

Vulvar venous malformations: surgical treatment remains an option

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Abstract

Background. Our aim was to evaluate the benefit of surgical resection of the venous malformation (VM) of the external female genitalia.

Methods. Over the period of 2009-2019, 18 consecutive females underwent surgical resection for vulvar VM. Evaluations included preoperative Doppler ultrasound, MRI, and pre-and post-operative photographic imaging. The main outcomes were: residual pain, cosmetic distortion, residual VM, and quality of life.

Results. Over a 10 year periods, 18 females, mean age 35 years (range 9-71) were included in this study. All patients were symptomatic: 16 had intermittent pain or discomfort, 1 had bleeding and 2 requested cosmetic treatment. Of these cases, there were 5 isolated vulvar VM, 12 associated VM: 3 of the clitoral hood, 3 troncular pelvic vein insufficiency and 12 of the lower limb. Eight patients had undergone previous procedures: 2 sclerotherapy treatments (1 to 3 sessions), 4 partial surgical resections. There were 18 single resections in the vulva (7 focal, 11 complete), 2 partial resections in clitoral hood and 2 had resection of a VM in the peritoneo-vaginal canal at the same time. The mean follow-up was 42.9 months (range 6-120). Two patients were lost to follow-up at 6 months. For all patients, elimination of pain and soft tissue redundancy was achieved. Two patients had persistent discomfort and 2 requested cosmetic treatment.

Conclusion. Surgical resection of vulvar VM can be the best approach with few postoperative complications, good functional and cosmetic results. Appropriate preoperative evaluation is required to identify isolated VM or VM associated with ovarian vein or internal iliac vein insufficiency requiring to be treated before surgery.

Key words: venous malformations, female external genitalia, vulva, surgery, gynecology

Résumé

Malformations veineuses de la vulve : le traitement chirurgical reste une option

Objet. Le but était d'évaluer le bénéfice du traitement chirurgical des malformations veineuses (MV) du périnée antérieur de la femme.

Méthodes. Entre 2009 et 2019, 18 femmes ont eu un traitement chirurgical de MV vulvaires. Les investigations préopératoires incluaient un doppler-échographie, une imagerie par résonance magnétique (IRM), et des photos pré- et post-opératoires. Les principaux critères de jugement étaient la douleur résiduelle, la déformation, la malformation résiduelle, et la qualité de vie.

Résultats. Sur une période de 10 ans, 18 femmes (moyenne d'âge 35 ans [9-71]) ont été incluses dans cette étude. Toutes les patientes étaient symptomatiques : 16 avec des douleurs intermittentes, une avait des saignements, et deux souhaitaient un traitement cosmétique. Parmi ces cas, cinq avaient une MV isolée vulvaire, 12 avaient des MV associées (trois de la

Tirés à part :
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couverture du clitoris, trois avaient une incontinence des veines du pelvis, 12 avaient des MV du membre inférieur ipsilatéral). Toutes les patientes ont eu une simple résection de la grande lèvre, deux une résection du capuchon du clitoris, et deux une résection d'une MV du canal péritonéo-vaginal durant la même chirurgie. Le suivi moyen a été de 42,9 mois (6-120). Deux patientes ont été perdues de vue à six mois. Toutes les patientes avaient une disparition des douleurs et de l'hypertrophie tissulaire, deux présentaient une gêne résiduelle, et deux étaient demandeuses d'une reprise cosmétique.

Conclusion. La résection chirurgicale des MV des grandes lèvres apparaît une stratégie valable en raison de faibles complications post-opératoires, de bons résultats cosmétiques et fonctionnels. Une évaluation pré-opératoire est utile pour identifier une MV isolée ou associée à une incontinence des veines du pelvis, cette dernière justifiant un traitement préalable.

Mots clés : malformations veineuses, organes génitaux externes féminins, vulve, chirurgie, gynécologie

The field of vascular anomalies encompasses a broad spectrum of lesions, the prevalence of this disease in the general population is 1.5% [1]. They are classified by their flow characteristics: slow flow malformations can be divided into lymphatic and venous malformations; a fast flow form is an arteriovenous malformation. Venous malformations (VM) can occur anywhere in the body, they account for more than 80% of all vascular malformations with less than 1% of these confined to the vulva. VM of the female external genitalia may cause significant patient distress in terms of self-image, pain, bleeding and impaired function [2].

Regional VM can involve the labia, clitoral hood, clitoris or vagina. There are some predictable patterns, including increased symptomatology during puberty and pregnancy, but trauma and spontaneous thrombosis can also play a role in exacerbating symptoms.

The treatment of VM often targets the labia and clitoral hood. The diagnostic and therapeutic modality of choice is yet to be determined because clinical experience remains relatively limited.

In the literature, VM were commonly treated with sclerotherapy or a combination with surgery, rarely by a single surgery. Sclerotherapy is the first line treatment [3-7], in many centers, for symptomatic lesions. It can alleviate symptoms with, often, short term improvement or the need of numerous sessions [3, 8].

Preoperative sclerotherapy has not been demonstrated to have a significant impact on blood loss at the time of surgery and quality of results [8]. Surgery currently seems to be the only way to potentially treat VM successfully.

Our hypothesis is that a better clinical result is obtained after single surgery rather than sclerotherapy only or a combination of the two techniques.

The aim of this study was to analyze the improvement in pain, cosmetic problem, residual VM and quality of life resulting of a single surgery.

Materials and methods

Study design

This was a retrospective, observational, single center study. Patients were selected by the same multidisciplinary team and operations were carried out by the same surgical team. Surgery was supervised and/or performed by the same surgeon.

A total of 18 consecutive symptomatic patients were included in this study between 2009 and 2019. Asymptomatic or minimally symptomatic patients as well as patients with infantile hemangioma of the labia were not included in the study.

All patients or legal guardian(s) were provided with oral and written information about the surgery and they all provided their written consent for surgery.

For this cohort, the review of data was approved by the ethics committee of Lariboisiere Hospital Paris.

Data collection

Data was collected from each patient's medical record: medical history, clinical findings, photographic images, Doppler ultrasound, MRI (magnetic resonance imaging), laboratory tests results and the assessment score of their quality of life (QoL). Further information was also collected: surgical procedures, postoperative complications, intraoperative blood loss, and surgery duration.

We used the following definition for the VM: focal VM developed in a part of the labia majora and superficial, extensive VM in all the labia majora with, in some cases, extension in the perineal cellular space.

An institutional score of QoL was determined using various factors: pain/heaviness, cosmetic distortion, sexual intercourse, educational and/or professional activities, and residual VM. Results were self-assessed by patients.

Additionally, at six months and at a later follow-up visit, pain was rated on a scale of 1 to 4: (1: no/minimal pain, 2: discomfort, 3: intermittent pain, 4: permanent pain [phlebothrombosis]). The score for cosmetic distortion was based on a scale of 1 to 3 (1: no/minimal distortion, 2: moderated distortion, 3: significant distortion).

Clinical protocol

The patients were selected based on their persistent symptoms following medical treatment (antiplatelet medication and elastic garment [panty], rarely venotonic drug).

All patients underwent the same imaging procedures which included: venous ultrasound (US), MRI, and hematology tests. A Doppler ultrasound was performed to detect troncular venous anomalies. A T2 fat-saturation weighted MRI consisted of two values:

- a diagnostic value;
- an appreciation of extent of VM. Phlebography was indicated if pelvic troncular venous insufficiency was identified by ultrasound.

Hematologic tests were done (D-dimers, fibrinogen, platelets) to evaluate the potential increased risk of hemorrhagic events due to the presence of a local intravascular coagulopathy (LIC).

A limited walking was recommended for two weeks to avoid delayed healing, low molecular weight heparin (LMWH) and elastic garment to be worn for 1 month were prescribed. Clinical follow-up exams and photographic images were carried out at 6 and 12 months after surgery. Most of the patients received an annual interdisciplinary evaluation.

Results

Clinical findings

Surgery was performed on 18 patients between 2009 and 2019 (18 females, mean age 35 years [range 9-71]). Seven had previously undergone procedures: 2 sclerotherapy treatments (1 to 2 sessions), 2 sclerotherapies follow by partial surgery, 2 partial resections of the labia majora, and 1 preoperative venous embolization of left ovarian vein.

Patients were symptomatic: 12 had pain or heaviness, 9 discomfort, 7 had cosmetic distortion, 5 had pain during sexual intercourse, and 3 experienced one or more episodes of bleeding. Two patients presented with segmentary thrombophlebitis with severe pain (*table 1*).

Diagnosis of VM was made at birth in 9 patients, 7 were symptomatic at puberty, and 7 reported significant progress of the VM after pregnancy.

Clinical examination provided the following information: soft tissue swelling was significant in 11 patients; and all lesions were confined to the labia majora. Seven patients had focal VM, and 11 had extensive lesions on all the labia (*figure 1*) There was no labia minora or vaginal involvement. Fifteen VM were unilateral, 3 bilateral with a contralateral focal asymptomatic component.

Associated lesions were:

- on the perineum, 3 on the clitoral hood, 2 on the clitoris, 3 varicosities in the mons pubis, and 3 presented venous insufficiency in the peritoneovaginal canal;
- in other areas, 11 had capillary-venous VM on the ipsilateral limb. Among patients of child-bearing age, no complications of pregnancy were noted during delivery.

Non-invasive investigations

US and MRI were used to evaluate associated lesions. Ultrasound was use to explore troncular venous anomalies such as: insufficiency of ovarian veins (1 patient), as well associated anomalies of veins of the peritoneovaginal canal (3 patients).

MRI depicted the extension of VM in the perineal cellular space, or associated VM or other pathology in the pelvis, and confirmed cavernous venous spaces in the labia on T2-weighted images (*figure 2*).

Hematologic tests did not show significant abnormalities (D-dimer test <1000ng/ml).

Operative surgical treatment

None of the patients had preoperative sclerotherapy and one patient had preoperative embolization of an incontinent ovarian vein. All patients underwent a single surgical procedure.

Skin resection was limited to superficial lesions, otherwise, all skin coverage was conserved. The medial border of the thin skin of the labia majora was always spared.

In the lateral part of the labia (first step), dissection was carried out at the edge of the VM with progressive ligation of venous drainages. Next, the dissection progressed to the upper part, then to the lower part of the labia where the main drainages through the perianal area is located.

In the medial area, a precise mobilization of the mass in contact with the thin skin covering was done with precise hemostasis using bipolar cautery (most difficult step of the resection). In the deep area, dissection was continued in the cellular space to obtain a complete resection of the VM.

Table 1. Clinical data.

SYMPTOMS	Pain and/or Heaviness	Discomfort	Cosmetic Distortion	Painful Sexual Intercourse	Bleeding
Patients n (%)	12 (66) ¹	9 (50)	7 (39)	5 (28)	3 (17)

¹ 2 with thrombophlebitis

We preserved as much of the underlying fat pad as possible to maintain a natural contour (*figure 3*).

A drain was placed in the wound exiting in the mons pubis. Redundant skin was never excised permitting physiologic remodeling. Two patients underwent resection of incontinent veins in the peritoneovaginal canal at the same time (*table 2*).

None of the patients needed a blood transfusion. The mean blood loss was 180mL (100-300mL). The median surgery duration was 3h 20min (range 2-6h). All patients had a histological examination that revealed a pure common VM according to ISSVA classification.

Postoperative course

In the early postoperative period, 7 patients experienced delayed healing, often at the lower part of incision. One

patient had a local hematoma without reoperation. Five reported hypoesthesia around the operated area.

Academic or professional activities were interrupted for 1 month for most patients.

Follow-up

At the 6-month follow-up, 15 patients had no pain or heaviness. Local soft tissue redundancy was eliminated in all patients, but 3 had dissymmetry of the anatomical area and requested cosmetic treatment (*table 3*).

At the final clinical visit (mean follow-up 42.9months, average 10-120), 3 patients were lost of follow-up after 6 months, 14 patients (93%) reported no residual pain or heaviness, no patients required additional surgery or sclerotherapy.

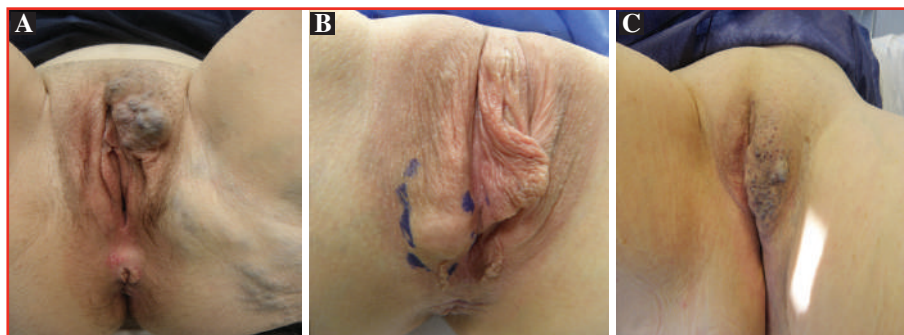


Figure 1. Clinical appearances of VM (labia majora). (A) Superficial focal VM of labia and mons pubis. (B) Multifocal VM with normal skin covering. (C) Extensive VM of the labia majora.

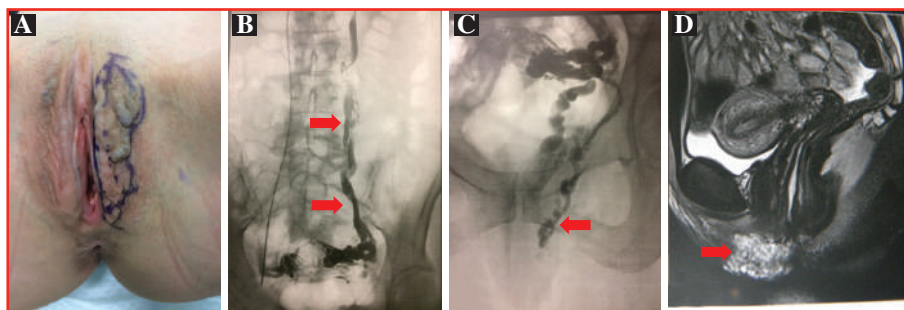


Figure 2. VM of the labia majora associated with venous troncular insufficiency. (A) Clinical appearance. (B) Phlebography: venous reflux of the left ovarian vein (arrow: ovarian vein). (C) Phlebography: venous reflux in the veins of the peritoneovaginal canal (arrow). (D) MRI on T2 fat saturation: heterogeneous hyper signal of labial VM (arrow).

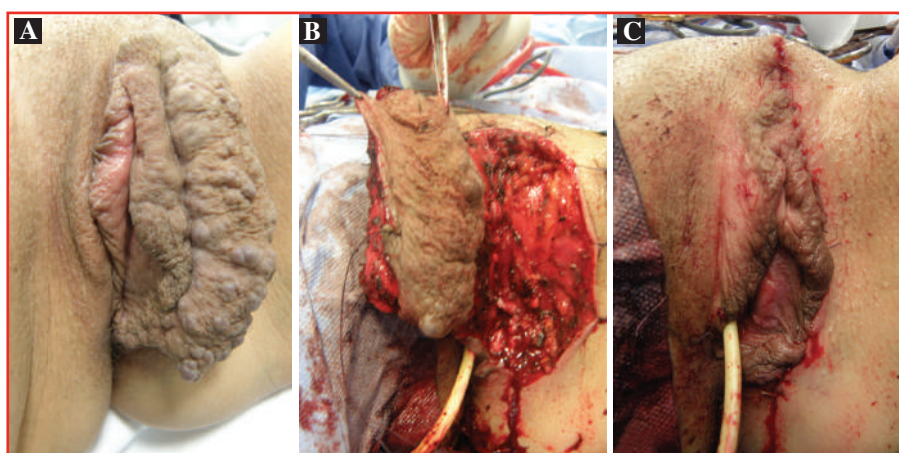


Figure 3. Operative views of the surgical procedure. (A) Preoperative clinical appearance. (B) Resection of extensive VM of the labia. (C) Reconstruction of labia majora, conservation of the median skin covering of the labia.

Table 2. Surgical procedures.

Number (%)		
VM resection	Focal VM of the labia	Extensive VM of the labia
Patients n (%)	7 (38)	11 (62)
Associated procedures	VM resection Clitoral hood	VM resection of peritoneovaginal canal
Patients n (%)	2(11)	2 (11)

Table 3. Pain score before and after surgery.

Pain score	Number (%)		
	Preoperative 18 patients	At 6 months 18 patients	At last follow-up 15 patients ¹
Minimal/ no pain	1(6)	15(83)	14(93) ²
Patient's n (%)			
Discomfort	7(39)	3(17)	1(7)
Patient's n (%)			
Intermittent pain	8(44)		
Patient's n (%)			
Permanent pain (phlebothrombosis)	2(11)		
Patient's n (%)			

¹ 2 patients were pregnant again at 2 and 3 years.

² Statistically significant change from baseline value (Student's t test p < 0, 05).

Quality of life

At the final follow-up, fourteen patients (93%) had no/minimal pain, 10 (67%) had no or minimal distortion, 4 (27%) had residual dissymmetry and 2 requested cosmetic treatment, 1 reported discomfort during sexual intercourse, 1 had recurrent VM occurred after pregnancy, 1 residual VM on the mons pubis (*figure 4*).

Discussion

In this report, we present the outcomes of 18 patients with VM of the female genitalia, who underwent single surgery at a single institution within a 10-year study period.

Sclerotherapy has reduced the indication of surgery in this anatomical area the last 10-years. The incomplete clinical results after sclerotherapy and the need of numerous sessions led, our team, to choose a surgical option [8, 9]. The limited number of cases and the frequency of previous treatment make it difficult comparative study with each procedure.

Clinical symptoms

Specific symptoms associated with this anatomical area are pain/heaviness and cosmetic distortion. The clinical course and assessment provide the clue for diagnosis in most instances. Superficial VM appear as soft, bluish, compressible masses on the labia majora which cause the labia to become distorted due to the unilateral vulvar mass. If deeper within soft tissue, they may not apparent until later in life. The overlying skin may be normal or may contain capillary-venous lesions. Spontaneous phlebothrombosis or trauma may lead to episodes of acute pain requiring a low molecular weight heparin drug (LMWH). The majority of VM are sporadic without somatic mutation.

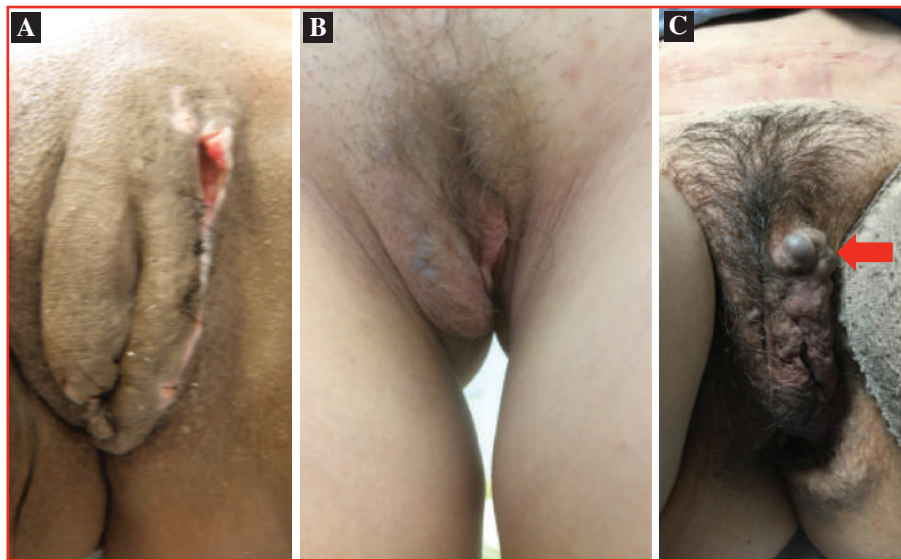


Figure 4. Post-operative follow-up. (A) Delayed healing in the proximal part of the labia. (B). Residual dissymmetry. (C) Recurrent VM after ten years follow-up and two new pregnancies.

Anatomical area

We report VM in the unilateral labia majora without involvement of the labia minora, but associated locations of VM may be found in the contralateral labia (4 cases with a nodular appearance), or in the clitoral hood (3 patients), the clitoris (2 patients) or vagina. In our cohort, 80% of the vulvar VM are associated with capillary-venous malformation of the ipsilateral limb.

Differential diagnosis

Because of the superficial position, radiological investigations for vulvar masses in our series were often unnecessary, except for extensive VM (VM in all the labia and or not extension in the perineal cellular space). Doppler-ultrasound and MRI could be selectively done for differential diagnosis of: vulvar varices [10], Bartholin's cyst or malignancies. Pelvic compression syndrome, common cause of chronic pelvic pain, is rarely associated with vulvar varices [11].

Treatment options

Three strategies were reported: single sclerotherapy, a combination of sclerotherapy followed by surgery, or a single surgical resection.

– *Percutaneous sclerotherapy* is the first line of treatment in some centers [5-7], but it is not a minimally-invasive

technique in this location: General anesthesia is needed to ensure adequate compliance (painful procedure), sessions are numerous, and there is limited tolerated products such as Polidocanol or STS (e.g. Thrombovar) [5]. In this location, it is difficult to control the venous drainage and there is a risk of non target embolization [3]. Residual VM is frequent and can be evaluated by patient satisfaction.

– *Combination of sclerotherapy and surgical resection.* The objective was to increasing the likelihood of clinical success and to minimizing intraoperative blood loss [3, 8]: sclerotherapy was made in the most symptomatic part of VM and surgical resection was limited to the embolized mass presenting risk of secondary extension of non excised vascular channels. In our cohort, 2 patients had a single previous sclerotherapy treatment and 4 had sclerotherapy followed by partial resection. All had poor clinical results and were scheduled for surgical resection.

– *Single surgical resection.* Surgery is the only way to potentially treat venous malformations successfully with a specific target on extensive VM of the labia. Good exposure with or without resection of the skin covering makes it possible to control venous drainage around the VM and to obtain a complete resection. Preservation of the medial thin skin coverage of the labia permits to avoid functional sequels. In our cohort, we report no excessive blood loss during surgery (mean 130mL), and few complications (no significant hematoma, delayed superficial healing). Intraoperative time could be longer than with others techniques.

Residual malformation

Residual VM frequent after sclerotherapy, possible after surgical resection, is not always a major concern, and is reported through patient satisfaction. Residual VM is a clinical evaluation, and MRI control is justified only for significant recurrence. During the follow-up (mean 42.9 months), 1 patient had focal recurrence of VM after a new pregnancy. No secondary surgery was requested by the patient.

Therapeutic indications

Conservative treatment is often proposed as a first step: aspirin and an elastic garment, or LMWH for focal phlebothrombosis. Indications for alternative treatment include recurrent pain, swelling, cosmetic concerns, and dysfunction.

– *Focal nodular VM* can be treated by first line sclerotherapy (interventional radiologist). For pediatric patients, sclerotherapy can be used for delaying surgery.

– *Painful phlebothrombosis* is treated by LMWH, and repeated episodes need to be treated by surgery.

– *Multi focal nodular VM*, or cavernous focal lesions could be treated by surgery. A gynecologic center with an experience of vulvar pathology can take in charge this group of patients.

– *Patients with extensive labial VM* underwent surgical resection without preoperative sclerotherapy in our serie. Resection of VM of the clitoral hood or of the peritoneovaginal canal can be performed at the same time. This procedure needs a competence in the treatment of vascular malformations. In our cohort multiple or stepwise resection was not required.

Conclusion

A wide spectrum of tumors can be found in the female external genitalia, and experience is needed to obtain the correct diagnosis and choose an appropriate management plan. Sclerotherapy is the first line treatment for focal VM

of the labia majora, and for pediatric patients. Surgery is the only way to successfully treat multifocal or extensive lesions of the labia. Carefully selected surgical indications can lead to successful outcomes. Surgical resection seems to be a valuable treatment option in our retrospective, single center analysis. ■

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Disclosures. None of the authors have any conflict of interest to declare.

References

1. Eifert S, Villavicencio JL, Kao TC, Taute BM, Rich NM. Prevalence of deep venous anomalies in congenital vascular malformations of venous predominance. *J Vasc Surg* 2000 ; 31 : 462-71.
2. Annam A. Female pelvic vascular malformations. *Semin Intervent Radiol* 2018 ; 35 : 62-8.
3. Vogel AM, Alesbury JM, Burrows PE, Fischman SJ. Vascular anomalies of the female external genitalia. *J Pediatr Surg* 2006 ; 41 : 993-9.
4. Herman AR, Morello F, Strickland JL. Vulvar venous malformations in a 11-year-old girl: a case report. *J Pediatr Adolesc Gynecol* 2004 ; 17 : 179-81.
5. Krokidis M, Ventucci P, Hatzidakis A, Iaccarino V. Sodium tetracycl sulfate direct intralesional sclerotherapy of venous, malformations of the vulva and vagina: report of five cases. *Cardiovasc Intervent Radiol* 2011 ; 34 : S228-31.
6. Marrocco-Trischitta M, Nicodemi EM, Nater C, Stillo F. Management of congenital venous malformations of the vulva. *Obstet Gynecol* 2001 ; 98 : 789-93.
7. Barral PA, Petit P, Bartolli JM. A rare case of a venous malformation of the clitoris. *Eur J Obst Gynecol Reprod Biol* 2018 ; 224 : 202-3.
8. Nassari N, Teresa MJO, Rosen RJ, Moritz J, Waner M. Staged endovascular and surgical treatment of slow-flow vulvar venous malformations. *Am J Obstet Gynecol* 2013 ; 208 : 366e1-3666.
9. Wang S, Lang JH, Zhou HM. Venous malformations of the female lower genital tract. *Eur J Obstet Gynecol Reprod Biol* 2009 ; 145 : 205-8.
10. Bell D, Kane PB, Liang S, Conway C, Tornos C. Vulvar varices: an uncommon entity in surgical pathology. *Int J Gynecol Pathol* 2007 ; 26 : 99-101.
11. Gaurilov SG, Karalkin AV, Turischeva OO. Compression treatment of pelvic congestion syndrome. *Phlebology* 2018 ; 36 : 418-24.