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Intramural uterine hemangioma: an insidious trap of a rare pathology. A case report

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Summary

Cavernous uterine hemangioma (UH) is an extremely rare benign pathology. The heterogeneity of symptoms and the rarity of such pathology render the diagnosis of UH quite difficult and it often relies on the final histologic examination. A case of a UH, previously diagnosed by ultrasound as a pelvic varicocele involving the uterine fundus, then revealed during a hysteroscopic endometrial resection, is described. The massive bleeding during the procedure caused a life-threatening which required an emergency abdominal hysterectomy. In case of an ultrasonographic diagnosis of atypical pelvic varicocele, more rigorous examinations should be planned before proceeding with an invasive uterine procedure. To the best of the authors' knowledge, this is the first report describing a UH discovered during an operative hysteroscopy, which could have dramatic consequences for the patient.

Key words: Hysteroscopy; Uterine hemangioma; Endometrial resection; Hemorrhage; Ultrasound.

Introduction

Cavernous uterine hemangioma (UH) is an extremely rare benign pathology [1]. To date, due to the different incidence reported [1-3], it is almost difficult to assess how many cases in the literature have been described. The correlated symptoms range from intermenstrual spotting, menometrorrhagia, and infertility to maternal and fetal demise from pronounced bleeding of the gravid uterus [3-5]. The rarity of such pathology and the heterogeneity of its symptoms render the diagnosis of uterine hemangioma quite difficult [1-3, 6].

Different treatments for the UH have been proposed, mainly depending on the presence and the severity of the bleeding. Conservative successful management as selective embolization or GnRH-analogue medical treatment as well as radical treatment (hysterectomy) have both been described [4, 6, 7].

Herein a case of uterine cavernous intramural hemangioma, previously diagnosed by ultrasound as a pelvic varicocele involving the uterine fundus, is described. The massive bleeding during a hysteroscopic endometrial resection caused a life-threatening which required an emergency abdominal hysterectomy. To the best of the authors' knowledge, this is the first report describing a UH discovered during an operative hysteroscopy, which could have dramatic consequences for the patient.

Case Report

A 42-years-old patient was referred to the present outpatient hysteroscopy unit for recurrent metrorrhagia. Unremarkable obstetric history with two uneventful spontaneous deliveries was reported. The woman suffered from recurrent heavy menstrual bleeding unresponsive to the medical treatment with levonorgestrel-releasing intrauterine system for approximately ten years. A transvaginal ultrasound scan previously performed showed multiple intramural uterine myomas, a thin endometrium, and the intrauterine device regularly placed into the uterine cavity. A noteworthy image extending from the posterior uterine wall to the fundus was reported (Figure 1). The detected blood flow was rather atypical but the finding was labelled as a pelvic varicocele by the sonographer. An outpatient free-anesthesia diagnostic hysteroscopy was performed: a regular uterine cavity with atrophic endometrium and the intrauterine device in a proper position were found. Considering the lack of response to the medical therapy, a hysteroscopic endometrial resection was planned.

A surgical pre-treatment of GnRH agonist (triptorelin 3.75 mg IM) was previously administered to the patient for three consecutive injections 28 days apart. The procedure was carried out using a nine-mm resectoscope with 0° optical system and glycine 1.5% as distending media. The electric loop was powered by 100 W monopolar current in pure cutting mode. After the insertion of the resectoscope, the endometrium of the uterine fundus, including 2-3 mm of myometrium, was initially resected by the electric cutting loop and a massive hemorrhage began immediately. Uterine massage and electric coagulation did not allow to control the bleeding. Considering the unrestrained hemorrhage, no other attempts to stop the bleeding were done and an emergency abdominal hysterectomy was immediately carried out. In a few minutes, the hemoglobin decreased from 13.7 g/dl to 5.5 g/dl. At the be-

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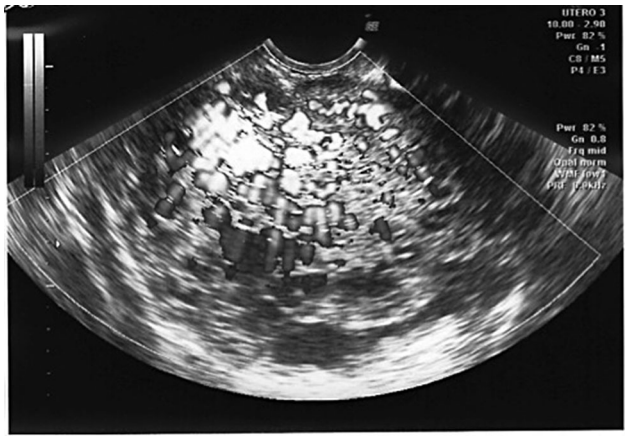

Figure 1. — Transvaginal ultrasound with Doppler velocimetry of the uterine posterior wall and fundus.

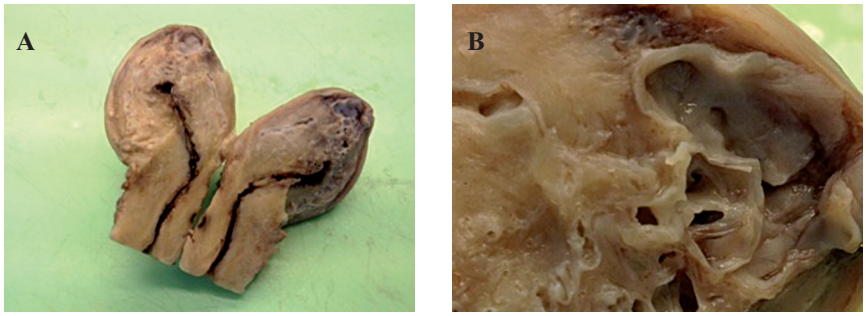

Figure 2A, B. — Longitudinal section of the uterus showing the hemangioma involving the posterior wall and the fundus.

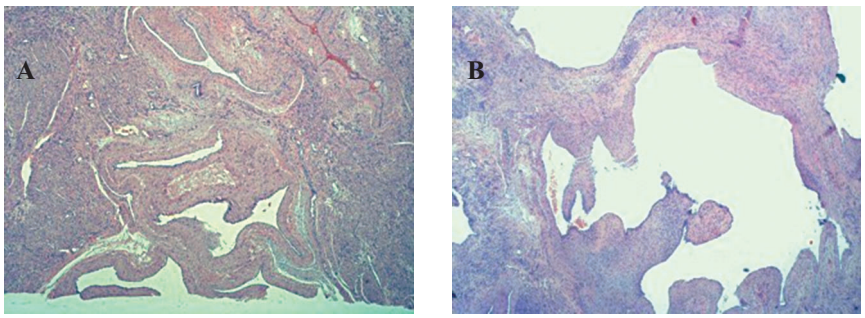

Figure 3A, B. — Microscopic aspects of the uterine hemangioma presenting a picture of irregularly shaped, cavernous vascular spaces infiltrating between the myometrial fascicles.

ginning of surgery, no blood was found in the peritoneal cavity. In order to stop the bleeding, uterine arteries were clamped and subsequently hysterectomy was performed. The patient received four units of blood and five units of plasma during surgery. During the first 24 hours after surgery four units of blood and eight units of plasma were administered and the level of hemoglobin rose to 9.9 g/dl. After six days of hospitalization the hemoglobin was 10.3 g/dl and the patient was discharged.

The pathologist described the uterus with a complex nodular growth and dilated vascular spaces within the myometrium, immediately below (3 mm) the endometrium (Figures 2A, B). Microscopically, UH presented a picture of irregularly shaped, cavernous vascular spaces infiltrating between the myometrial fascicles. The large vascular spaces were walled by flat endothelial cells and distended by blood. No cell atypia was seen (Figures 3A, B).

Discussion

A rare case of uterine intramural hemangioma, previously diagnosed by ultrasound as a pelvic varicocele involving the uterine fundus, was described. The massive bleeding during a hysteroscopic endometrial resection caused a life-threatening event which required an emergency abdominal hysterectomy. To the best of the authors' knowledge, this is the first report describing a UH discovered during a hysteroscopic procedure, which could have dramatic consequences for the patient. The peculiarity lies also in the fact that the surgical procedure was began with an incorrect ultrasonographic diagnosis, which was mainly due to the rarity of the uterine pathology rather than to a medical error. This led to a life-threatening event, which could have been

avoided with more rigorous examinations.

Cavernous UH is an extremely rare benign pathology, first incidentally described in 1897 during an autopsy of a young women died 24 hours after delivering twins [1]. UH is a benign tissue malformation, which may be with a localized or diffuse appearance. From the anatomical viewpoint, it is possible to detect it in the cervix but it involves more frequently the entire wall of uterine corpus, from the endometrium to the serosa [1, 5]. Due to its anatomical conformation characterized by a vascular dilatation, UH has to be differentiated from other benign lesions, such as adenomatoid tumor, lymphangioma, and arteriovenous malformation[1].

The pathogenesis of UH may be congenital or acquired. Congenital hemangioma is usually associated with congenital diseases as Klippel-Trenaunay syndrome, Maffucci syndrome, as well as Kasabach-Merritt syndrome [1, 5, 8]. On the contrary, hormonal changes seem to be the cause of the acquired variety. Estrogen could play a role in the vasculogenesis and angiogenesis of hemangiomas via an indirect pathway of angiogenic factors [9]. In the present case, the patient referred two uneventful pregnancies with spontaneous deliveries. Although several cases of UH were diagnosed in pregnant women [2], the authors speculate that at the time of pregnancies, the malformation was possibly not present or at least not so highly developed. Nevertheless, in subsequent years a levonorgestrel-releasing intrauterine device was inserted to treat a recurrent metrorrhagia. Despite several ultrasound scans performed, only in the two last examinations, a remarkable vascular alteration was observed.

Due to the heterogeneity of symptoms and the rarity of such pathology, the diagnosis of UH may result quite difficult and often relies on the final histologic examination [1-3, 6]. Indeed, although an instrumental diagnosis could be established, the rarity of the pathology places it at risk of missed identification [7, 10-12]. Vaginal examination, uterine curettage specimens, ultrasonography, and hysterography are usually uninformative [1]. Color Doppler ultrasonography may provide an initial suspect of UH, but MRI represents a more accurate option and a valid alternative to angiography and CT scan in case of pregnant women [2, 3]. In the present case, the ultrasonographic diagnosis of pelvic varicocele significantly influenced the surgeon's choice to proceed with a hysteroscopic endometrial resection. Furthermore, contrary to previous cases in which the intrauterine malformation was hysteroscopically observed [12, 13], the diagnostic hysteroscopy revealed a normal cavity with atrophic endometrium.

Considering the rarity of the pathology and the few cases reported in the literature, it is difficult to assess which is the best option to treat a UH. In order to preserve fertility, several treatments as laser or knife excision, cryotherapy, electrocauterization, internal artery ligation, uterine artery embolization, laser ablation, as well as radiotherapy have been described [2, 3, 13, 14], but in cases that do not respond to conservative treatments, hysterectomy should be considered [1, 3].

In conclusion, the UH is a dangerous and insidious condition where, due to the rarity of the pathology, a proper diagnosis is unlikely made. The authors hope that the description of this case may help to avoid similar life-threatening situations, and that in case of ultrasonographic diagnosis of atypical pelvic varicocele, more rigorous examinations can be planned before proceeding with an invasive uterine treatment.

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