Perianal atypical leiomyoma
A case report

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1. Introduction

Atypical leiomyoma also known as singular leiomyoma or pleomorphic leiomyoma is more commonly found in the uterus and esophagus; its presentation as perianal disease is relatively rare. As this tumor lacks specific clinical manifestations, the rate of clinically correct diagnosis is low and it is often missed or misdiagnosed; the incidence and malignancy rates increase gradually with age.[1]

2. Case presentation

A 28-year-old female, G1P1, was hospitalized in The First Affiliated Hospital of Guangxi Medical University with a perianal mass found more than 4 years ago. The patient denied contraceptive use, injury, obvious pain, chills, or fever, and change in urine or stool. At the local hospital in 2013, a perianal incision and drainage procedure was performed on the diagnosed perianal abscess (the specific process was unknown). The postoperative incision had healed and the mass had decreased in size. The mass increased during a 2015 pregnancy. Postpartum the patient used no contraceptives, had no injury, pain or other symptoms; the untreated mass was smaller after childbirth. With the mass now larger than that in 2015, the patient presented to our hospital in 2017 for treatment. The patient had a history of broad bean disease and a past caesarean section. The patient had no family history of genetic atypical leiomyoma. The diagnosis was a perianal subcutaneous mass (nature to be investigated). The patient was signed share her perspective or experience consent for treatment and surgery, and since this article belongs to the clinical retrospective study, an ethical approval was not required.

2.1. Physical examination

The 5 cm × 4 cm × 4 cm sized mass was located on the left side of the anus and vagina; it was firm upon palpation, had distinct boundaries, was approximately 1.5 cm from the edge of the anus and vagina, was fixed and nontender. On rectal examination, the rectal mucosa was smooth and, similar to the mass in the intestinal wall, was extremely difficult to palpate the pole.

The magnetic resonance imaging (MRI) scan revealed: (1) an occupying lesion in the left perineum; the specific process was unknown. A 4.1 cm × 5.2 cm × 4.9 cm sized round mass was observed on the left side of the circumference (unclear if this was a fibrous tumor, tendon sheath giant cell tumor, or ossifying myositis); lesions and the vaginal wall on the left side of the left pubic rectal muscle boundary were unclear; the urethral and corpora cavernosum clitoridis boundary was clear. (2) There was a small amount of fluid in the pelvic area (Fig. 1).
In order to alleviate the patient’s symptoms, an anal peripheral mass resection was performed under lumbar anesthesia on January 12, 2017.

2.2. Surgical procedure

On intraoperative exploration, a 7 cm × 4 cm × 4 cm sized round mass was observed on the left side of the circumference; it was firm upon palpation, was adhered to the capsule, and the anal canal sphincter had a tight adhesion (Fig. 2). The perineum left approach: an approximately 6-cm incision was made along the outside of the capsule, followed by the separation of subcutaneous tissue, partial ligation of the sphincter, and then an approximately 1-cm incision was made partially through the rectal wall, followed by complete removal of the mass. This was followed by full hemostasis, 3–0 absorbable suture line repair of the rectal perforation, placement of a negative pressure drainage tube, and then layer-by-layer suture repair of the incision.
2.3. Pathological examination

To the naked eye, the mass size was approximately 7 cm × 4 cm × 3 cm in size and appeared gray, smooth, firm, with an intact capsule. No tumor necrosis or bleeding occurred. All the mass were taken out, and 1 piece was taken every 1 cm. After the materials were drawn, the tissues were fixed, dehydrated, soaked in wax, embedded, and stained with HE. (B) Atypical leiomyoma with normal leiomyoma in the presence of a bizarre nucleus (as indicated by the arrow). HE, medium magnification. (C) The arrow shows bizarre nuclei, HE, high magnification. (D) Tumor cells express SMA, EnVision, medium magnification. (E) Tumor cells express Desmin, EnVision, and medium magnification. (F) Tumor cells express Calponin, EnVision, and medium magnification. (G) Tumor cell Ki67 positive index 2%, EnVision method, medium magnification. (H) Tumor cells did not express CD117, EnVision method, medium magnification. HE=hematoxylin and eosin staining.

Figure 3. Pathology of perianal atypical leiomyoma. (A) Surgical resection of pathological specimens. All the mass were taken out, and 1 piece was taken every 1 cm. After the materials were drawn, the tissues were fixed, dehydrated, soaked in wax, embedded, and stained with Hematoxylin and eosin staining. (B) Atypical leiomyoma with normal leiomyoma in the presence of a bizarre nucleus (indicated by the arrow). HE, medium magnification. (C) The arrow shows bizarre nuclei, HE, high magnification. (D) Tumor cells express SMA, EnVision, medium magnification. (E) Tumor cells express Desmin, EnVision, and medium magnification. (F) Tumor cells express Calponin, EnVision, and medium magnification. (G) Tumor cell Ki67 positive index 2%, EnVision method, medium magnification. (H) Tumor cells did not express CD117, EnVision method, medium magnification. HE=hematoxylin and eosin staining.

3. Discussion

Perianal mass more common to benign tumors such as epidermoid cysts, sebaceous glands, anal canal papilloma, and so on; benign lesions are common, less malignant.

Bell et al[2] studied 43 cases of uterine singular leiomyoma. After more than 2 years of follow-up, with only 1 case (2%) identified as a clinical malignancy, and they concluded that such tumors should be named “recurrence of atypical leiomyoma.”
Academician Liu Tonghua studied the 42 cases of atypical leiomyoma with complete follow-up data from Peking Union Medical College Hospital and reclassified them according to Bell’s standard. The results showed that 28 cases were common type leiomyomas and 14 cases were leiomyosarcomas. In the 2 studied groups, the main difference was in the number of nuclear divisions in the tumor cells.[3] Zhuang et al[4] have reported 2 cases of surgically resected perianal leiomyoma. Postoperatively the pathological diagnosis of leiomyoma was confirmed; the patient was discharged after surgery, and no recurrence was observed during 3 months of follow-up.

Perianal atypical leiomyoma should be considered when a perianal abscess is present. Perianal abscess incidence is characterized by acute onset, severe pain; it is often accompanied by chills and fever, and after rupture, formation of an anal fistula is observed. Perianal atypical leiomyoma is round, has a smooth mucosa, is firm upon palpation, and generally lacks symptoms of infection. Perianal atypical leiomyoma may be related to genetic predisposition and can occur at any age. The clinical manifestations of atypical leiomyoma and the general appearance are undistinguishable from ordinary leiomyoma without microscopic examination. The pathology of this case also suggested that atypical leiomyoma is scattered and patchy distribution of exotic nuclei cell common leiomyoma cells in the background, the cell morphology of odd shape, singular nuclear pleomorphism, nuclear large and hyperchromatic, with multinucleated giant cells, but few mitotic figures, 0 to 1/10HPF. If there is a pregnancy or the administration of large doses of progesterone, the fibroids can appear similar to the singular cells. These singular tumor cells are usually focal in the fibroids, often near the degenerative zone, but sometimes most of the singular cells are diffused in the majority of fibroids. However, rarely is the entire fibroid constituted of such cells. In the case of this diagnosis of atypical leiomyoma, caution is indicated. To distinguish this diagnosis from leiomyosarcoma, leiomyosarcoma of the tumor cell atypia is obvious, nuclear fission is easy to see, generally >10/10HPF, common tumor necrosis and bleeding. It is necessary to observe carefully and draw more specimens for exclusion.

In our case, the patient had a history of perianal surgery 2 years prior to presenting at our hospital with a relapse. The MRI indicated a tumor; thus, we decided to perform surgery and confirm the diagnosis by immunohistochemistry. When a local resection of an anal fissure leiomyoma is feasible to prevent fecal incontinence, particular attention should be paid to avoid injury to the anal sphincter.[5] If during the resection there is sphincter injury, the repair should be timely. In cases with tumor and tight anorectal sphincter adhesions, to ensure complete resection of the tumor outside the capsule, partial ligation of the external sphincter and puborectalis muscle, followed by postoperative repair of the anal pubis rectal muscle and external sphincter, should be performed to promote postoperative recovery of anal function.

Perianal atypical leiomyomas are benign tumors, but with the clinically atypical leiomyoma, it is sometimes difficult to distinguish between potential malignant smooth muscle tumors, and there may be malignant changes. Surgery should ensure complete resection, and to avoid postoperative recurrence, there should be a regular follow-up.

After a thorough search of the literature, we found that leiomyomas are more common in the uterus and esophagus, and perianal leiomyomas are rare. It should be considered that perianal leiomyomas must be confirmed by immunohistochemistry.

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References
[1] Alonso GJ, Membrives OA, Martínez CD, et al. Real anal leiomyoma: a case report. J Gastrointest Cancer 2011;42:54–6.
[2] Bell SW, Kempson RL, Hendrickson MR. Problematic uterine smooth muscle neoplasms. A clinicopathologic study of 213 cases. Am J Surg Pathol 1994;18:535–58.
[3] Liu T. Comments on a better understanding of smooth muscle neoplasms in the uterus. Zhonghua Bing Li Xue Za Zhi 1996;25:259–62.
[4] Zhuang JB, Zhu HB, Huang XF. Perianal smooth muscle tumors: two case report. Clin Educ General Pract 2013;6:698–9.
[5] Ruiz SS, Beltrán JG, Parés D. Perianal leiomyoma. Rev Esp Enferm Dig 2009;101:209–11.