



## Transperineal excision of a sizeable angiofibroma of the ischiorectal fossa. A case report



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### ABSTRACT

Cellular angiofibroma is a recently described rare benign soft-tissue tumor more commonly presenting in middle-aged women, often mimicking malignancy. The vulva is most common location. Complete local excision is the best curative treatment and usually there is no recurrence after surgery. We describe a 49-year-old woman with a painless tumor in the left ischiorectal fossa. It was a random finding in a routine computed tomography (CT) scan after resection of ear melanoma 3 years previously. Ultrasonography showed a solid mass, and further magnetic resonance imaging (MRI) suggested a rhabdomyosarcoma. Altogether, these findings indicated malignant disease. An uncomplicated simple excision of the tumor was done in the operating theatre. The mass measured 7×5×5 cm and the histopathological examination found that it was a cellular angiofibroma, a benign lesion. There were no postoperative complications. This case report highlights the need for multidisciplinary team management of rare tumors such as cellular angiofibromas.

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## 1. Introduction

Cellular angiofibroma is a rare benign mesenchymal soft-tissue tumor. It was first described in 1997. Macroscopically these tumors are usually well circumscribed and are characterized by two main components: bland spindle cells and small to medium-sized vessels with mural hyalinization [1].

The Bartholin's cysts are the most common vulval lesions. Other differential diagnoses apart from an angiofibroma include aggressive angiomyxoma, angiomyofibroblastoma, spindle cell lipoma, solitary fibrous tumor, perineurioma, and leiomyoma [2].

Genital angiofibromas occur mainly in middle-aged women (39–50 years), are less than 3 cm in size, and are localized in the vulva [3]. Currently there are only two case reports of an angiofibroma of the perineum in the literature [4].

We report the case of a woman with a large angiofibroma of the ischiorectal fossa that was excised transperineally.

## 2. Case Presentation

### 2.1. Clinical Presentation

A 49-year-old, Caucasian postmenopausal parous woman, with a history of ear lobe melanoma three years previously, was diagnosed with a perineal mass compatible with potential Bartholin's cyst on her annual computed tomography (CT) imaging procedure [Fig. 1a].

There was no family history of gynecological or breast cancer.

### 2.2. Clinical Findings

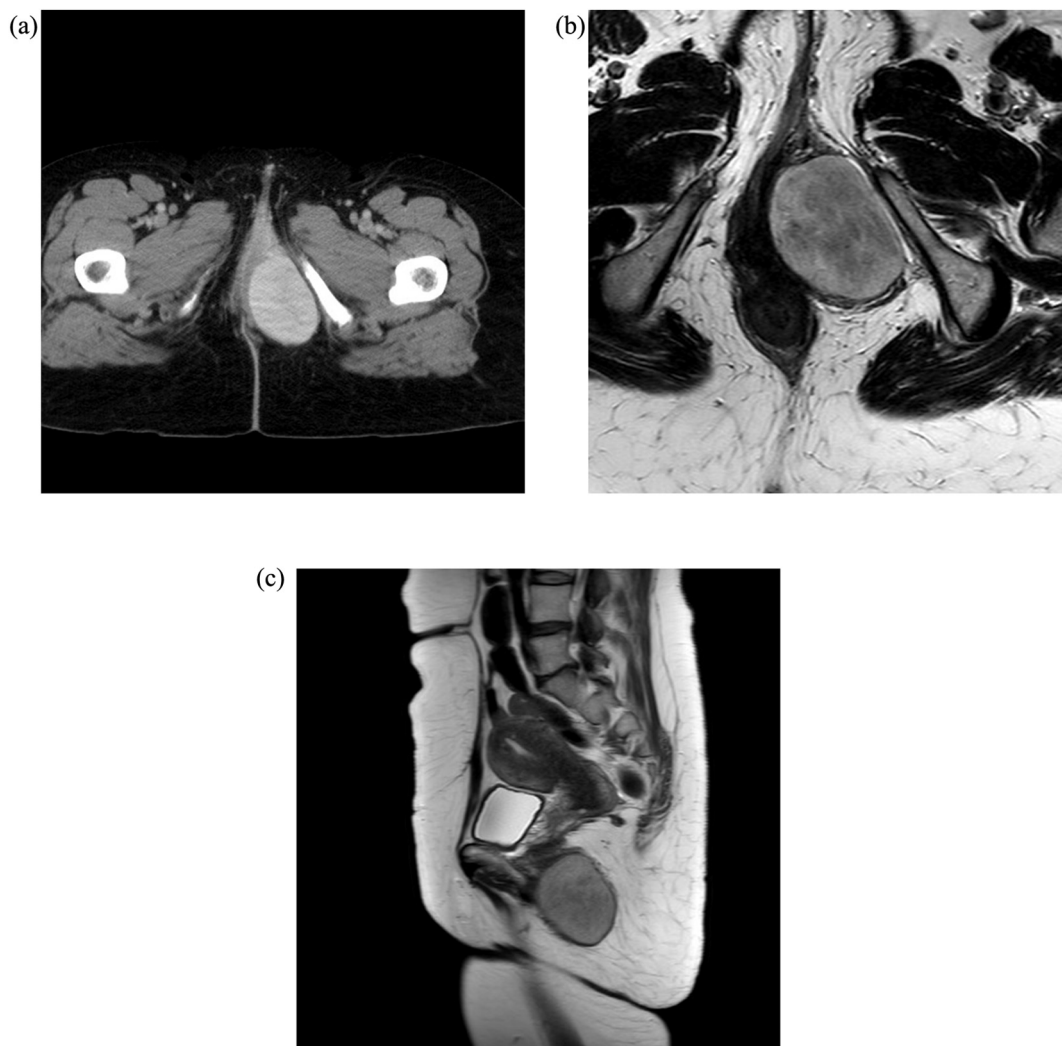
Gynecological examination revealed a large (5 cm in diameter), asymptomatic, left ischiorectal fossa lump and no lymph nodes were palpated on groin examination. As ultrasound examination found that the mass appeared to have solid characteristics, the patient underwent magnetic resonance imaging (MRI) of the lower abdomen [Figs. 1b and c and 2a]. The initial diagnosis of benign Bartholin's cyst was revised, and the differential diagnosis included a rhabdomyosarcoma, a lipoma, a perineal metastasis of the melanoma, or an ectopic fibroma.

### 2.3. Timeline

#### 2.3.1. Diagnostic Focus and Assessment

The MRI scan revealed a well circumscribed mesenchymal tumor (6.7 × 5.0 × 6.0 cm) deriving from the levator ani or the musculus

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**Fig. 1.** (a) CT scan transverse image of the mass. (b) MRI transverse image of the mass. (c) MRI sagittal image of the mass.

sphincter ani externus, with no abnormal lymph nodes in the pelvis, suggesting rhabdomyosarcoma as the primary diagnosis. All gynecologic neoplastic tumor markers were negative. After multidisciplinary evaluation with oncologists and general surgeons, the patient was scheduled for a full excision of the tumor for both therapeutic and diagnostic purposes.

### 2.3.2. Therapeutic Focus and Assessment

The surgical team comprised a gynecologist and a colorectal surgeon. The initial plan was for transperineal surgical exploration and if the lesion was inaccessible via the perineal route, convert to laparoscopy/laparotomy in order to complete a clear-margin surgical excision and lymph node biopsy. In the operating theatre, the patient was placed in the lithotomy position under regional anesthesia and a left paravaginal incision was made 2 cm laterally to the left labia majora (Fig. 2a, b). The mass was resected *en bloque* with no complications and minimal blood loss (Fig. 2c–f). Surgical time was 40 min. The tumor was located in the left ischioanal fossa and was in direct contact with the levator ani, the anus and the left vaginal wall. It measured 7×5×5 cm, weighed 118 g, was pale white, and was surrounded by a rim of normal tissue (Fig. 2f). Initial pathological examination showed the lesion to be benign. The patient was discharged after 48 h with no postoperative complications.

The lesion was well circumscribed, there was no capsule, and it was surrounded by a layer of adipose, connective, and smooth muscle tissue. Histopathology is shown in Fig. 3a, b and c. The spindle-shaped cells were positive for Vimentin, estrogen and progesterone receptors. Stains for CD10, SMA, S-100 protein, c-kit, CD34, Desmin and HMB45 were all negative. Ki-67 was between 1% and 3%. The final diagnosis was cellular angiofibroma.

### 2.3.3. Follow-Up and Outcomes

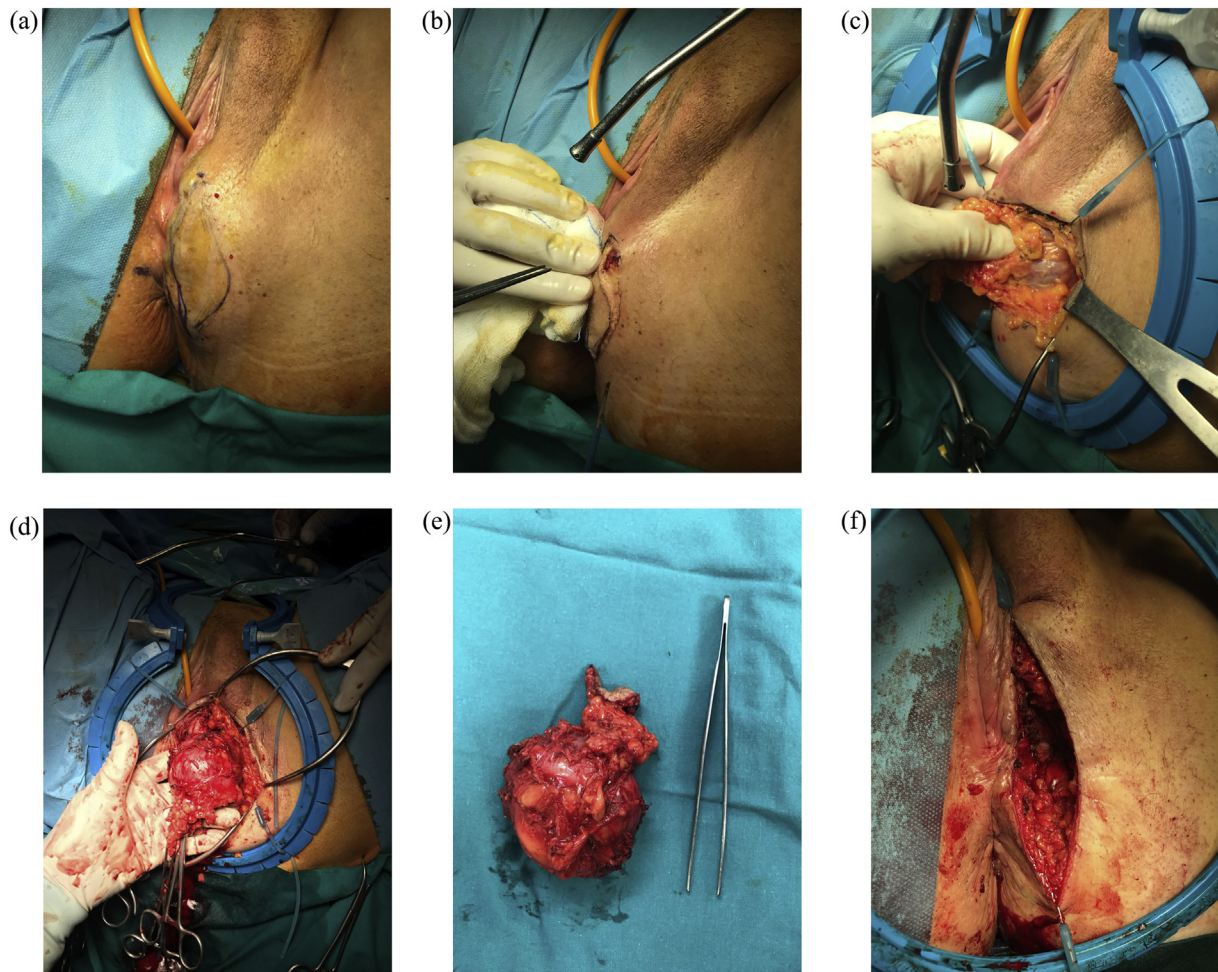
There were no postoperative complications. Eighteen months after surgery an ultrasound scan and a speculum vaginal examination showed no evidence of tumor recurrence.

## 3. Discussion

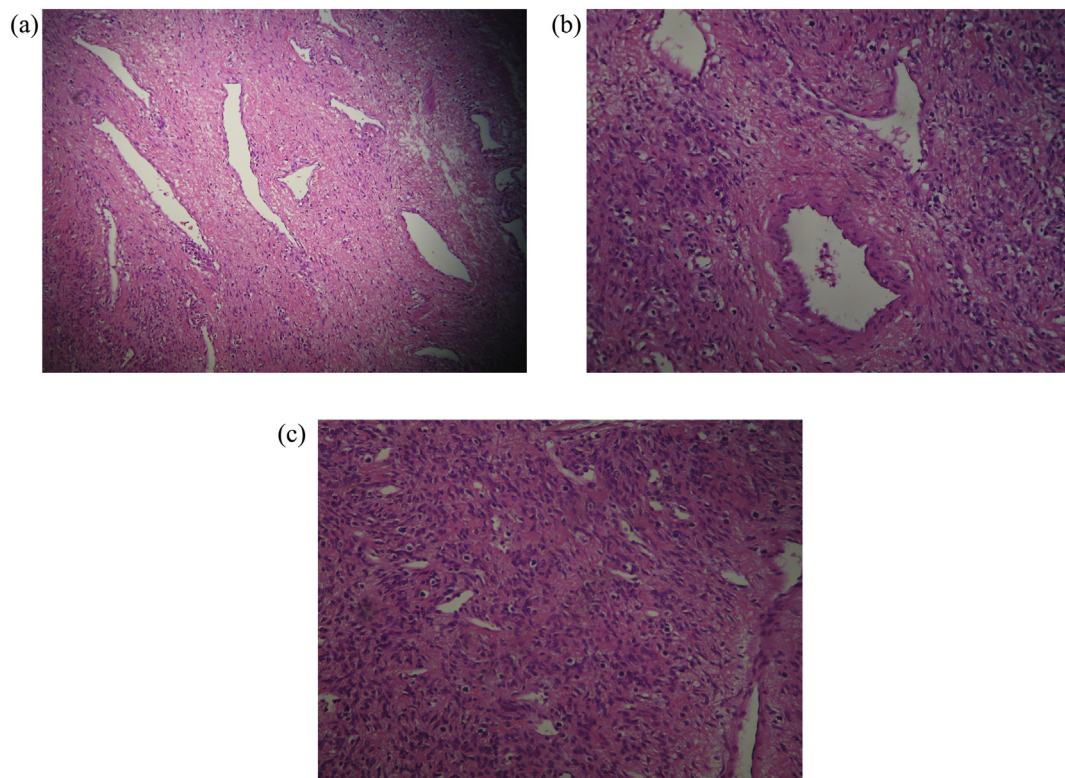
Soft-tissue angiofibroma is a benign, fibrovascular soft-tissue tumor with a specific cytogenetic translocation [5]. It is more common in males, but in women the most frequent location is the vulva, and the average size is less than 3 cm [6]. In the reported case, the location (left ischioanal fossa) and the size (7×5×5 cm) led to a high initial suspicion of malignancy.

Malignant change in angiofibromas (atypia, sarcomatous) is reported to be low. There is a study of 13 cases with atypia or sarcomatous





**Fig. 2.** (a) The perineal mass protrudes like a lump in the genital area. (b) Initial sphenoidal left paravaginal incision. (c) Initial dissection through the left ischiorectal fossa. (d) The mass has been mobilized from the surrounding tissues. (e) *En bloqué* excision of the 7×5×5 cm tumor. (f) The surgical trauma before closure.



**Fig. 3.** Histopathology of the angiofibroma: (a) fusiform cells, blood vessels and adipose tissue (hematoxylin-eosin; 100 X); (b) small to medium-size vessels with hyalinized walls, showing fusiform cells with bland nuclei and clear (hematoxylin-eosin; 400 X); (c) bland spindle cells with uniform nuclei and pale indistinct cytoplasm (hematoxylin-eosin; 400 X).

transformation among 154 typical cellular angiofibromas in both males and females [7]. The biologic significance of malignant transformation remains uncertain [7].

Both the radiologic and pathologic differential diagnoses for this lesion include benign and malignant lesions. Thus, detailed histopathological assessment is essential.

Treatment is surgical, removing the whole tumor, preferably with free surgical margins. If the mass is near or involves the anus, general surgeon support is needed. The most common approach for lesions located in the ischioanal fossa is via the transperineal route. However, depending on the location and tumor size, transabdominal, laparoscopic, and vaginal access offer alternatives. In this case, the surgical approach was pre-operatively unpredictable because (a) of the size of the lesion, and (b) of the possibility of being rigidly adherent to the surrounding tissues, a finding suggesting malignancy and necessitating clear margins for excision as well as pelvic/inguinal lymph node biopsies. Thus, the operation started with a transperineal incision, tumor dissection preserved the integrity of the rectum, and the procedure was completed successfully with a minimally invasive approach only after confirming the benign identity of the lesion intra-operatively with a pathologist. A major complication of this procedure could be the development of rectovaginal or vesicovaginal fistulae, because of the proximity of the lesion to the rectum or the urinary bladder. Meticulous dissection of the lesion and the joint approach skills of a colorectal and gynecological surgeon reduced this risk.

Usually, this tumor does not recur, although there was one report of recurrence in a 49-year-old woman who had a simple excision of a 4 cm tumor and she subsequently developed a recurrent swelling at the site of the previous excision [7,8]. The woman described in this case report had had no recurrence after 18 months.

#### 4. Conclusion

Angiofibroma of the perineum can be excised transperineally, even if it is unusually large. An individualized approach, a meticulous diagnostic set-up, and the presence of a multidisciplinary team are important for the management of complex cases such as the present one.

#### Contributors

Themistoklis Mikos was responsible for project development, clinical follow-up, data collection, and manuscript writing.

Iakovos Theodoulidis contributed to data acquisition and manuscript writing.

Kalliopi Dampala contributed to data analysis and interpretation, and revising the article.

Costas P Spanos contributed to data acquisition and analysis, and drafting the article.

Sofia Tsiapakidou contributed to data acquisition and interpretation, and approval of the final version.

Grigoris F Grimbizis contributed to manuscript writing/editing and data analysis.

#### Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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#### Patient Consent

Obtained.

#### Provenance and Peer Review

This case report was peer reviewed.

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