CASE REPORT

Angiomatosis of the uterus, cervix and fallopian tubes: a rare and benign cause of intractable, heavy menstrual bleeding

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SUMMARY

Angiomatosis of the uterus, cervix and fallopian tubes is a rare and benign entity that has not been reported in the literature previously. We present a case of a 27-year-old patient with severe and intractable heavy menstrual bleeding unresponsive to all conservative and conventional forms of treatment. Following a laparoscopic hysterectomy, the histopathological finding of angiomatosis, a vascular abnormality in the uterus, cervix and fallopian tubes, provided a plausible explanation in this situation.

BACKGROUND

Our case report describes an uncommon and benign histopathological finding of a vascular abnormality in a postoperative specimen after hysterectomy. Specifically, on examination of the uterus, cervix and fallopian tubes, features consistent with angiomatosis were detected, providing a plausible explanation for the patient's unusual presentation and resistance to all forms of treatment, including an endometrial ablation.

A literature review has revealed that angiomatosis affecting the upper part of the female genital tract has not been previously reported in the literature.

In this instance, after an endometrial ablation, a second-look hysteroscopy revealed a uterine cavity that had been optimally ablated. Despite that, the patient claimed that her periods were unchanged; this was most unexpected.

We feel that this case should be published as a histopathological finding of angiomatosis of the pelvic organs has never been reported and may have future implications for the diagnosis, management and understanding of heavy menstrual bleeding.

CASE PRESENTATION

A 27-year-old, para 2 presented with heavy, painful periods unresponsive to medical management, including oral hormone therapy and the levonorgestrel releasing intrauterine system. The patient was requesting surgical management by way of a hysterectomy, feeling certain that she had completed her family. Both of her pregnancies were uneventful, continued to term and resulted in uncomplicated, spontaneous vaginal deliveries. There was no medical or surgical history of note.

Following counselling, she underwent a hysteroscopy, curettage and NovaSure endometrial ablative

procedure. Preoperative investigations, including coagulation profile, were all normal. At the time of surgery, the hysteroscopy revealed a normal endometrial cavity. A repeat hysteroscopy, following the NovaSure, confirmed an 'optimal' result.

The histopathology on the endometrial curettings was reported as 'normal proliferative endometrium'. Subsequent to this, the patient re-presented with ongoing severe, heavy periods and was requesting definitive surgical management. Initially, there was a reluctance to subject her to a hysterectomy, given her young age, but she was desperate for definitive treatment. Following further extensive counselling, she underwent a total laparoscopic hysterectomy. Intra-operatively, the pelvic organs were of normal appearance, and the laparoscopic hysterectomy was straight forward; her ovaries were conserved. The postoperative course was uneventful.

INVESTIGATIONS

Histopathological examination of the uterus and fallopian tubes revealed a haphazard excess of blood vessel formation within the stroma of the cervix and myometrium, with higher vascular density in the outer layers. The vessels were frequently arranged in groups around an artery or vein, while the surrounding vessels were thin walled. The fallopian tube muscularis was almost entirely replaced by networks of thin and thick blood vessels. The lesional vessels were not circumscribed but distributed diffusely throughout the uterus, cervix and fallopian tubes (figures 1 and 2). There was no evidence of malignancy, the cervical epithelium was normal and the endometrium was 'inactive'. The final histopathological diagnosis was 'Angiomatosis of the uterus, cervix and fallopian tubes'.

OUTCOME AND FOLLOW-UP

At postoperative follow-up 6 weeks and subsequently 5 years post hysterectomy, the patient reported complete resolution of bleeding and satisfaction with her treatment.

DISCUSSION

Angiomatosis is a rare, non-neoplastic vascular malformation presenting with diffuse infiltration of soft tissues. It is characterised by a proliferation of blood vessels with accompanying mature fat, fibrous tissue, lymphatics or nerves, that may involve skin, subcutaneous tissue, skeletal muscle, bone and

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Figure 1 Uterine corpus medium magnification. Higher magnification showing muscular vessels (upper right) with numerous smaller dilated vessels (left and centre).

rarely, smooth muscle. This benign vascular lesion affects tissues in a contiguous fashion by either vertically involving multiple tissue types or by involving similar tissue types.¹ Angiomas typically presents within the first 20 years of life, and the precise aetiology of the disorder is unknown. It likely represents a congenital vascular malformation and is thought to arise in early intrauterine life, affecting females more often than males. It most commonly affects the tissues of the lower limbs, followed by the abdomen, chest wall and upper limbs.

Although benign, most angiomas will eventually become symptomatic, and presenting complaints vary according to the location of the lesion. These can include pressure effect, diffuse swelling, pain, discolouration and bleeding.² This vascular proliferation tends to grow at a slow rate, and the definitive diagnosis is made by histopathological examination of affected tissue. When detectable on CT, it is visualised as serpiginous densities within low density areas corresponding to tortuous vessels. The only form of treatment is complete surgical excision, which is usually curative. Should surgery not be feasible due to the location of the lesion, embolisation can provide temporary arrest of growth and symptomatic relief. Following treatment, recurrences can occur at a rate of up to 50%, and surveillance appropriate to the location of the disease is recommended. Recurrences are thought to largely represent incomplete excision. Malignant transformation and metastases have not been reported.²

Figure 2 Uterine corpus low magnification. Scanning view of myometrium showing variably sized vascular channels with larger congested vessels more prominent towards the serosal surface (right).

A careful history and examination are required to rule out angiomas as part of a more systemic disease. While this benign vascular lesion can rarely present as an isolated lesion, it is typically associated with Von Hippel Lindau disease and Klippel-Trenaunay-Weber syndrome, both rare genetic disorders featuring abnormal vascular growths and malformations, among other systemic anomalies.³ The patient in question did not display any other features of these syndromes, and thus they were excluded.

Angiomas must also be distinguished histopathologically from other vascular tumours, such as capillary haemangioma, cavernous venous haemangioma, arteriovenous malformation (AVM), papillary endothelial hyperplasia (PEH) and bacillary angiomas.⁴ Histopathologically, haemangiomas are neoplastic lesions demonstrating endothelial hyperplasia.³ AVMs are characterised by the presence of malformed arteries and veins with dilated lumens involving submucosa but not muscularis propria.⁴ PEH is a focal lesion and does not involve arteries.⁴ Bacillary angiomas is a form of angiomas associated with bacteria of the *Bartonella* genus, rarely occurring in the immunocompetent population, and characterised histopathologically by lobular vascular proliferations lined with endothelial cells. Angiomas, in contrast, displays a highly characteristic pattern of a haphazard proliferation of vessels of varying sizes, particularly large veins. The latter have irregularly attenuated walls and intimal redundancies. However, the most distinctive feature is the presence of clusters of capillary vessels residing within, or just adjacent to, the vein walls.⁵

In general, the failure rate for NovaSure is relatively low and commonly quoted as 5%–10%, over a period of 5–10 years.⁶ This case demonstrates an unusual situation whereby a patient was deemed suitable to undergo an endometrial ablation and, from a technical point of view, the procedure was routine with a global treatment effect. Despite this, she reported that her bleeding pattern was unchanged.

Angiomas has rarely been reported to involve the female genital tract^{4,7} and has never been reported to involve the uterus. While this condition remains an extremely rare cause of heavy menstrual bleeding, it should be considered in a patient such as this one presenting with early-onset, heavy menstrual bleeding unresponsive to standard treatments.

The histopathological findings of a vascular abnormality in the uterus, cervix and fallopian tubes provided a plausible explanation for such an outcome. Treatment of angiomas is purely surgical and, in this case, was curative.

Learning points

- ▶ Angiomas of the uterus is a rare and benign condition.
- ▶ This finding is significant as it has not been previously reported in the literature and highlights the importance of maintaining a broad differential for excessive menstrual bleeding in a young, otherwise healthy, patient.
- ▶ The role of histopathological examination is crucial to the diagnosis of this condition.²

Contributors JCP: performed literature review, reviewed patient file/case notes/histopathology, wrote case report, arranged discussion and slides with histopathologist and wrote resubmission. BM: primary gynaecologist managing patient, primary surgeon at all surgeries, assisted with revision of case report, arranged discussion and slides with histopathologist and edited resubmission. VO: histopathologist.

Competing interests None declared.

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