Vaginal angiomatosis: differential diagnosis of a rare case

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Abstract

Vaginal angiomatosis is regarded as part of a very rare entity of benign vascular tumors of the female genital tract. The incidence of these tumors is extremely low. The rarity of this disease and lack of distinctive features poses a problem of differential diagnosis. We present the case of a 51-year-old female with grade III uterine prolapse and a bleeding vaginal wall mass. Violaceous irregular soft tissue with hemorrhagic spots was observed in the lower third of the posterior vaginal wall. The patient underwent surgery for colpohysterectomy with vaginal wall mass excision. Surgical excision was curative, and no recurrences were observed after 12 months of follow-up. The aim of our study is to present a rare but representative case. This will hopefully increase the level of awareness regarding this condition so that physicans will keep it in mind during differential diagnosis of similar clinical cases. Furthermore, it highlights the important role of pathological examination for the definitive diagnosis of angiomatosis. (J Turk Ger Gynecol Assoc 2015; 16: 189-91)

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Introduction

Vaginal angiomatosis is considered as a very rare entity of benign vascular tumors of the female genital tract. The incidence of these tumors is extremely low and this percentage is further reduced if we take into account only vaginal angiomatosis. The rarity of this disease and lack of distinctive features poses a problem of differential diagnosis. Benign vascular lesions of the female genital tract are localized more frequently in the ovary followed by the vulva and more rarely in the cervix and vagina (1). Symptoms depend on the size, location, and histological type of the lesion. Small benign vascular lesions can be asymptomatic, whereas more extensive lesions may be associated with important clinical manifestations such as abnormal vaginal bleeding, infertility, spontaneous abortions, and fetal loss. Benign vascular lesions of the female genital tract can rapidly grow during pregnancy; therefore, they may cause important complications (2), and pregnant women with genital angiomatosis may require cesarean section (3). The histological type of the lesion may affect the onset of symptoms because bleeding from arteriovenous malformation can be more intense and difficult to control (4). Color Doppler ultrasound examination is often able to add useful features to diagnosis (4). Benign vascular lesions of the female genital tract may occur as isolated lesions or coexist with other hemangiomas involving the skin or other organs or can be part of a complex syndrome such as Klippel–Trénaunay–Weber syndrome (4), tuberous sclerosis, hereditary hemorrhagic telangiectasia, and blue rubber bleb nevus syndrome. Vaginal angiomas are classified on the basis of histological appearance as capillary, cavernous venous, or arteriovenous (5). The aim of our study is to present a rare but representative case. This will hopefully increase the level of awareness regarding this condition so that physicans will keep it in mind in the differential diagnosis of similar clinical cases.

Case Presentation

We present the case of a 51-year-old female with grade III uterine prolapse and a bleeding vaginal wall mass. The patient reported symptoms, such as difficulty in evacuation caused by the uterine prolapse and vaginal bleeding caused by ulcerated posterior vaginal wall mass. The patient history had no specific feature associated with angiomatosis, and all laboratory parameters are within the normal range. She had two spontaneous vaginal delivery without episiotomy or lacerations. Our patient had no history of neoplasia or other diseases, such as hypertension, chronic venous insufficiency, or acquired immune deficiency syndrome. The physical examination revealed grade III uterine prolapse with an increased volume of uterus, a hypertrophic elongated cervix, and bilateral labial hypertrophy. Violaceous irregular soft tissue is present in the lower third of vaginal wall (Figure 1). The lesion had

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a maximum longitudinal diameter of 20 mm with hemorrhagic spots in the posterior vaginal wall. Ultrasound examination confirmed the findings of the physical examination. Color Doppler ultrasound examination of the pelvic region revealed high-grade pelvic venous varicosity without arteriovenous shunts or vascular malformations. The patient underwent surgery for colpohysterectomy with vaginal wall mass excision, left salpingo-oophorectomy (because of the presence of a benign ovarian cyst), and vaginoplasty. Microscopic examination was performed on paraffin-embedded sections that were stained with hematoxylin and eosin (H&E) (Bio-Optica, Milano, Italy). The histopathological examination revealed marked acanthosis and hyperkeratosis of the cervical and vaginal epithelium that were related to uterine prolapse. In addition, histological examination of both the anterior and posterior vaginal walls demonstrated diffuse fibrosis of the sub epithelial connective tissue containing numerous vascular elements lined by flattened endothelium. These predominantly included not only venous vessels with irregularly dilated lumens with occasional herniations forming vascular lacunae but also vessels of arterial and capillary type (Figure 2). The former had thick walls, including smooth muscle cells; the latter appeared to form foci of proliferation that were clustered adjacent or in the thickness of the wall of some venous lacunae or within an organized thrombus (Figure 3). Some venous lumens demonstrated intravascular papillary proliferation of the endothelium. Endothelial cells revealed no nuclear atypia or mitotic figures. Immunohistochemistry was performed on formalin-fixed paraffin-embedded tissue block with a panel of monoclonal antibodies against different antigens (Bio-Optica, Milano, Italy). Staining for immunohistochemical endothelial markers cluster of differentiation 34 (CD34) and Wilms' tumor 1 (WT1) highlighted the increased vascular density and confirmed the endothelial nature of the almost solid clusters of capillaries. The muscular wall of the arterial vessels was positive after staining for smooth muscle actin and desmin. Staining for proliferation marker Ki67 revealed focal positivity in the basal layer of the vaginal epithelium; however, no positivity was detected in endothelial cells. These morphological and immunohistochemical characteristics enabled a diagnosis of vaginal angiomatosis. After 12 months follow-up, no relapse occurred. Our institution is a teaching hospital, and the patient signed an informed consent for the use of examination results and/or biological material for educational purposes as well as for the publication of this case report and any accompanying images.

Discussion

Angiomatosis of vagina is very rare and is often an incidental finding because of the small size of the tumors and their asymptomatic nature (6). It is difficult to clinicoradiologically differentiate angiomatosis from other neoplastic conditions. Angiomatosis should be distinguished from the other vascular tumors, particularly capillary hemangioma, cavernous venous hemangioma, arteriovenous malformation (AVM), and papillary endothelial hyperplasia (PEH) apart from bacillary angiomatosis, juvenile hemangioma, pyogenic granuloma, angiokeratoma, Kaposi's sarcoma, and angiosarcoma.

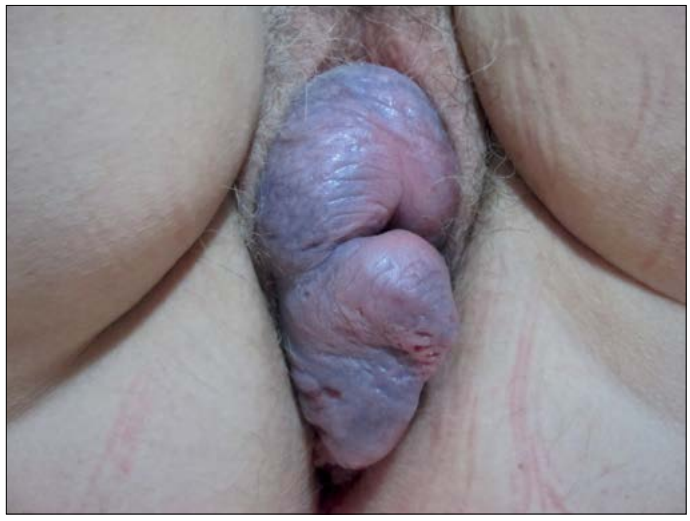

Figure 1. Violaceous irregular soft tissue with hemorrhagic spots in the lower third of the vaginal wall

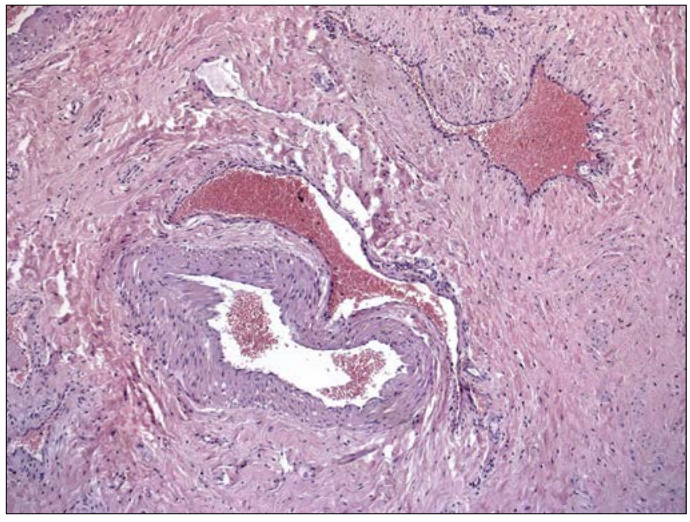

Figure 2. Arterial vessel with thick walls, including smooth muscle cells coexisting with venous vessels [hematoxylin and eosin (H&E), 5×]

PEH (Masson's tumor) occurs in the lumen of a dilated venous vessel in the vicinity of an organizing thrombus. It is probably a reactive lesion rather than a neoplastic one that incidentally arises during the organization process of older thrombi (7). In our case, histopathological exam revealed the presence of similar papillary structures in some vascular lumens; however, PEH is a focal lesion and usually does not involve arteries.

AVMs are fast-flow vascular malformations comprising a complex vessel network that directly connects feeding arteries to draining veins. The intervening normal capillary network is absent, and the microscopic appearance is characterized by the presence of both veins and arteries with dilated lumens. AVM shows similar morphological characteristics of angiomatosis; however, it is a focal but not well demarcated lesion, whereas angiomatosis occurs all over the body at multiple sites (5). Furthermore, our case demonstrated some features that were always absent in AVM-like clusters of capillaries.

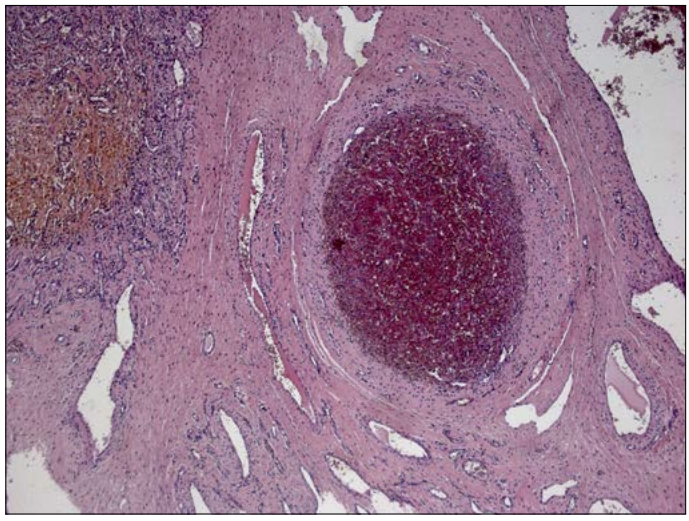

Figure 3. Proliferation of capillaries within an organized thrombus (right) and scattered in the deeper layers of the vaginal wall (left) [hematoxylin and eosin (H&E), 5×]

Non infiltrating margins of the lesion, absence of remarkable nuclear atypias, or mitosis rules the diagnosis of malignancies.
Majority of the cases of angiomatosis involving the pelvis and thigh are reported in Klippel–Trenaunay–Weber syndrome (8). This syndrome is characterized by localized angiomatosis, venous varicosities, and asymmetric osseous hypertrophy of the ipsilateral extremity (3). Our patient did not demonstrate any other systemic abnormality; therefore, she could be an even rarer presentation of angiomatosis outside Klipper–Trenaunay–Weber syndrome. A large angiomatosis could carry a high risk of intravascular coagulation and Kasabach–Merritt syndrome (9). This syndrome, usually affecting infants and also known as hemangioma with thrombocytopenia, leads to decreased platelet counts and sometimes other bleeding problems that can be life threatening. Patients uniformly demonstrate severe thrombocytopenia, low fibrinogen levels, high fibrin degradation products (due to fibrinolysis), and microangiopathic hemolysis (10). Hence, early diagnosis is required to avoid such disastrous complications.
Bacillary angiomatosis is a form of angiomatosis associated with a bacteria of the *Bartonella* genus. It can manifest in people with acquired immune deficiency syndrome and rarely appears in those who are immunocompetent. Cutaneous bacillary angiomatosis is characterized by the presence of lesions on or under the skin. Appearing in numbers from one to hundreds, these lesions may take several forms: papules or nodules that are red, globular, and non blanching with a vascular appearance; purplish nodules sufficiently similar to Kaposi's sarcoma; and purplish lichenoid plaque or subcutaneous nodule that may have ulceration similar to a bacterial abscess. Fever and bacteremia are other conditions frequently observed. In our case, features indicative of bacillary angiomatosis were absent.
In conclusion, accurate clinicoradiological examination is required to appropriately define the lesion and to detect associated congenital anomaly. Pathological examination is essential to arrive at a definite diagnosis and to exclude a possibility of malignant vascular tumor. Surgical excision is curative, and no recurrences are usually observed.

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