MANAGEMENT OF AN AESTHETICALLY DISABLING COMPLEX VULVAR VENOUS MALFORMATION IN A YOUNG WOMAN

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Abstract

Venous malformations (VM) of vulva, perineum and pelvis are uncommon condition which may present with cutaneous varices or aesthetically disabling swelling of external genitalia. Herein, we report a young woman who presented with a large left vulva bluish tinged swelling, progressively increasing in size since birth. Computed tomography of the pelvis and lower limbs confirmed the diagnosis of extensive VM of pelvis and perineum. She underwent selective angiogram which revealed venous malformations of left vulva and gluteal region with no arterial supply to the lesion. Surgical excision was performed but complicated with bleeding which necessitate multimodal hemostatic procedures and blood transfusion. There is no recurrent swelling after 5 years follow up. Surgical excision of vulvar venous malformation offer good cosmetic outcome. However, appropriate planning with axial scan, angiogram and operative technique including multimodal hemostatic measure are important steps to achieve good result with no recurrence**.** We discussed on challenges on management of this case in relation to the currently available literature.

Keywords: *Venous Malformation, Vascular Malformation, Perineum and Pelvis*

Introduction

Venous malformations (VMs) of the vulva, perineum and pelvis are rare. Although most patients are asymptomatic, it may potentially cause sexual dysfunction due to its increasing size and awkward position. The management of this condition remains a challenge due to the unpredictable progression and high recurrence rate. We present a case of VM of the vulva which was treated with surgical excision and associated with torrential bleeding requiring multimodal hemostasis. The challenges and complications encountered are discussed.

Case Report

A 28-year-old woman presented with a history of increasing vulva swelling since birth. It was a painless swelling and caused a severe disfigurement of her perineal area. She sought surgical treatment when she was getting married. On examination, there was a non-pulsatile swelling at the left labia measuring 6 cm x 5 cm (Figure 1) consisting of dilated veins which was soft and compressible. The left lower limb was swollen with varicosities involving the entire limb. Her platelet count was 108 x 10^9/L, hemoglobin level was 11.6 g/dL. Computed tomography angiogram (CTA) of the lower limb revealed extensive vascular malformations involving the left pelvis and left lower limb with two foci of venous malformations in the vastus intermedius and adductor magnus muscle respectively (Figure 2 and 3).

Angiogram was done with selective left common iliac run revealed delayed contrast filling up venous channel at left vulva and left gluteal region. There was no arterial supply to the lesion. Hence, no embolization was performed. According to the International Society for The Study of Vascular Anomalies, her lesion was classified as a common venous malformation. Surgical excision of the venous malformations was performed with combination of argon plasma and advanced bipolar energy device (LigaSure, Covidien, USA), and the hemostasis was complimented with tissue glue. The wound was primarily closed. Histopathology report was consistent with venous malformation with no evidence of angiosarcoma. Post-operatively, she had complications of wound dehiscence and bleeding. The bleeding was major and revealed

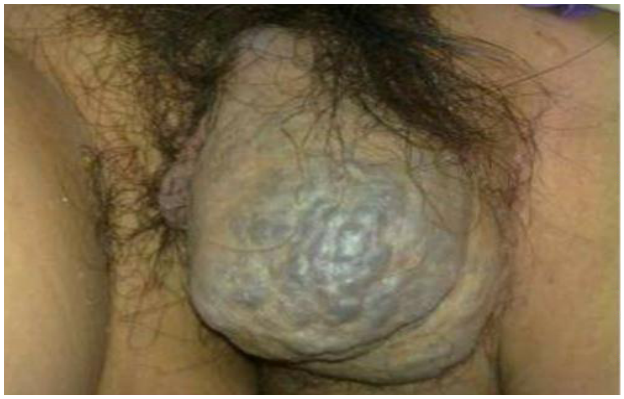

Figure 1: Left labial majora bluish swelling due to the extensive venous malformation

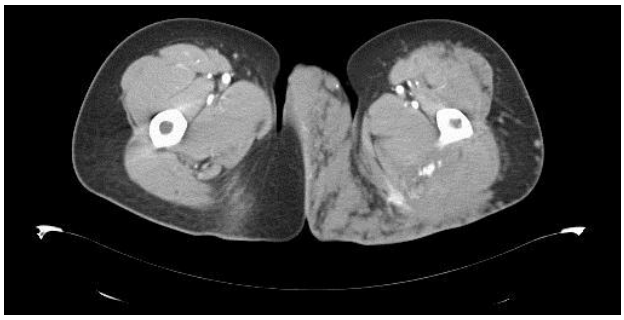

Figure 2: Pre-operative computed tomography angiogram revealed venous malformation over the left vulva and left thigh

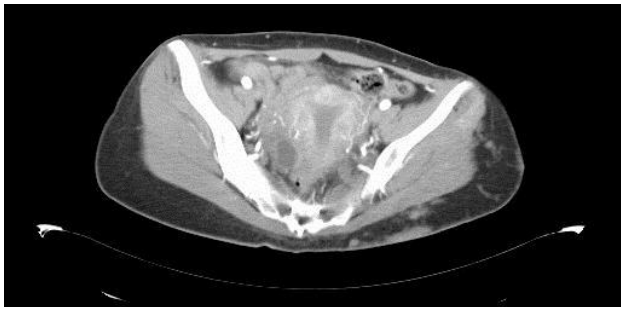

Figure 3: Pre-operative computed tomography revealed extensive venous malformation in the pelvis

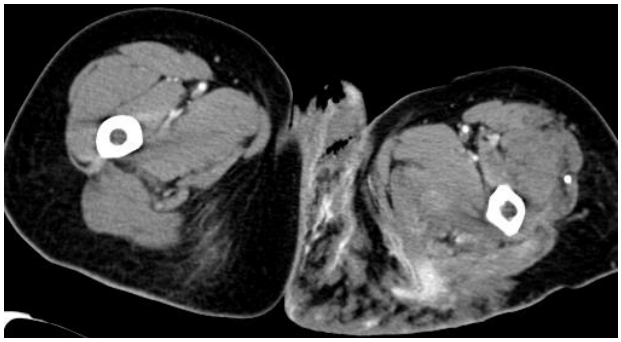

Figure 4: Left labium is filled with residual clot with no new vascular malformation

Figure 5: Post-op picture of the left labium revealed no recurrent swelling after 5 years

coagulopathy. The prothrombin time was prolonged at 19 seconds (11.4-14.2), international normalized ratio (INR) was 1.72 and activated partial thromboplastin time (APTT) was 48.9 seconds (31.3-46.1). Hemoglobin level dropped from 11.6 to 8.3 g/dL. She was transfused with 4 units of fresh frozen plasma and 1 unit of red blood cells. Multiple surgeries for hemostasis using argon plasma coagulation and application of underrunning sutures over the bleeding areas were performed. The wound was left open with vacuum dressing which subsequently healed via secondary intention. Computed tomography scan was done 1 month following surgery and revealed no recurrence at left labium (Figure 4). Follow up to 5-year period revealed no recurrent swelling at left labium (Figure 5).

Discussion

Congenital vascular malformations are rare, with reported incidence of around 1.4/100,000 per year. It can be further classified into slow-flow lesions that include capillary, lymphatic and venous malformations and high-flow lesions that include arterial malformations, arterio-venous fistula and arteriovenous malformations (1). Although mostly asymptomatic, its aesthetic appearance maybe disabling (2). The most common presenting features of this condition is pain and ecchymosis especially during menses or pregnancy. This is believed to be the sequelae of chronic consumptive coagulopathy secondary to blood stagnation within the venous malformation (3).

External genitalia venous malformations are frequently associated with lower limb involvement as observed in this current case (3). This condition can be treated symptomatically, with surgical excision, injection of sclerosant or a combination of multiple modalities (4). The aim of surgery in this patient is to improve cosmesis which contributes to a better psychosexual function. Surgery is often challenging due to the location of this VM involving the pelvic area. Although complete resection is pertinent in preventing recurrence, a 'radical' approach may be associated with large amounts of blood loss (5). This could be concurred as venous malformations are often associated with coagulopathy (5). Therefore, diathermy and energy

devices are used to facilitate surgery with minimal blood loss. There is lack of reported outcome pertaining energy device usage in VM resection.

Sclerotherapy is the latest therapeutic modality accepted as monotherapy, especially for surgically inaccessible lesions. It has also been implicated as an adjunct to surgery (4). Examples of sclerosants used are 80% ethanol, *N*-butyl cyanoacrylate (NBCA), polidocanol, sodium tetradecyl sulphate, coils and contour particles. Direct intralesional injection of sclerosing agents causes destruction of endothelial surfaces and subsequent intimal thrombosis, ultimately resulting in the obliteration of the lumen of the VM. Complications of sclerotherapy include severe pain, extensive tissue necrosis and nerve damage. Polidocanol and sodium tetradecyl sulphate are preferred compared to ethanol in small and superficial venous malformations and lesions close to major nerves, as ethanol usage has an increased risk of tissue necrosis and nerve damage. Sclerotherapy can be performed by clinical palpation or guided with colour duplex scan or venography (2). Direct injection can be used in small superficial lesions. However, in deep and large VMs, the injection of the sclerosants should be guided with imaging modalities. Direct injection and venography is more invasive and expensive compared to colour duplex. However, it has the advantage of adequate visualization of the lesion in its dilated state. This provides a road map for better planning of treatment. There have been reports of recanalization of VMs after sclerotherapy (5, 6). However, this treatment modality can be repeated safely or as per necessary, followed up with surgical resection (6).

In the current case, no angioembolization was performed as it was a venous malformation with no single feeding arterial supply. Sclerotherapy was not offered due to the concern of severe pain in the awkward location of the vulva which can be disastrous to the patient and the swelling is too large for sclerosant injection to be effective.

Conclusion

Surgical excision of the venous malformation is effective to clear the lesion at the vulva despite the challenges of extensive bleeding encountered. Though there was anticipation of bleeding with extra precaution performed (tissue glue, argon plasma coagulation and advanced bipolar energy device), the hemostasis in surgical excision of such lesion has remained challenging.

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Competing interests

The authors declare that they have no competing interests.

Informed Consent

Written informed consent was obtained from the patient for inclusion in this report. Research and ethics committee approval for case reports is not a requirement in Universiti Kebangsaan Malaysia Medical Centre (UKMMC).

References

1. Nosher JL, Murillo PG, Liszewski M, Gendel V, Gribbin CE. Vascular anomalies: A pictorial review of nomenclature, diagnosis and treatment. World J Radiol. 2014;6(9):677-92.
2. Revicky V, Maina W, Cockburn J, Stanley K. Vulval low-flow arteriovenous malformation. Arch Gynecol Obstet. 2009;280(2):271-3.
3. Enjolras O, Ciabrini D, Mazoyer E, Laurian C, Herbretau D. Extensive pure venous malformations in the upper or lower limb: A review of 27 cases. J Am Acad Dermatol. 1997;36:219-25.
4. Guex JJ, Allaert FA, Gillet JL, Chleir F. Immediate and midterm complications of sclerotherapy: report of a prospective multicenter registry of 12,173 sclerotherapy sessions. Dermatol Surg. 2005;31(2):123-8.
5. Lee BB, Do YS, Yakes W, Kim DI, Mattassi R, Hyon WS. Management of arteriovenous malformations: a multidisciplinary approach. J Vasc Surg. 2004;39(3):590-600.
6. Yamaki T, Nozaki M, Sasaki K. Color duplex-guided sclerotherapy for the treatment of venous malformations. Dermatol Surg. 2000;26(4):323-8.