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CASE REPORT

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Case report of experience of misdiagnosis of Currarino syndrome as ovarian cyst

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Abstract

The medical information was collected for Currarino syndrome. The patient had anal surgery for congenital anal stenosis when 2 years old. Clinical manifestations were pelvic cystic mass and low abdominal pain. The pelvic mass was found with a diameter of about 20 cm during the transabdominal exploration. At the second day after operation, the patient complained of neck pain. Neurosurgeon performed surgical treatment and diagnosed it as anterior sacral meningocoele. Currarino syndrome has female pelvic mass, sacral malformation, and congenital anorectal malformation, blindly puncture or drainage before operation should not be permitted.

KEYWORDS

case report, currarino syndrome, ovarian cyst

1 | BACKGROUND

ASM is due to embryonic mesodermal ganglia hypoplasia, only about 300 cases have been reported since 1937. ASM is divided into congenital and acquired.[1] Acquired ASM is characterized by abnormal dural structure and is associated with connective tissue disorder.[2] Because the part of meningocele is concealed in the pelvic cavity, if there are no symptoms such as nerve injury or nerve compression, the clinical diagnosis is difficult. Currarino syndrome is usually characterized by anterior sacral mass, sacral malformation, and congenital anorectal malformations. It is an autosomal dominant genetic disease HLXB9 gene mutation on chromosome 7, short arm 36[3] . More than 95% patients have constipation symptoms that occur in infancy.[4] Early diagnosis is difficult, and tumor enlargement may rupture, affecting the quality of life of patients. A misdiagnosed Currarino syndrome was analyzed. An informed consent was signed by the patient.

2 | CASE PRESENTATION

A 25-year-old Chinese woman was admitted to hospital because "she was found pelvic mass 6 years ago with lower abdominal pain for 19 h." Current medical history: she felt

consciously lower abdomen enlargement 6 years ago, the mass bottom was above umbilical two fingers and was not untreated. The patient had sudden lower abdominal pain 19 h ago with dizziness, nausea, and no vomiting. Blood HCG was negative. B ultrasound: A giant cystic anechoic area was seen at the posterior side of the uterus, the size was about 178 × 136 mm, the boundary was clear, its internal echo was clear, and the septal echo was seen inside (Figure 1).Past history: she had anal surgery for congenital anal stenosis in Xi'an Children's Hospital when 2 years old. She had sex history and did not marry. Family history: she denied family history of hereditary diseases. Body examination: there was about 10 cm long longitudinal incision in the left lower abdomen. There was abdominal bulge and muscle tension. a diameter of about 20 cm cyst, clear boundary, tenderness, no rebound pain, poor activity in middle and lower abdomen was touched. Anal: Anus mucosal was valgus.

CT:1. pelvic giant cystic space occupying lesion. 2. sacrococcygeal deformity. During the transabdominal 10 cm longitudinal exploration, the pelvic mass was found with a diameter of about 20 cm, cystic, clear boundary and complete capsule, located behind the peritoneum. The tumor protruded upward through the posterior rectal space, with the right ovary and fallopian tube structure in the upper left, small uterus, dense bladder adhesion, and normal left adnexa. We punctured cyst for decompression, extracting clear and transparent fluid, and then cystectomy with cooperating gastrointestinal surgeon. The patient was indwelled 1 pelvic drainage tube postoperative. At the second day after operation, the patient complained of pain neck back, and at the third day after operation, the patient complained of chest pain after sitting up about 1 min, feeling dizziness, dyspnea, difficulty in lifting the right upper limb, and relieving after bed rest. Daily pelvic drainage after operation was about 300–400 ml with pale yellow color, clear. Body examination: no neck ankylosis, Kernig sign negative. Postoperative pathology: there was no coated epithelium in the fibrous cystic wall, and lymphocytic infiltration was seen in some areas of the cystic wall, which was consistent with benign cysts (Figure 2). Lumbar vertebrae, sacrococcygeal CT plain scan showed sacrococcygeal deformity (Figure 3). Pelvic drainage fluid assay: coagulant positive (+), the total number of cells $2000*10^6$/L,WBC $809*10^6$/ L. CSF Biochemistry: M-TP (CSF)5259.70 mg/L, Cl (CSF)112.3 mmol/L, GLU (CSF)2.5 mmol/L. Lumbosacral plexus nerve MRI plain scan + enhancement: lumbar lateral bending, lumbar 5, sacral 1 vertebral body mismatched with sacrococcygeal

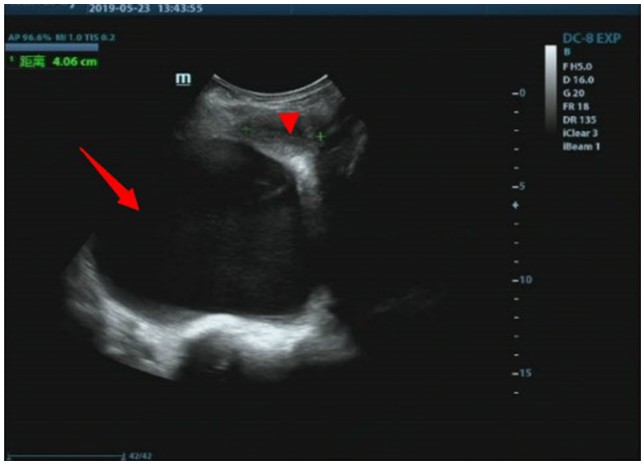

FIGURE 1 Preoperative ultrasound findings showed giant cystic mass in the pelvic cavity

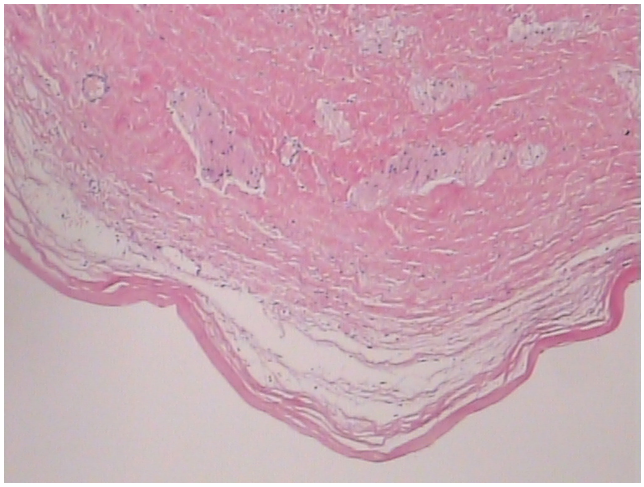

FIGURE 2 Pathology showed benign cysts

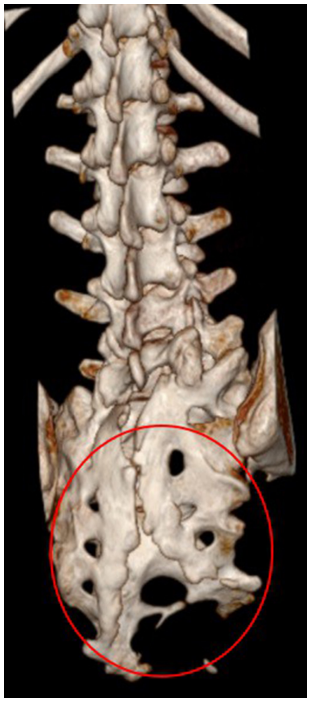

FIGURE 3 CT showed sacrococcygeal vertebra deformity

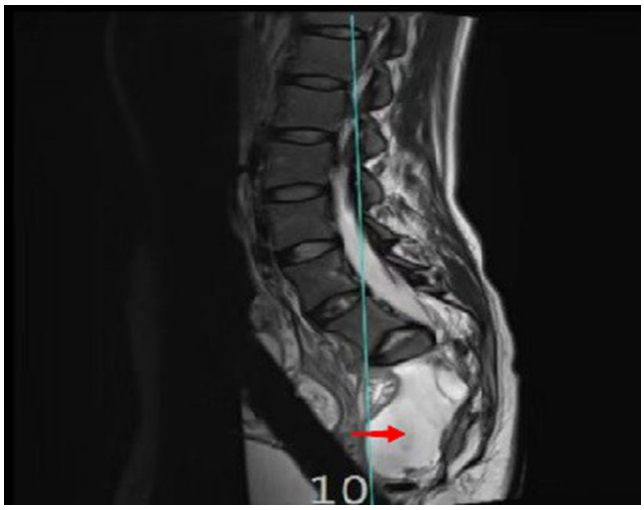

FIGURE 4 Lumbar lateral bending, lumbar 5, sacral 1 vertebral body mismatched with sacrococcygeal vertebrae shape disorder in MRI

vertebrae shape disorder(Figure 4). Multi-disciplinary treatment was carried. Neurosurgeon performed surgical treatment: lumbosacral approach microscope subacral anterior meningocele repair + dural repair + spinal canal decompression + right thigh fat resection. We modified diagnosis: 1 neural tube developmental malformation: ASM; 2 cerebrospinal fluid leakage; 3 sacrococcygeal malformation; 4 uterine dysplasia. The patient recovered and was discharged. The patient followed up without discomfort, pelvic B ultrasound was not abnormal.

3 | DISCUSSION

Constipation, abdominal pain, urinary retention, and lower back pain were the first symptoms of Currarino syndrome.[1] About two thirds have symptoms, 27 per cent have neurological symptoms, 30 per cent have symptoms of the genitourinary system, and these symptoms usually occur at 20 ~ 30 years old.[5]The clinical manifestation of this patient was pelvic mass and abdominal pain. CT showed sacral defect, congenital anal stenosis, which met the triple sign, but the preoperative clinician and imaging physician did not diagnose the Currarino syndrome. The main reason was that the disease was not well understood, and the neurosurgeon was not consulted before operation.

Dvthel review 11 cases summarized the following clinical characteristics: (l) Dysfunction of defecation and urine. (2) Lower extremity phlebitis and increasing intracranial pressure. (3) Sciatica.[6] We do not blindly carry out puncture or drainage before operation, avoid sudden intracranial pressure drop or central nervous system infection, and endanger the patient's life, especially transanal or vaginal puncture.[7] This patient's preoperative headache was not obvious, we consider postoperative headache reason was the cerebrospinal fluid leakage after surgery to cause low intracranial pressure. It should not affect the menstruation and ovulation. Premature delivery may be caused by mass compression for part of patients.

It should not affect the menstruation and ovulation. Premature delivery may be caused by mass compression for part of patients. The mortality rate of patients with conservative treatment reached 30%, mainly due to meningitis,[8] so surgical treatment is recommended. There are three main types of surgical approaches: (1) posterior sacral approach: it is suitable simple ASM, the main purpose of the operation is to repair the fistula mouth to relieve the symptoms of compression. (2) transabdominal approach: it is due to preoperative failure to identify pelvic ASM. The anterior transabdominal approach has the risk of visceral injury, and the nerve structure is not clearly displayed. However, the anterior approach is suitable for complex cases with large lesions or involving adjacent visceral cases.[6](3) Cystic cavity abdominal shunt: Simple drainage cannot relieve the symptoms of compression for the larger cyst. Some scholars advocate the first sacral approach to repair the expansion outlet and then resect teratoma through the abdominal.[6] In this case, the patient was performed first through the abdominal approach because of misdiagnosing pelvic mass, then through the sacral approach for meningocele repair + dural repair.

To sum up, the patient should be taken into account the Currarino syndrome with female pelvic mass, sacral malformation, and congenital anorectal malformation and is not been blindly punctured or drainaged before operation, especially through the anus or vagina puncture. MRI examination and neurosurgery consultation should be considered. In the future, early diagnosis is researched with neurosurgery and imaging department.

AUTHOR CONTRIBUTIONS

Aiwen Le carried out the studies, participated in collecting data, and drafted the manuscript. Ya hong Xu participated in acquisition, analysis, or interpretation of data. Zhonghai Wang participated in reviewing and proofreading papers. All authors read and approved the final manuscript.

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CONFLICT OF INTEREST

The authors declare that they have no competing interests.

DATA AVAILABILITY STATEMENT

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

ETHICAL APPROVAL

All procedures performed in studies involving human participants were in accordance with the ethical standards of the Huazhong University of Science and Technology Union Shenzhen Hospictal, Shenzhen Nanshan People's Hospital research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. Publish this information was obtained from study participants.

CONSENT

The consent for publication was obtained from the patient who has signed the consent form.

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