Review

Venous malformation in the uterus

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Abstract

Venous malformations are benign vascular lesions that rarely appear in the uterus. They are made up of abnormal veins, of different sizes and proportions, with spongiform configuration and random disposition. In the literature, some cases have been previously reported, using the term "cavernous hemangioma", but according to recent changes in terminology, approved by the International Society for the Study of Vascular Abnormalities (ISSVA), the use of this term is discouraged, and the diagnosis of Venous malformation is suggested, if the histopathological findings are met. We present the case of a 44-year-old woman, with abnormal vaginal bleeding and a clinical diagnosis of myomatosis and myoma aborted by the internal cervical orifice, in whom the histopathological study revealed the presence of a venous malformation that compromised the myometrium and endometrium, with subsequent formation of a polyp.

Key words: Vascular malformations, hemangioma cavernous; uterus; pathology.

Malformación venosa del útero

Resumen

Las malformaciones venosas son lesiones vasculares benignas infrecuentes que se presentan en el útero. Están conformadas por venas anormales, de diferentes tamaños y proporciones, con configuración espongiforme y disposición al azar. En la literatura, han sido previamente reportados algunos casos, usando el término "hemangioma cavernoso", pero según los cambios recientes en la terminología, aprobados por Sociedad Internacional para el Estudio de las Anormalidades Vasculares (ISSVA), se desaconseja el uso de este término y se sugiere el de "Malformación venosa", si se cumplen los hallazgos histopatológicos al momento de hacer el diagnóstico. Presentamos el caso de una mujer de 44 años, con cuadro de hemorragia vaginal anormal y diagnóstico clínico de miomatosis y mioma abortado por el orificio cervical interno, el estudio histopatológico reveló la presencia de una malformación venosa que comprometía el miometrio y endometrio, con formación subsecuente de un pólipo.

Palabras clave:

Malformaciones vasculares; hemangioma cavernoso, útero, patología.

Introduction

Vascular malformations are structural abnormalities of the vessels, caused by errors in vascular morphogenesis. They are classified according to the type of distorted vessel and can compromise any of the vascular structures (capillaries, veins, arteries, and lymphatic vessels), and often involve more than one component (1).

Venous malformation is a very common benign vascular lesion (1,2) and encompasses lesions that have been reported under different names, previously labeled as venous hemangioma and cavernous hemangioma. Most are sporadic, although a subset is related to specific genetic mutations (3) that can be inherited as an autosomal dominant trait (1).

It usually occurs on the head and neck, extremities and trunk (4), but rarely within the uterus (5), where it can affect any layer of the uterine wall, especially the myometrium. Most are discovered incidentally, but can present with abnormal vaginal bleeding or be associated with gynecological and obstetric complications (6). They are found at any age, without predominance for any decade (the youngest patient described was a 14-year-old girl) (7) and should be suspected in patients with a history of infections, curettage, therapeutic abortions, pelvic surgery, endometrial carcinoma or Gestational trophoblastic disease (8).

In the literature, some cases of this entity located in the uterus have been reported, such as "cavernous hemangiomas",

but the use of this term is discouraged, since these lesions do not represent a proliferative neoplasm as its name indicates (2). The exact incidence of the entity is unknown, due to the low number of reported cases (6).

We present a case of myometral venous malformation in a 44-year-old patient with abnormal vaginal bleeding and an ultrasound diagnosis of uterine myomatosis, which led to the performance of a hysterectomy. The vascular abnormality described in this case was not clinically suspected and was only detected in the histopathological study.

Case description

A 44-year-old woman (pregnant 3, deliveries 2, ectopic 1), consulted for 1 year of abundant daily vaginal bleeding (constant), associated with intermittent pain in the hypogastrium of moderate intensity. In her gynecological medical history, she reported menarche at age 14, two vaginal deliveries and a right ectopic pregnancy, with right salpingo-oophorectomy and uterine tubal ligation. She had no history of sexually transmitted diseases or abortions. But they had undergone a gynecological curettage a year earlier for metrorrhagia and a thickened endometrium of 17 mm, with a pathology report indicating endometrial polyps.

At the ultrasound evaluation, the uterus was irregular with diameters: longitudinal 15.6 cm, anteroposterior 9.5 cm and transverse 10 cm, for a volume of 780 cm3. Myometrium with heterogeneous echostructure and altered echogenicity due to the presence of some irregular rounded areas compatible with myomatous nuclei, the largest located intramural: 50 x 46 mm on the posterior surface, 16 x 20 mm on the uterine fundus and 25 on the anterior surface x 31 mm. Irregular, heterogeneous endometrium with a maximum thickness of 31.3mm. Pelvic vessels did not evidence any alterations on color Doppler examination. On gynecological examination, a mass was observed protruding from the internal cervical os, with fetid serosanguineous discharge, suggestive of a superinfected aborted myoma.

They performed a hysterectomy with a left salpingo-oophorectomy. In the Pathology laboratory, upon macroscopic study of the surgical specimen, a purplish polypoid lesion with necrotic and hemorrhagic areas was observed, which depended on the uterine fundus and measured 4 x 3 x 2.5 cm. When the polyp was cut, multiple congestive vascular structures were observed, some of them with intraluminal thrombi; These abnormal blood vessels also focally compromised the myometrium, in the area underlying the location of the polyp.Additionally, intramural nodulations suggestive of leiomyomas were identified (Figure 1).

On histopathological study, a benign vascular lesion was observed, made up of abundant irregular vascular structures, dilated and filled with blood, with thin walls, lined by a flat endothelium without atypia; some of these vessels were surrounded by bundles of smooth muscle and were observed in others intraluminal thrombi. The lesion was found in the middle of the myometrial smooth muscle bundles and the endometrial stroma in the polypoid formation, where ulcerated areas, ischemic and liquefactive necrosis were identified (Figure 2). Mitotic figures, cell proliferation, or pleomorphism were not observed.

Figure 1. Surgical specimen

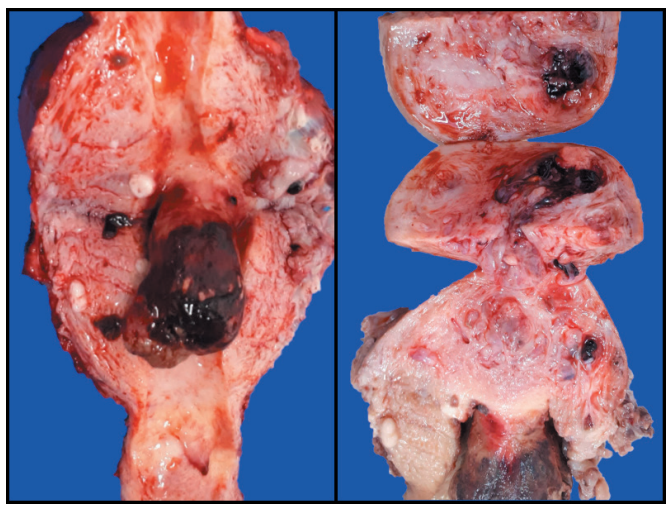
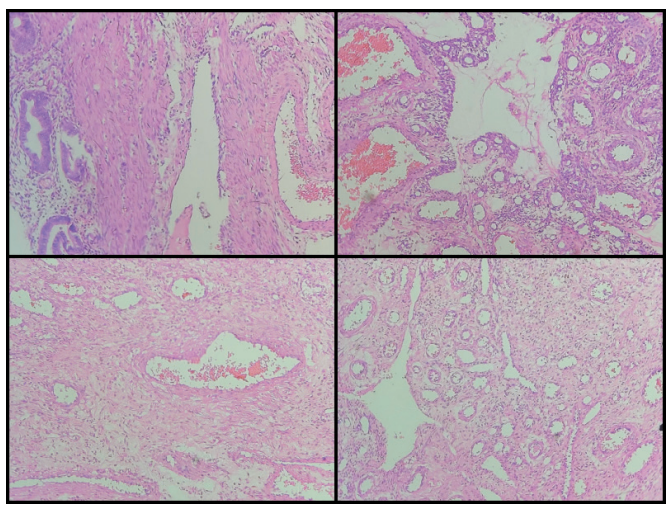


Figure 2. Microscopic findings



Discussion

Venous malformations of the uterus, although uncommon, are not rare to be found. They are lesions that should be suspected in women of reproductive age and postmenopausal stage with unexplained vaginal bleeding (8). Symptoms are variable, asymptomatic cases may occur or may present with abdominal pain, fertility disorders, pregnancy-related complications (6), and bleeding that can vary, from abnormal vaginal bleeding, menometrorrhagia, postcoital spotting and postmenopausal bleeding (1).

This entity typically presents as solitary, localized or segmental, superficial or deep lesions (9). Ranging from superficial varicosities to large and complex masses that infiltrate soft tissues, formed by groups of venous vessels, which due to slow flow develop intraluminal thrombi, calcification, and phleboliths (3). It can be diagnosed by ultrasound, showing a thickened uterine wall composed of cavernous spaces with turbulent flow (10). Also, they can present as an endometrial polyp (7), which can easily ulcerate, aggravating vaginal bleeding (1).

The definitive diagnosis requires the histopathological study of the surgical specimen (6,7). Microscopically, they are poorly circumscribed lesions, composed of abnormal veins of different sizes and proportions, often in a spongiform configuration, arranged randomly in the stroma (3). The endothelium is thin and mitotically inactive, the vessel walls do not have an internal elastic layer and contain a variable amount of smooth muscle. And the dilated vascular lumens fill with red blood cells and thrombi in various stages of organization. These thrombi help distinguish these low-flow lesions from high-flow arteriovenous malformations (9).

Some of the vessels are large with thick, irregularly attenuated muscular walls, while others are large but thin-walled. Lesions in which muscular-walled vessels predominate were classically called venous hemangiomas, while cases with thin-walled vessels were diagnosed as cavernous hemangiomas. According to recent changes in terminology, approved by the International Society for the Study of Vascular Abnormalities (ISSVA) (11), it is recommended to use the term hemangioma be used in lesions that arise as a result of cell proliferation, under the assumption that they are true neoplasms (3), which are characterized by returning spontaneously and inevitably before puberty (11). In contrast, vascular malformations do not have a growth phase or an involution phase and endothelial turnover is stable (1), which is why they are considered developmental defects, composed of dysmorphic vessels, which never return, but persist or grow progressively over time (11).

In the immunohistochemical study, endothelial cells in venous malformations are highlighted with CD31 (paraendothelial cell marker). Smooth muscle actin enhances pericytes and smooth muscle within the vascular wall. The Ki67 cell proliferation index is low and GLUT1 (a marker for diagnosing infantile hemangioma) is negative, as are the PROX-1 and D2-40 markers (1).

In our case, the patient had a history of gynecological curettage one year prior to the date of the consultation. We consider that the venous malformation could develop spontaneously and after intracavitary manipulation secondary to curettage. Due to the fact that the lesion developed a polypoid formation, which protruded from the internal cervical os, the ulceration and necrosis caused constant bleeding and superinfection of the lesion. As mentioned above, the definitive diagnosis of vascular malformation was made after the histopathological study.

Although vascular malformations are rare in the uterus, it is recommended to evaluate them with Color Doppler Ultrasound, the endometrium and myometrium, in patients with a history of menorrhagia or metrorrhagia, with long-term use of oral contraceptives, history of curettage, therapeutic abortions and pelvic surgery, since they are at risk of developing these injuries (8).

Conclusion

Although venous malformations are rare in the uterus, they should be considered among the differential diagnoses in the cases of women with vaginal bleeding of unclear cause. Additionally, we recommend following and using the new terminology for vascular lesions, proposed and accepted by the International Society for the Study of Vascular Abnormalities (ISSVA), available at https://www.issva.org/classification

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