MRI during pregnancy (Sagittal, T2WI). A presacral cystic mass (8×7.5×8.8 cm) with high signal intensity in the T2WI. There was no solid part.

On day 3 of hospitalization, a fever had suddenly developed, the tenderness had increased, and blood tests showed elevated WBC (13,940 /μL) and CRP (8.72 mg/dL). Because the dorsal aspect of the vaginal wall was strongly compressed by the cyst, the vagina was extremely tender and could not be inserted with a speculum. Therapeutic and diagnostic centesis was performed via the vaginal wall on day 3, and a yellow, foul-smelling fluid was collected. The fluid contained many Gram-negative and Gram-positive bacteria, including anaerobes, but no malignant cells. The cyst soon decreased to 4×5 cm in diameter and the tenderness became less severe on the next day. Antibiotics (sulbactam/ampicillin) were administered, and vaginal irrigation was started. Although each condition improved for eight days, fever and elevated WBC count recurred 9 days after centesis.

A non-contrast-enhanced MRI examination of the pelvis revealed a presacral cystic mass without the solid part (**Fig. 1**) on day 12 of hospitalization. Surgical treatment, which included a percutaneous fenestration and placement of an indwelling drain into the cyst under spinal anesthesia, was performed with informed consents on the same day. The fenestration, drainage, and antibiotics worked well against the infection of the cyst; the patient could be discharged without a drainage tube at 13 weeks' gestation; however, the intrapelvic cyst maintained its size (approximately 4×5 cm in diameter). The fetus had no obvious abnormalities and showed appropriate growth.

Before delivery, the intrapelvic cyst was thought to be a presacral epidermoid cyst because of its site of occurrence and existence of many squamous epithelial cells detected in samples taken from aspirated fluid. The intrapelvic cyst was followed-up via vaginal ultrasonography and maternal blood samples, including some tumor makers. The cyst did not increase in size and there were no other signs of malignancy, such as newly developed papillary mass in the cyst and elevated tumor markers, between 14 weeks' gestation and 37 weeks' gestation. The tumor markers were within the normal range (squamous cell carcinoma antigen, 0.6–0.9 ng/mL; cancer antigen 125, 13.6–30.2 U/mL; and carbohydrate antigen 19–9, 1.6–2.3 U/mL).

A healthy 2,805-g female neonate was delivered with CS at 37 weeks' gestation, with Apgar scores of 9 at 1 minute and 10 at 5 minutes. The indication of CS was the patient's previous CS. The intrapelvic cyst could not be detected in the abdominal cavity; both ovaries were observed to be normal during CS.

Three months later, laparoscopic total cyst excision was performed with the patient under general anesthesia. The adjacent mesosigmoid was exfoliated from the inside to the lateral side, leading to the mobilization of the sigmoid to the rectum. Circumferential exfoliation of the mesorectum preceded toward the anus. The cystic mass was found in a deep presacral space, which firmly adhered to the puborectalis, external anal sphincter, and left posterior side wall of the lower rectum. By using laparoscopic coagulation scissors we achieved complete excision of the cyst without rectal wall injury. The cyst was a monoloculated mass adhering to the rectum and sacral bone and extending into the presacral area. The lesion was completely removed (**Fig. 2**). Microscopic and histopathological examination of the cystic mass revealed it to be a nonmalignant epidermoid cyst (**Fig. 3, 4**).

The patient has been regularly followed up for 15 months and has shown no signs of recurrence.

Discussion

Presacral epidermoid cysts are categorized as developmental cysts. A precise classification and description of each feature have been described in detail previously[1]. We searched for the previous reports on I-CHU-SHI (a database of Japanese literatures) and PubMed. In Japan, 104 adults with presacral epidermoid cysts were reported from 1983 through October 2015. Seventy-six of the patients were women. The common chief complaints were tenderness, palpable mass, and compression symptoms

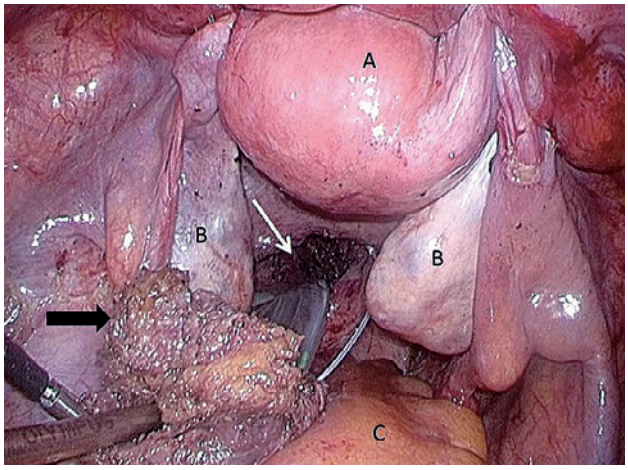

Fig. 2 Laparoscopic finding in the pelvic cavity. Presacraltumor (**broad arrow**) was resected from the presacral cavity (**narrow arrow**). A represents the uterus, B represents the ovaries, and C represents the rectum.

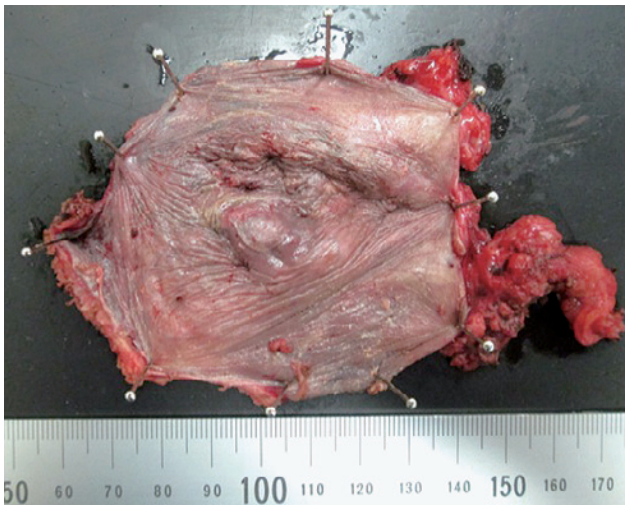

Fig. 3 Macroscopic view of the cyst

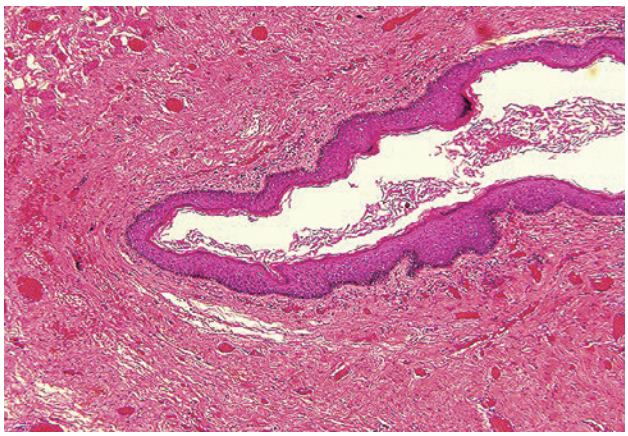

Fig. 4 Pathological feature of the presacral epidermoid cyst; H.E. stain

Table 1 Adult patients with presacral epidermoid cysts in Japan from 1983 through October 2015

Clinical characteristics	Number of patients (percentage of 104 patients)
Age, years	48 (range, 17–83)
Sex	
Male	28 (26.9%)
Female	76 (73.1%)
Chief Complaint	
Tenderness	24 (23.1%)
Palpable mass	20 (19.2%)
Compression symptoms (Disorder of urination or defecation)	20 (19.2%)
Asymptomatic	28 (26.9%)
Tumor size, centimeters	8.3 (range, 2–30 cm)
Malignancy	7 (6.7%)
Surgical approach	
Transsacral	63 (60.6%)
Transabdominal	22 (21.2%)
Laparoscopic	7 (6.7%)
Others	12 (11.5%)

(disorder of urination/defecation), but 28 cases had no subjective complaints. Seventy percent of the cases were of cysts 10 cm or larger (**Table 1**).

Three important factors should be considered in the follow-up of pregnant patients with presacral developmental cysts. First, an estimated diagnosis of developmental cysts should be made. In the case of a presacral cyst detected by ultrasonography, there are some possible diagnoses, such as ovarian tumors, sacral meningocele, anorectal abscesses, and pilonidal diseases. In our case, therapeutic and diagnostic centesis of the infected cyst during pregnancy contributed to a correct diagnosis due to the detection of epithelial cells in the fluid. However, a clinical case of malignant metastasis due to centesis of a developmental cyst with squamous cell carcinoma has been reported[3]. Centesis also has a risk of bacterial infection. Therefore, rapid diagnostic centesis of the cyst should not be recommended. At present, the accurate test to judge the malignancy of presacral developmental cysts is unknown. An increase in tumor markers in the serum and content fluid of the developmental cyst does not always indicate malignancy[4]. Some oncofetal antigens (e.g., α-fetoprotein, human chorionic gonadotropin, carcinoembryonic antigen, cancer antigen 125) could be affected by a change of fetal development and differentiation. Usually, the levels of those antigens are elevated during gestation period. In case of abnormal placentation or fetal

Table 2 Pregnant women with presacral developmental cysts

Authors reporting	Year	Diagnosis	Tumor size (cm)	Delivery mode	Treatment after delivery
Gerwig WH Jr[7]	1954	Presacral dermoid or teratoma	2	Vaginal delivery	Resection
Shimanuki et al[8]	1984	Epidermoid	9.4×8.6×10.4	Vaginal delivery	Resection
Akimoto et al[9]	1984	Dermoid	12×9×7	Unknown	Resection
Shinmyo et al[10]	1993	Epidermoid	8×6×6	Cesarean section	Resection
Toriguchi et al[11]	2010	Dermoid	7	Vaginal delivery	Resection
Chung et al[12]	2012	Tailgut cyst	4.5	Vaginal delivery	Resection
Present case	2015	Epidermoid	6×4×4	Cesarean section	Resection

complications such as preeclampsia, trisomy 21, and open neural tube defect, they may be abnormally elevated[5]. Computed tomography would be less useful because it often leads to ovarian cyst diagnosis, and the effect of radiation must be considered in pregnant women. A pelvic MRI may be more useful for assessment because it can determine accurate position, size, and presence or absence of a solid part of presacral cysts. Epidermoid cyst generally appears hypointense on T1-weighted images and hyperintense on T2-weighted images[6].

The second important factor is the delivery mode. Seven cases, including our own, of pregnant women with presacral developmental cysts have been reported in detail in the Japanese and English literature by searching on I-CHU-SHI and PubMed. Four patients delivered vaginally and two underwent CS (both because of prior CS; **Table 2**)[7–12]. There is no suggested criterion for the delivery mode. However, we considered that a presacral epidermoid cyst in itself is not always a contraindication for vaginal delivery; therefore, most patients can deliver vaginally with careful observations. With regard to our case, we would select vaginal delivery if she had no past history of CS.

The final significant factor is treatment. Complete surgical resection should be performed essentially as soon as possible because this disease has a risk of pressure symptoms due to enlargement, infection, and malignancy. However, surgical treatments during pregnancy have a risk of premature delivery and fetal death. Therefore, surgical resection would be better performed after childbirth. Although there is no clear evidence, surgery for pregnant women should be considered when malignant signs, such as an acute elevation of tumor markers (SCC antigen, α-fetoprotein, and carcinoembryonic antigen) in serial measurement, development of a solid part in the presacral cyst, and rapid increase in size, are detected. According to Japanese reports, 7 cases (6.7%) were diagnosed in which carcinoma (all squamous cell carcinomas) developed in the epidermoid cyst. Various surgical approaches can be selected depending on the location, size of the tumor, and degree of an inflammation and adhesion[2,13,14]. Laparoscopic surgery can be selected for complete resection. In Japan, a transsacral surgery was the most popular approach.

Epidermoid cyst is a frequent benign cutaneous tumor, but it can be detected rarely at the presacral region as the developmental cysts, and it also can occur in pregnant women. The mass should be removed completely to reduce the risk of recurrence and malignant progression.

Conflict of Interest: The authors declare that there is no conflict of interests.

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