Case Report

Perianal Angioleiomyoma: A Case Report and Review of the Literature

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Abstract

Angioleiomyomas are rare benign tumors, which take origin from smooth muscle fibers of the tunica media of veins. Even though angioleiomyomas can appear anywhere in the body, these masses are rarely occurred in the gastrointestinal system. This is the first reported case of perianal angioleiomyomas, where the tumor in close relation with the anal canal was investigated with endoanal ultrasonography. Local excision of such lesion is generally curative.

Keywords: Angioleiomyoma, anus, endoanal ultrasonography, perianal, vascular leiomyoma

Introduction

Angioleiomyomas are rare benign tumors, which take origin from of smooth muscle fibers.[1] Angiomyoma, dermal angioma, or vascular leiomyoma are various names of angioleiomyomas which are vascular variants of leiomyomas.[2] The tunica media of veins is the histological location where these rare, benign tumors take origin. Even though angioleiomyomas can appear anywhere in the body, these masses are rarely occurred in the gastrointestinal system.

Regarding the histological division, angioleiomyomas can be classified into four types: (1) capillary/solid, which is the most common type and is characterized by the presence of smooth muscle fibers that surround slit-like vascular channels (preponderance in females); (2) venous, which is characterized by abundant vascular small channels with thickened muscular walls (preponderance in males); (3) cavernous, with a thin layer of smooth muscle cells that enclose wide vascular small channels (preponderance in males); and (4) a type of angioleiomyomas with combined characteristics between capillary and venous.[1,3]

We are presenting a rare case of perianal angioleiomyomas, the first reported in the English literature, where the tumor is in close relation with the anal canal and it is the first reported case, which was investigated with endoanal ultrasonography (EAUS).

Case Report

A 71-year-old woman presented to our proctology practice, complaining of a palpable, painless, perianal lump and a pressure sensation at the perineum. The patient referred that she began noticing a small lump 2 years earlier, located at the anterior part of the anus and posteriorly to the vagina [Figure 1]. In the past 10 months, the lump has grown up and has become firm. During the examination, the lump was presented hard at palpation, painless, and without fluctuation. Bilateral inguinal lymph nodes were not enlarged. In addition, at anoscopy of the anal canal, there was not noticed any inflammation of the anal mucosa. Both EAUS and transanal ultrasonography were also performed, showing a mass with a visible capsule and lobular parenchymal structure, in close relation with the anal canal [Figures 2 and 3]. The tumor appeared well circumscribed and well defined with dimensions of 30.8 mm × 28.1 mm × 18.8 mm. Fine-needle

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aspiration biopsy was inconclusive to permit a definite diagnosis. Magnetic resonance imaging (MRI) (T1-weighted magnetic resonance [MR]) presented mixed areas (isointense or hyperintense to skeletal muscle) [Figure 4a and b], whereas in T2-weighted MR, the lesion had a heterogeneous appearance and was slightly hyperintense to skeletal muscle [Figure 4c]. In addition, in T2-weighted MR images, there was a peripheral rim with low signal intensity, which corresponded to a fibrous pseudocapsule of the lesion.

Local excision was performed, preserving a great part of both the internal and the external sphincter. The postoperative course was uneventful and the patient was discharged from the hospital on the 2nd postoperative day. Three months after the operation, during the follow-up visit, the patient did not show any incontinence or stenosis. Furthermore, the patient did not show any signs of the recurrence of the disease 1 year after the operation.

Histopathologically, gross examination of the surgical specimen revealed a submucosal sharply demarcated grey-tan tumor, which at the microscopic level showed numerous slit-like, thick walled and dilated – Frequently with "staghorn" configuration – vascular channels with intervening bundles of spindle cells with minimal nuclear pleomorphism and the absence of mitoses [Figure 5]. Results of immunohistochemical analyzes revealed that the spindle cells were negative for C-kit, estrogen receptor, and CD34 and positive for smooth muscle actin (SMA) and desmin [Figure 6]. The findings were compatible with angioleiomyoma, mainly of cavernous and a smaller proportion of solid (capillary) and venous type.

Figure 1: Clinical appearance of perianal angioleiomyoma (arrows)

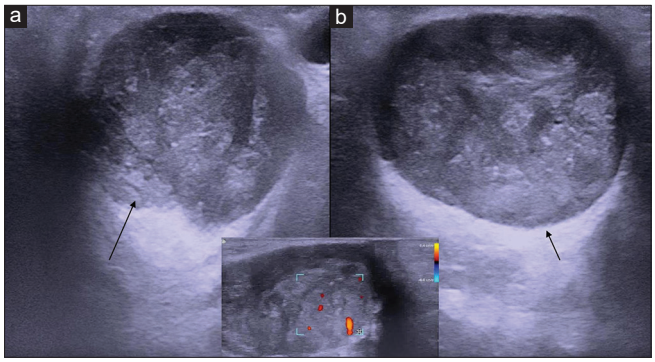

Figure 3: Perianal angioleiomyoma viewed with linear ultrasound probe in (a) Vertical (arrow), (b) Transverse sections (arrow) (presence of heterogeneous echotexture combined with posterior acoustic enhancement) and (c) Color Doppler examination of the lesion (vascular flow signal on color Doppler) (insert)

Discussion

Angioleiomyomas are benign soft tissue tumors, which derive from tunica media of small blood vessels, and they are composed of smooth muscle cells.[4] Angioleiomyomas are usually solitary lesions, with a female preponderance, and a major incidence between the third and sixth decades of life.[2] These benign tumors represent 5% of all benign lesions of soft tissues.[5] Another interesting characteristic is that the preoperative diagnosis of angioleiomyomas rarely occurs. Moreover, these benign neoplasias are often presented on the skin of lower extremities,[6] while less common localizations are hands, female reproductive organs, scrotum, mouth cavity, kidneys, and rectum.[3,7-11] Regarding

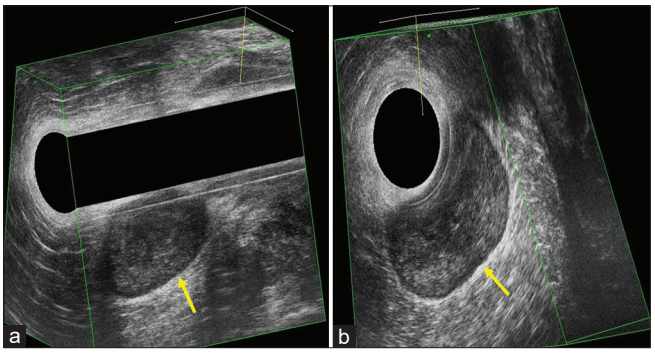

Figure 2: The tumor was presented as circumscribed lesion with homogeneous structure at EAUS (a and b) (marked by arrow). EAUS: Endoanal ultrasonography

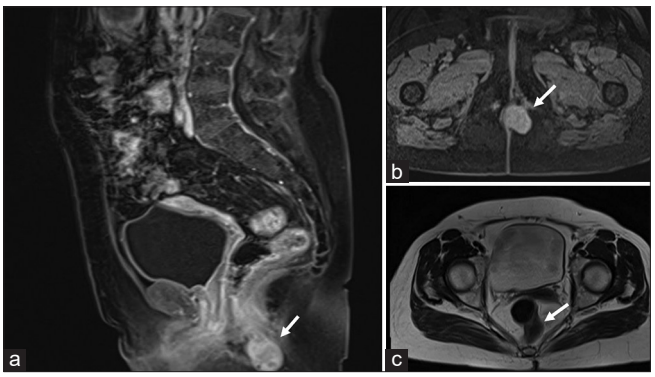

Figure 4: MR images (T1-weighted MR) of perianal angioleiomyoma, (a) Sagittal (T1-weighted fat suppressed axial section image with contrast enhancement) (marked by arrow), (b) Transverse section (T1-weighted fat suppressed axial section image without contrast enhancement) (marked by arrow) and (c) Transverse section (T2-weighted MR with heterogeneous and slightly hyperintense to muscle characteristics) (marked by arrow). MR: Magnetic resonance

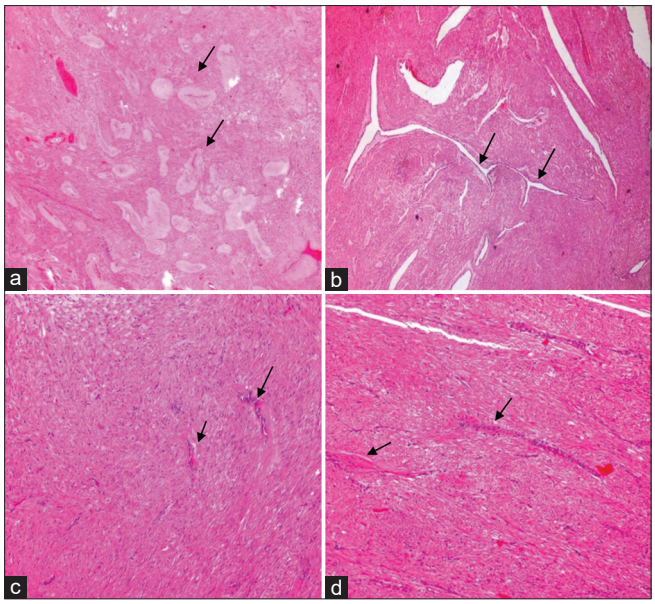

Figure 5: Hematoxylin-eosin stain of different sites of the tumor where there are smooth muscle fascicles and (a) Thick-walled vascular channels (×40) (arrows), (b) Dilated "stag-horn" vascular pattern (×40) (arrows), (c and d) Slit-like vascular channels (×100) (arrows)

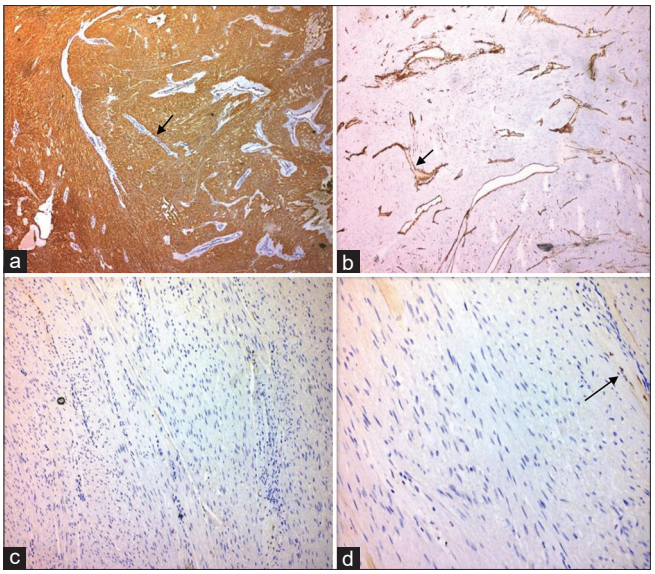

Figure 6: (a) SMA positive smooth muscle bunds (×40) (arrow), (b) CD34-positive vascular channels and negative tumor cells (×40) (arrow), (c) ER-negative tumor cells proving no Mullerian derivation (×100) (d) Ki67 generally <1% (×200) (arrow). ER: Estrogen receptor, SMA: Smooth muscle actin

the treatment of angioleiomyomas, local excision of the lesion and ligation of feeder vessels are the most common ways of treatment.[2] Nonetheless, our presented case appeared as a painless lump, and pain is the most referred clinical symptom of angioleiomyomas.[6] Cold and intense physical activity may be the cause of the dynamic contraction of smooth muscle and, as a result, the provocation of pain.[12] In the literature, another reported symptom is the swelling of lesion, especially after physical activity or long standing on feet. This symptom could be explained due to the accumulation of blood in the vascular channels of the tumor.

As far as the pathogenetic origin of angioleiomyomas, there is still a great deal of controversy. Among the proposed estiological causes of angioleiomyomas are venous stasis, local minor trauma, as well as estrogen variations.[13-15] Some authors have proposed that the proliferation of smooth muscle cells within a hemangioma creates an angioleiomyoma.[16] In addition, many authors agree that angioleiomyoma derives from veins, so there is a hypothesis that these lesions may be hamartomas and as a consequence, a great number of angioleiomyomas are not true proper tumors but are basically a type of vascular malformations.[6,13,16]

EAUS is considered to be a reliable technique for the assessment of perianal neoplasia and valuable in distinguishing tumor-related conditions from other benign conditions that may affect the perianal area.[17,18] Nevertheless, the use of EAUS in the diagnosis of angioleiomyoma has not yet been reported. During EAUS examination, angioleiomyoma is presented as a tumor with well-defined margins and homogeneous structure, which is suggestive of the benign nature of the lesion [Figures 2 and 3]. In addition, color Doppler examination of the lesion usually presents high resistance inside the angioleiomyomas' arteries, making obvious the presence of muscular arteries.[5] Furthermore, the presence of posterior acoustic enhancement of the mass as well as the absence of any septations inside the tumor, are two additional ultrasonographic charateristics that may assist in the identification of angioleiomyoma and its differential diagnosis to other well-defined masses (such as hemangiomas and angiolipomas.).

MRI characteristics of angioleiomyomas are well identified.[19] Specifically, MRI (T2-weighted MR) presents mixed areas which were hyperintense and isointense to skeletal muscular fibers. Hyperintense areas show increased enhancement after intravenous contrast injection, while isointense areas do not show enhancement after the contrast injection. Moreover, hyperintense areas correspond to the skeletal muscle bundle cells, while isointense areas are related to either fibrous tissue or intravascular thrombi. MRI has not the ability to differentiate between different histological types of angioleiomyoma.[2]

Histological findings include mainly the presence of well-differentiated smooth muscle cells with intervening vascular channels usually delimited by a thin capsule.[6,15,20] Three major histological subcategories are solid, cavernous, and venous type.[21] In angioleiomyomas has also been reported the presence of both myxomatous and hyaline degeneration as a consequence of circulatory disturbance/ischemia.[6,20] Additional histological features of angioleiomyomas that may also be found are mature fat cells, organizing thrombus, and lymphocytic infiltrate, due to marked degenerative changes inside these benign lesions.[2] Tumor cells express SMA,

muscle-specific actin, calponin, usually desmin, and variably h-Caldesmon.[21]

Among the differential diagnosis of angioleiomyoma, there are lesions that also should be considered, such as fibromas, hemangiomas, lipomas, schwannomas, ganglions, foreign-body granuloma, glomus tumor, inclusion cyst, and pseudoaneurysm and rare type of tumors which originate from sweat glands like digital papillary adenocarcinoma.[2]

Conclusion

Angioleiomyomas are benign lesions that take origin from the smooth muscle of the tunica media. Their perianal localization is extremely rare. Preoperative diagnosis is difficult to be made. EAUS is considered to be a reliable technique that can assist in the preoperative assessment of angioleiomyomas. Local excision of such tumors generally has curative intent.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent form. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal the identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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