



Angiomatosis of the Uterus in a Young Female Presenting with Heavy Menstrual Bleeding

Wei Ling Yeoh*, Ruchira Fernando

Department Anatomical Pathology, Launceston General Hospital, Tasmania, Australia

ABSTRACT

Angiomatosis is a vascular malformation characterized by a diffuse network of variably sized blood vessels involving multiple tissue planes. It is known to affect skin, deep soft tissue, bone and is often clinically extensive. Angiomatosis of the uterus is extremely rare with a paucity of literature describing this phenomenon. We present an unexpected case of angiomatosis of the uterus and cervix in a young female presenting with menorrhagia. 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.

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Introduction

A 29 year old lady presented with a long-standing history of dysmenorrhea and menorrhagia resulting in iron-deficiency. She had five uncomplicated vaginal deliveries and has no other relevant medical history. A transabdominal and transvaginal ultrasound demonstrated a normal endometrial thickness of 4.4 mm and a homogenous myometrium with no focal lesion. A trial of the hormone releasing intra-uterine device was poorly tolerated as it worsened her symptoms. She declined other conservative measures and opted for an elective total hysterectomy for definitive treatment.

Case Report

Macroscopically, the uterus appeared symmetrical and mildly enlarged with a dusky appearance. Sectioning showed a band-like dark purple discoloration of the outer one third of the cervix and myometrium associated with prominent blood vessels (Figure 1). Histological examination featured angiomatosis, characterized by a diffuse permeation of the outer half of the myometrium and cervix by a mixture of thick and thin walled vascular channels of varying sizes, lined by flattened, benign endothelial cells (Figure 2). The endometrium showed early secretory changes with no other abnormalities to account for abnormal bleeding. The Fallopian

tubes that were also removed did not demonstrate angiomatous lesions. She recovered well from the surgery. Angiomatosis, which is regarded as a diffuse form of hemangioma, is not included in the standardised nomenclature as a disease of the uterus. As a result, this condition is likely to be underdiagnosed and it continues to represent a pathological curiosity. Moreover, clinical examination and routine investigations for menorrhagia including hysteroscopy, endometrial curettage and ultrasonography are likely to be unhelpful in this instance. Therefore, definitive diagnosis of angiomatosis relies heavily on histopathology of hysterectomy specimens. Angiomatosis must also be distinguished histopathologically from other vascular tumors, such as capillary haemangioma, cavernous venous haemangioma, arteriovenous malformation (AVM), papillary endothelial hyperplasia (PEH) and bacillary angiomatosis.

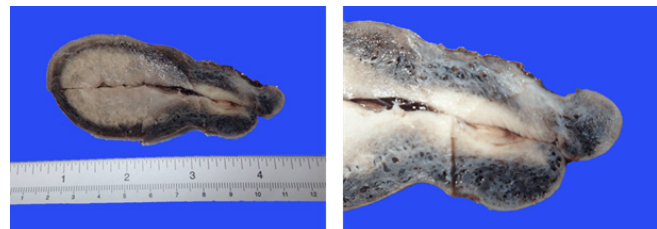


Figure 1. (Right) The uterus shows a continuous peripheral rim of dark purple discoloration. (Left)

The area of discoloration appears to be representing numerous congested blood vessels.

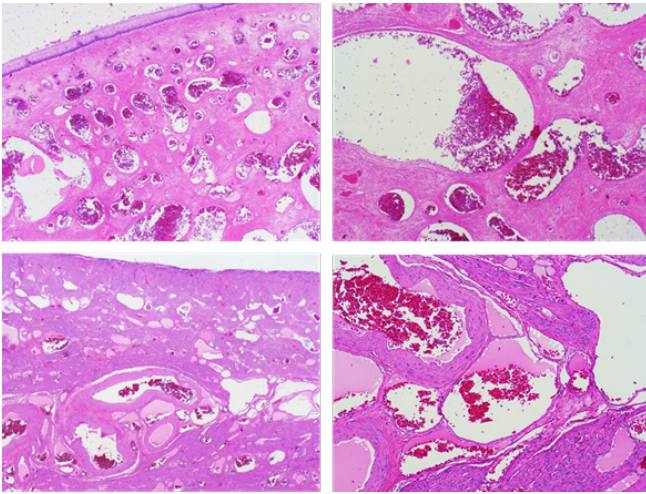


Figure 2. (Top right and left, H and E) Uterine cervix shows numerous ectatic vascular channels lined by benign endothelial cells. (Bottom right and left, H and E) Uterine corpus similarly shows vascular channels of varying sizes extensively involving the myometrium.

Discussion

Angiomatosis in the usual deep soft tissue sites can cause pain and persistent swelling that is worsened with strenuous exercise. Moreover, conventional angiomatosis can be associated with certain syndromes such as Klippel Trenaunay syndrome, Sneddon's syndrome or Gorham disease [1]. However, the clinical presentation and significance of this histological finding in the uterus is largely unknown. In a similar case report of a 27 year old with angiomatosis of the uterus, cervix and Fallopian tubes by C Pontre et al [2], the patient presented with intractable heavy menstrual bleeding that is unresponsive to all conventional and conservative forms of treatment. On a slightly different note within the same spectrum of vascular lesions, a retrospective study conducted in Taiwan on uterine haemangiomas, it was found that one of the most common presentations is menorrhagia [3]. Likewise, heavy, painful menstrual bleeding represents the chief complaint of our index case. Therefore, it appears that angiomatosis of the uterus represents a feasible explanation for heavy menstrual bleeding in young females with no other identifiable cause [4]. This is especially true in patients who are not responding to standard treatments. A rare case of

pregnancy in a patient with the syndrome where arterio-venous anomalies in this patient originally occurred in her right leg, but in the course of her first pregnancy she also developed circumscribed angiomatosis at the left and right side of the uterus. Her pregnancy was uneventful. However, because of prominent vascular changes in the cervix and lower uterine segment, there appeared to be a cephalo-pelvic discrepancy. 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 [5]. The lesional vessels were not circumscribed but distributed diffusely throughout the uterus, cervix and fallopian tubes [6]. Although benign, most angiomatoses will eventually become symptomatic, and presenting complaints vary according to the location of the lesion. These can include pressure effect, diffuse swelling, pain, discoloration and bleeding. Angiomatosis has rarely been reported to involve the female genital tract 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 will unresponsive to standard treatments [7-10].

Conclusion

As histopathological examination is the key to definitive diagnosis of angiomatosis of the uterus, maintaining a broad differential diagnosis and having a good understanding of various benign vascular lesions of the uterus is of paramount importance. More research into understanding the pathogenesis of this condition in the uterus will be the next step going forward. This should be followed by the development of diagnostic criteria and management guidelines to prevent undertreatment and/or overtreatment.

Disclosure Statement

The authors state that there are no conflicts of interest to disclose.

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