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CASE REPORT

A giant Müllerian duct cyst in the perineum: a case report

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Abstract **The authors present a case report of a giant Müllerian duct cyst in the perineum. A 37-year-old man presented with a mass with the size of 50 cm × 40 cm × 30 cm in the perineum. Computed tomography (CT) scan and magnetic resonance imaging (MRI) of the pelvis and perineum found a huge multilocular cystic mass. X-ray film of the pelvis showed a giant soft-tissue shadow of high density in the perineum. Voiding cystourethrogram did not show any cyst interlinked to posterior urethra. After related examinations, the patient underwent open surgical resection of the cyst. Pathologic examination confirmed the presence of a Müllerian duct cyst. Huge Müllerian duct cyst is uncommon in clinic. Treatments of Müllerian duct cysts depend on their sizes and symptoms.**

Keywords Müllerian duct cyst; clinical symptoms; diagnosis

Background

Müllerian duct cysts result from an abnormality in regression of the Müllerian system. They are rare congenital anomalies with an autopsy incidence of 1% in men [1] and clinical prevalence of 5% in urologic patients [2]. As they are asymptomatic in about 60% cases, they are probably underreported according to Coppens [2]. However, Müllerian duct cyst which can discharge 5 000 ml liquid that we have ever seen is extremely rare in clinic.

Enlarged prostatic utricles and Müllerian duct cysts both originate from Müllerian duct remnants [2,3]. Although these entities share a similar embryologic origin, they are clinically different. So when diagnosing a Müllerian duct cyst, we must exclude enlarged prostatic utricle. And due to the different symptoms and sizes of cysts, treatments of Müllerian duct cysts are not all the same. Recently we diagnosed and treated an unusual case of a giant Müllerian duct cyst in the perineum. The purpose of this report is to describe our experience with diagnosis and treatment and to review the literature in order to provide a comprehensive assessment of Müllerian duct cysts.

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Case report

A 37-year-old male from Jiangxi Province with a 5-year history of a mass in the perineum was investigated in our department in May 2010. The patient was diagnosed as pelvic cyst six years ago for frequent urination and difficult defecation, and an exploratory laparotomy with excision of cyst was performed. The pathologic diagnosis after the operation was Müllerian duct cyst. In the next year, the pelvis cyst recurred two times followed by two cyst excisions. And the pathologic diagnoses were the same as before. Since 2005, a chestnut-sized lesion on the patient's right buttock progressively enlarged. Multiple paracenteses were performed and liquid discharge was up to 5 000 ml at most. The cystic fluid chyle test was negative. And no tumor cell and sperm were found within the fluid.

In May 2010, the mass with the size of 50 cm × 40 cm × 30 cm in the perineum and right buttock was witnessed (Fig. 1A). Large veins were seen on its surface, and there is no pressing pain and diabrosis. CT scan and MRI revealed a huge multilocular cystic mass in the perineum and pelvic, and the thickness of cystic walls were uneven (Fig. 2). X-ray film of the pelvis showed a giant soft-tissue shadow of high density in the perineum. Voiding cystourethrogram did not show that the cyst interlinked to posterior urethra.

Upon completion of related examinations after admission to hospital, the patient agreed to perform perineum cyst excision. After a 50 cm skin incision was done in the right

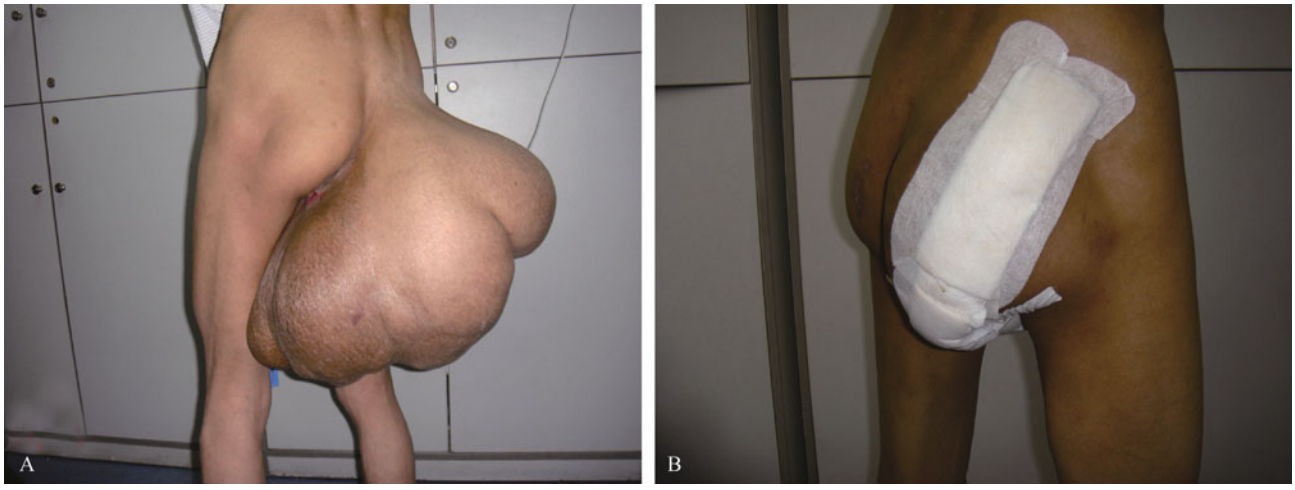

Fig. 1 The pre- and post-operative images of the patient. (A) Pre-operation. The patient could not even walk for a short time before operation and had symptoms such as frequent and arduous urination and so on due to the huge cyst, which seriously impacted the quality of life of the patient. (B) Post-operation. The patient could stand and move spontaneously, and symptoms of difficult urination and defecation had been significantly improved.

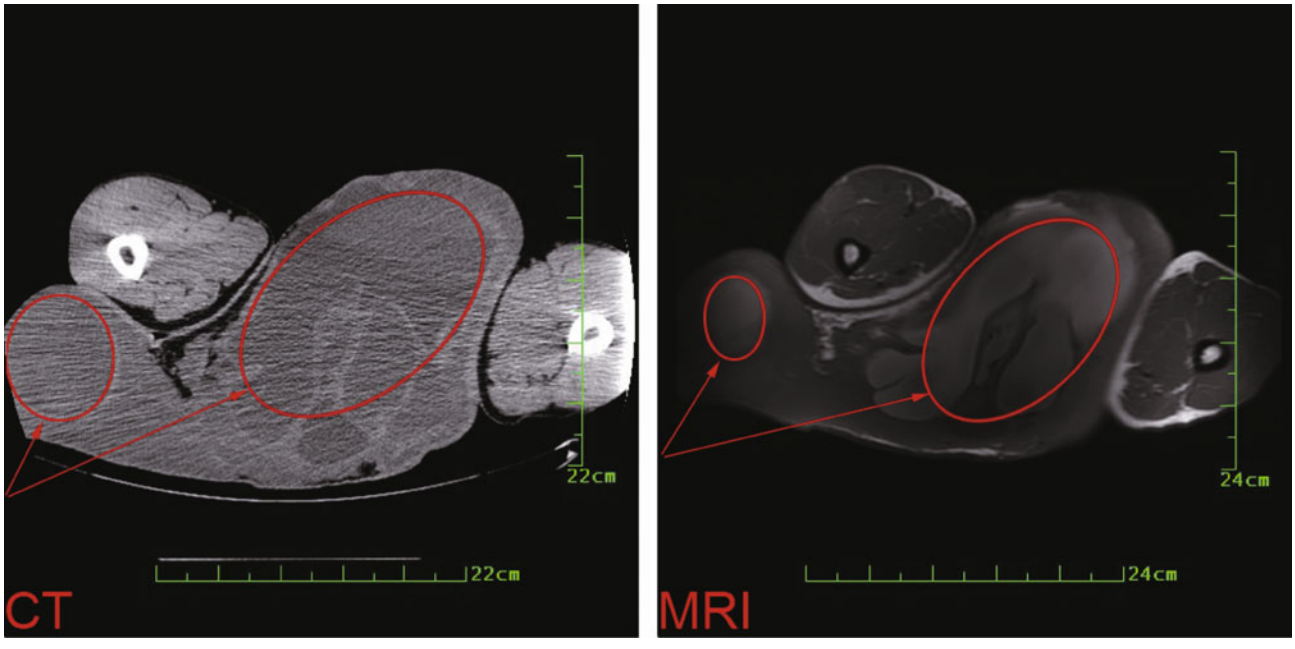

Fig. 2 The CT scan and MRI images of the cyst. CT scan and MRI suggested a huge cystic mass in perineum and pelvic, which was exactly a multilocular cystic mass, and the thickness of cystic walls were uneven.

buttock and perineum, we detected a multilocular cystic mass with diameter between 2 cm and 30 cm, and abundant blood supply. The diameter of the largest blood vessel was up to nearly 1 cm. Due to the size of the wound surface and errhysis, 26 units of suspended red blood cells and 1 600 ml blood plasma were transfused. The cyst was cut open after the surgery and part of the cyst fluid was clear, and part was brown. The cyst fluid weighed 7 kg in all. And the cyst wall weighed 4 kg, of which thickness ranged from 1 mm to 6 mm.

Post-operative course was uneventful. The patient recovered well and regained normal voiding function (Fig. 1B). When discharged from the hospital he could stand and move spontaneously. The pathologic diagnosis after the operation confirmed Müllerian duct cyst (Fig. 3).

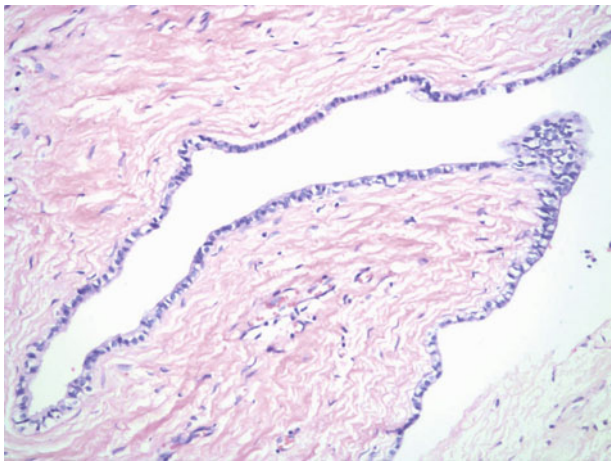

Fig. 3 The pathological sections of patient's cyst. The pathological sections of patient's cyst walls indicated that cyst walls were mainly constituted by fibrous connective tissue, and simple columnar epithelium is visible on the local scale. Different parts of the cyst walls showed edema, glassy degeneration and infiltration of inflammatory cells (H&E, original magnification 200 ×).

Discussion

Gyneducts turn up during the 6th week of embryo and differentiate to genital organs for female or male rapidly. Then they begin to atrophy from the 10th week of embryo as sertoli cells secreting Müllerian inhibiting substance (MIS) [3]. Hyposecretion of MIS or sinusurogenitalis, or male phenotype deficiency may cause incomplete degradation, causing Müllerian duct remnant [3].

Cyst originated from Müllerian duct remnant can be differentiated to enlarged prostatic utricle or Müllerian duct cyst [1–3]. There is some confusion in the literature as the terms "enlarged prostatic utricle" and "Müllerian duct cyst" tend to be used interchangeably[4]. For the former, 90% is accompanied by hypospadias or hermaphroditism, which most often happens to children with cystic cavity connecting to posterior urethra [1]. The latter is most common with adult male with normal external genitals. Its cystic cavity is not connected to posterior urethra. Instead, it is located behind prostate or bladder as a cystic mass and its size could be huge [2,3]. Around 60% of Müllerian duct cyst patients have no symptoms related to cyst. Onset of illness usually happens to 30 to 40 years old people who have symptoms. The major symptoms include difficulty in urinating, frequent micturition, urinary retention, urinary tract infection, perineal pains, thin stool, and intrapelvic mass [5]. Acute urine retention caused by Müllerian duct cyst also was reported [6].

The mass of this patient initiated from the pelvic cavity, and related clinical manifestations were difficulty in urination and defecation. It was confirmed that the recurred mass was not only located in the pelvis, but also expanded to the perineum. CT scan and MRI suggested a huge cystic mass in perineum. Voiding cystourethrogram showed that the cyst did not interlink to posterior urethra and the patient had normal external genitals. Without hypospadias, and according to the post-operative pathological report, the diagnosis conclusion was Müllerian duct cyst [1–3].

Treatments of Müllerian duct cysts depend on whether patients have any clinical symptoms, complications and/or the sizes of the cysts. Small-size Müllerian duct cysts without symptoms can be monitored regularly, and don't need treatment. For those infected, antibiotics should be used. For bigger size cysts but confined to prostate and bladder, transurethral unroofing or bladder cyst marsupialization can be performed. It was reported that some cases were cured via abdomen or rectum percutaneous ethanol injection (PEI) guided by ultrasound [2]. For patients with huge cyst like this case, open surgical excision is the choice [3,5]. With the development of laparoscopic techniques, laparoscopic excision of Müllerian duct cyst is feasible for surgeons. Compared with open surgical approaches, laparoscopic excision can afford minimally invasive access to the retrovesical space, a clear view of the deep pelvic structures, reduce the incidence of injury to adjacent structures, and decrease the time of recovery [1,7,8].

As far as we know, there are no published reports of Müllerian duct cyst recurrence after operation. This may relate to the low reported rate [2] and few follow-ups after surgery [5,9]. In this case, the cyst recurring three times should arouse our attention. But whether the recurrence is related to surgical methods is unclear. Okur *et al.* used open surgery excising Müllerian duct cysts in 6 patients, and had found no recurrent evidence during 2–4 years of follow-up [10]. Aminsharifi *et al.*, Hong *et al.* and Lima *et al.* reported using laparoscopic removal of Müllerian duct cysts in 2, 6 and 6 patients with 7 months, 3–56 months and 8–48 months follow-up, respectively. And there were also no cyst recurrence [1,7,8]. Besides, after our surgery, this patient did not recur in 2 years follow-up. So there still need greater sample sizes and longer follow-up period to determine whether the cyst recurrence is related to surgical options.

From this case report, we can learn that Müllerian duct cysts should be diagnosed in time and treated thoroughly. Otherwise, the cyst may recur or progressively enlarge like this case.

Conflict of interest All the authors confirm that there is no conflict of interest.

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