Case Report
Chordoma of vagina: a case report and review of the literature

Wei He, Shuaibin Liu, Xiaoling Gan, Lina Hu

Department of Obstetrics and Gynecology, The Second Affiliated Hospital of Chongqing Medical University, Chongqing 400010, P.R. China

Received October 29, 2017; Accepted March 27, 2018; Epub May 15, 2018; Published May 30, 2018

Abstract: Chordoma is a rare, low-grade malignant tumor originating from the notochordal remnant, usually located in axial skeleton, especially sacral and cranial. Chordoma of vagina is extremely rare and has never been reported up to now. In this case report, we discuss the clinical characteristics, diagnosis and therapeutic strategies of vaginal chordoma combined with literature review, to further explore the features of vaginal chordoma and raise awareness of vaginal neoplasms. A 58-year-old woman presented with a three year history of vaginal mass and a 6 day history of colporrhagia after menopause for 16 years. After the surgical excision, the histopathological examination and immunohistochemical confirmed it to be vaginal chordoma. With one year follow-up, local recurrence and metastasis has not yet been observed.

Keywords: Vaginal neoplasms, chordoma, clinical manifestation, histopathology, treatment

Introduction

Chordoma is a rare, low-grade and locally invasive malignant tumor arising from the embryonic remnant of notochord, which often occurs in the axial skeleton, especially cranial, spinal, and sacral. Morbidity is reported to be 0.8/100000 and the incidence ratio between male and female was 2:1. Within the axial skeleton chordoma, 32% of cases were cranial, 32.8% spinal, and 29.2% sacral [1]. Chordoma in the extra-axial skeleton such as nasopharynx, soft tissue and skin are far less frequent. Rarest of all is the chordoma arising in the vagina, and as a particular case it has never been reported before. We analyze the clinical characteristics of vaginal chordoma combining with some relevant literatures to summarize some new diagnose and treatment experiences in vaginal tumors.

Case report

A 58-year-old woman admitted to The Second Affiliated Hospital of Chongqing Medical University with complaints of a vaginal mass over 3 years and colporrhagia for 6 days after menopause for 16 years. Three years ago, the patient found an asymptomatical tumor mass in her vagina which was about 2 cm in diameter, but she left it untreated. Six days before entering our hospital, she suffered from vaginal bleeding and the volume was as much as menstruation while she was walking. So she sought medical attention in the Maternal and Child Health Hospital of Hechuan county. The ultrasound revealed the tumor mass to be a vaginal cyst. Under anti-inflammatory and hemostasis treatment for 3 days, bleeding stopped and then she transferred to our hospital. The transvaginal ultrasound showed that the endometrium was 3 mm thick. A 44 mm × 43 mm × 51 mm mixed echo mass with clear edge could be found on the right wall of vagina, in which moderate and high level echoic mixed with some low echo area could be seen on the right wall of vagina. Gynecological examinations found an approximate 6 cm × 5 cm × 5 cm hard mass on the right wall of vagina whose...
Chordoma of vagina

Surface was smooth without ulceration or active hemorrhage. It had a distinct border and was close to pubic bone and ischium, and had neither space to vagina nor rectum. The vagina and rectum wall adherent to the mass were immovable. The cervix, corpus uteri and bilateral adnexa were normal.

On October 3rd, 2016, she underwent a pelvic computed tomography (CT) which revealed a 10 mm × 57 mm × 59 mm irregular solid cystic mass with mixed density image on the right wall of vagina. Enhancement scanning showed a heterogeneously enhancing mass. The tumor mass had no boundary to the right wall of vagina and clinged to right external obturator muscle. The fatty space surrounding the lesion is clear. It indicated a malignant tumor mass which might come from vagina (Figure 2).

On November 7th, the pelvic magnetic resonance (MR) showed an irregular slightly long T1 and long T2 mixed signal on the right pelvic floor-ischiorectal space, in which a 67 mm × 51 mm lumpy and schistose short T1 and long T2 signal could be seen. Enhanced scan showed inhomogeneous enhancement. The lesion and the right boundary between mass and vagina was not clear. The boundary between the mass and the right lateral obturator muscle was not clear. The surrounding fat space was vague, and the right inferior ramus and the ischium were not changed. It indicated a tumor mass in the ischiorectal space of pelvic floor and vagina (Figure 3).

After the consultation of gastroenterologist and orthopedist, the patient got vaginal chordoma resection surgery under general anesthesia. In the surgery, we found a 6.0 cm × 5.0 cm × 5.0 cm movable tumor mass on the right wall of vagina limitless to the surrounding tissues and organs. It had clear edge and was close to pubic bone and ischium, and had neither space to vagina nor rectum. The vaginal wall and rectum wall clining to the mass could not be moved. We incised the vaginal wall toward the surface of tumor and tried to isolate it to other tissue, but we found it difficult to make blunt separation for no clear boundary existed. So we

Figure 1. Transvaginal ultrasonography: A. A 44 mm × 43 mm × 51 mm mixed echoic mass on the right wall of vaginal wall. B. The inner construction is even with abundant blood flow signals.

Figure 2. Plain CT abdomen and enhanced CT of abdomen: A 10 mm × 57 mm × 59 mm irregular solid cum cystic mass with mixed density image can be seen on the right wall of vagina.
used ultrasound knife to sharp dissected till the mass was totally resected. We put negative pressure drainage tube in the residual cavity. Then we stitched the wound normally and packed a piece of gauze in the vagina followed by pressure dressing. The total hemorrhages quantity was 200 ml. The dissected specimen was a 6.0 cm × 5.0 cm × 5.0 cm solid mass with some irregular cystic structures filled with coffee liquid. The postoperative pathologic examination defined it to be chordoma (Figure 4). The patient’s postoperative recovery was well and no recurrence or metastasis has occurred during the one year follow-up.

Discussion

Chordoma’s clinical symptoms depend on the location of origin and the nonspecific symptoms often lead to a delay in diagnosis. Intermittent pain of lumbosacral portion and the local compression are the main symptoms of sacral chordoma. If the rectum is compressed, the patient would have a change in bowel. If bladder pressed, dysuria, urinary frequency, and urgency are common. All above could be misdiagnosed as cystitis or rectitis. If the nerve root is pressed, it would induce radiculalgia and paraesthesia. For some patients, a hard and unmovable tumor mass in the presacral region can be found by rectal examination, who may suffer from perianal skin hypoesthesia, weakness of muscular tension, or looseness of the anal sphincter. Nasopharynx chordoma’s clinical manifestation include headaches, rhinobyon, Epistaxis, dysphagia, or throat pain [2, 3]. Cranial base chordoma may lead to headaches, blurred vision, hypomnesia, emotional lability, ptosis, tongue weakness, ataxia, or dysphagia [4-6].
In this case, the lesion was on the vaginal wall, protruding into the vagina without limitation to the vagina and was not connected with sacral. The patient had a painless swelling for three years without any treatment. She presented just for postmenopausal bleeding but found cyst of vagina under vaginal ultrasound incidentally. The pelvic magnetic resonance image showed small patchy abnormal signal and enhancement in S4 and S5 vertebrae whose nature was unsure. Combined with her medical history, accessory examination and intraoperative findings, there was little possibility that the tumor originated from sacral. We didn’t find any abnormal sacral condition during the surgery, and there was still a little space between the mass and sacral. The mass also had clear limitation to other tissue in pelvic. All of them above support the view that this chordoma was not from sacral. Because the mass had no space to the vagina and the vaginal wall clinging to the mass could not be moved, we speculated that the vaginal wall was invaded by the tumor. Patients suffer from malignant vaginal tumor usually presented with colporrhagia and vaginal mass, which was similar to a malignant vaginal tumor. So we prefer to diagnose it as vaginal chordoma. Though rare, this case gives us some cogitation about the differential diagnosis of for complex vaginal lesions and we may deepen the understanding of vaginal tumor. Since no cases about vaginal chordoma were reported before, and vaginal chordoma was not included in WHO classification of tumors of female reproductive organs in 2014, whether this case was exact vaginal chordoma or not need further study and more cases to support.

Computed tomography could show the scope and internal structure of chordoma. CT is good at visualize bonylysis and intratumoral calcification. Chordoma of sacral, skull base, spine are often present as expansive bone destruction with clear boundary and lobulated structure in which punctiform or schistose residual bone and dot-like calcification can be found. MRI is considered to be the gold standard in pretreatment and post- treatment evaluation of chordomas. It demonstrates the entirety of the chordoma and its relationship to vital structures [5]. On T1-weighted MR images, chordomas demonstrate intermediate to low signal intensity; On T2-weighted MR images, they demonstrate high signal intensity. Both T1-weighted image and T2-weighted image are high signal when tumor hemorrhage happens, and low signal when tumor calcified [6].

The diagnosis of vaginal tumor should be confirmed by histopathology. In this report, we did ultrasound-guided biopsy at first but it did not show abnormal change. The reason may be that the puncture position is not exact or the needle biopsy tissue was so little. But the following vaginal mass biopsy and postoperative histopathological examination with immunohistochemistry confirmed this to be a vaginal chordoma. The gross morphologic of chordoma often present to be round mass with lobulated

---

**Figure 4.** A-C. Hematoxylin and eosin section shows that the lobule is consist of myxoid stroma and physaliferous cells, the typical cells of chordoma filled with abundant mucoprotein, floating in the pale blue mucoid stroma background. D. The expression of S-100 on vaginal mass was detected by immunohistochemical staining.

---
Chordoma of vagina

structure separated by fibrous band. It was soft and developed expansively. The cut surface of this tumor was canous to blue and white color in which focal hemorrhages and cystic zone could be seen. The tumor mass was well-circumscribed with pseudocapsule [5-7]. The tissue in lobular arrangement was separated by fibrous connective tissue at low power. The lobule consisted of classic physaliferous cells suspended in myxoid stroma, the typical cells of chordoma were filled with abundant mucoprotein. The tumor cells were arranged in cords, nests and sheets [8, 9]. Under the electron microscope, specific mitochondria-endoplasmic reticulum complex and desmosome junction could be seen, which indicates that the cells to are epithelioid. The immunohistochemical examination of chordoma always reveals S-100, Vimentin, CK, EMA positive [8-10]. Recent studies show that brachyury is a maker that can be used to distinguishes chordoma from some other tumors like parachordoma [10]. Chordoma cells increase slowly and often invade surrounding tissues. The metastatic incidence is reported to be 19%-37% [11, 12]. Common metastatic sites are pulmonary, liver, bone, lymphatic node, stomach etc. The cases of metastases in central nervous system, heart, skin, ovary, muscle also have been reported [4, 7, 12-14]. Hematogenous metastasis is the main metastasis route followed by lymphatic metastasis [1, 7].

Standardized treatment for vaginal chordoma is not clearly defined because neither primary nor metastatic vaginal chordoma has been reported up to now. As for the treatment of vaginal chordoma in our hospital, we referenced the treatment of malignant vaginal tumor and chordoma located in other sites. We suggest the patient receive postoperative adjuvant radiotherapy in other hospitals for we don’t have radiotherapy equipment, but she didn’t take advice for some personal reasons. The therapy of primary vaginal carcinoma is acknowledged to be surgery and radiotherapy or combined with chemotherapy where appropriate. Whereas the standard treatment of primary bone and skull base chordoma is surgery. Complete surgical resection with wide, negative surgical margin is of great importance to reduce local recurrence, prolong the disease-free interval and increase tumor-free survival [1, 5, 15-21]. Chordoma has been thought to have a poor radio and chemotherapy sensitivity. But more investigators hold that surgery combined with intensity-modulated radiation therapy, proton beam radiation, heavy ion radiotherapy, carbon ions radiotherapy, and stereotactic body radiation therapy is effective in increasing 5-year survival rate and total survival rate, prolonging the disease-free interval and decreasing the local recurrence rate, especially for patients who get incomplete resection [15, 19, 22-28]. Some studies also show superior local control rate using carbon ion re-irradiation and proton therapy in recurrent skull base chordoma [29, 30]. Unified chemotherapy of chordoma is not acknowledged according to existing literature and chemotherapy is often regarded as supportive treatment in an advanced stage. Some research has shown that Gleevec-based chemotherapy could slow down tumor growth which may be an effective chemotherapeutic drug under further research [31]. The study from Huang D and Guan J Y indicated that percutaneous intra-tumoral injection with pingyangmycin lipiodol emulsion is effective to control the growth of recurrent sacrococcygeal chordomas and relieve pain caused by the tumor. It may be an effective method to treat recurrent chordoma in the future [32, 33].

Chordoma is easy to relapse for due to its local aggressiveness and difficulty of radical resection. The overall median survival is 6.29 years [1]. Five-year overall survival was 60.0%. Overall survival correlates significantly with treatment modality, with 44% surviving at 5 years with no treatment, 52% with radiation alone, 82% surgery alone, and 78% surgery and radiation [15]. Patients from zero to nineteen years old and more than seventy years old age groups have the poorest survival rate. Hispanic and high socioeconomic status people’s survival was higher than others [34]. It is reported that the 5-year recurrence-free survival rate is 59% and 46% at 10 years [16]. Patients may have poor prognosis after relapse and metastasis. In our case, the patient got a complete resection of chordoma mass and her prognosis depends on further long-term follow-up.

Acknowledgements

We acknowledge the patients providing informed consent for the publication of their medical details. The authors would like to thank Dr. Zheng’ai Xiong (Department of Obstetrics
and Gynecology, The Second Affiliated Hospital of Chongqing Medical University), Dr. Jia Li (Department of Pathology, The Second Affiliated Hospital of Chongqing Medical University) for their assistance.

Disclosure of conflict of interest

None.

Address correspondence to: Lina Hu, Chief of The Obstetrics and Gynecology Department, The Second Affiliated Hospital of Chongqing Medical University, 76 Linjiang Road, Chongqing 400010, P.R. China.
E-mail: cqhulina@126.com

References

[22] Yasuda M, Bresson D, Chibbaro S, Cornelius JF, Polivka M, Feuvret L, Takayasu M, George B. Chordomas of the skull base and cervical...
Chordoma of vagina


