Slow-Flow Venous Malformation of Vulva in a Pre-menarcheal Girl

Dear Editor,

Venous malformations (VMs) characteristically present as asymptomatic compressible soft tissue masses that can become swollen and painful with injury, activity, or changes in the hormonal milieu as in pregnancy or puberty [Table 1].[1-5] Vulval VMs account for about 1% of cases and are extremely rare in the pre-menarcheal age group. Herein, we report such a case.

An 8-year-old girl born to a non-consanguineous couple presented to our dermatology outpatient department with the chief complaints of a bluish swelling in the external genitalia noticed in the past 4 months, gradually progressing in size. It was associated with throbbing pain, aggravated by day-to-day activities such as straining for stools, cycling, or long hours of standing. There was no history of trauma at the site of the lesion. The mother gave a history of atrial septal defect at birth, which closed spontaneously. There was no history suggestive of sexual abuse. On clinical examination, a well-defined, bluish, soft, compressible, reducible swelling measuring 1.5 × 1 cm was present over the left labia, and a similar swelling of 0.5 cm in diameter was present over the right labia [Figure 1]. A cough impulse was present, and the swelling increased with the deepening of the bluish hue on the Valsalva maneuver. This process increases the intra-thoracic pressure, resulting in a decrease in venous return, dilating the abnormal venous channels in the body, thus confirming a VM.

With the above history and clinical features, we considered the possibility of vulval VM, vulval varicocele, and canal of Nuck hernia. The non- cystic consistency of the swelling rules out the possibility of a Bartholin's cyst. Similarly, a negative history of trauma excludes traumatic cysts. Ultrasound with Doppler study revealed a slow-flow VM in the left labia measuring 1.13 × 0.8 cm [Figure 2]—a similar evolving lesion in the right labia. The lesions revealed a monophasic pattern indicating a slow venous blood flow [Figure 3]. The two-dimensional echocardiogram was normal. Due to financial constraints, the patient's caregivers refused computed tomography and magnetic resonance imaging (MRI) scans. An informed consent was obtained from the parents of the patient. Following baseline blood investigations, sclerotherapy with bleomycin injection was the preferred treatment. Bleomycin was preferred considering its efficacy, easy availability, and available literature data that favor bleomycin in combination with triamcinolone

Table 1: Reported cases of vulvar VMs in the pre-menarcheal age group[1-5]

Study	Study period	Total Cases
Nassiri *et al*.	7 years	11
Wang *et al*.	10 years	8
Herman *et al*.	2004	Single, 11-year-old
Van der Woude DA *et al*	2011	Single, 13-year-old
Focseneanu MA *et al*.	2012	4
Present study	2022	Single, 8-year-old

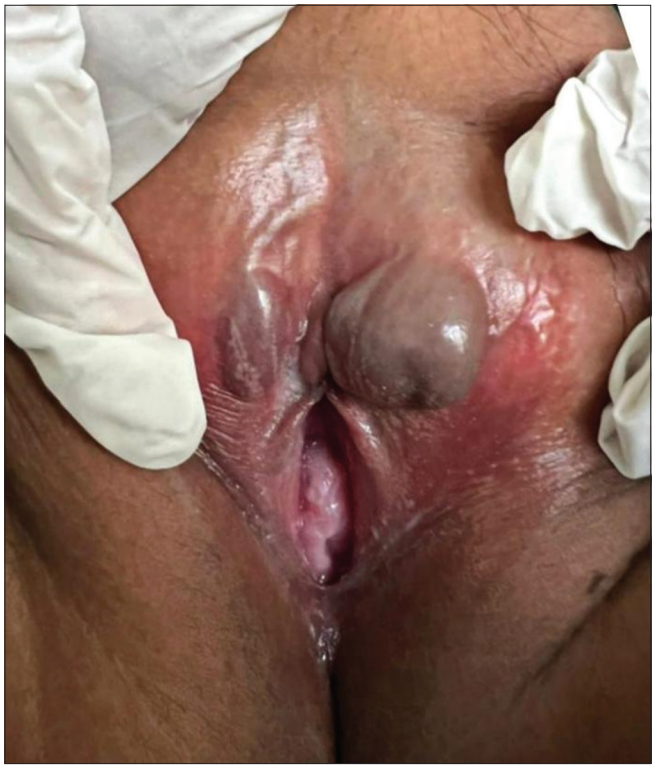

Figure 1: Bluish swellings over bilateral labia

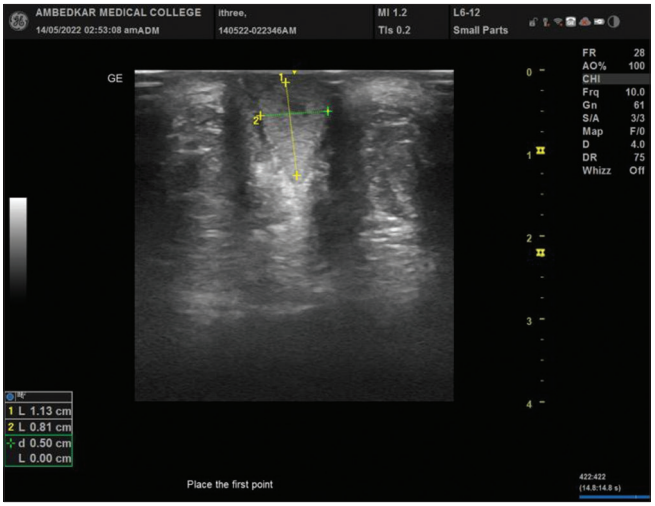

Figure 2: Ultrasonography of the swelling

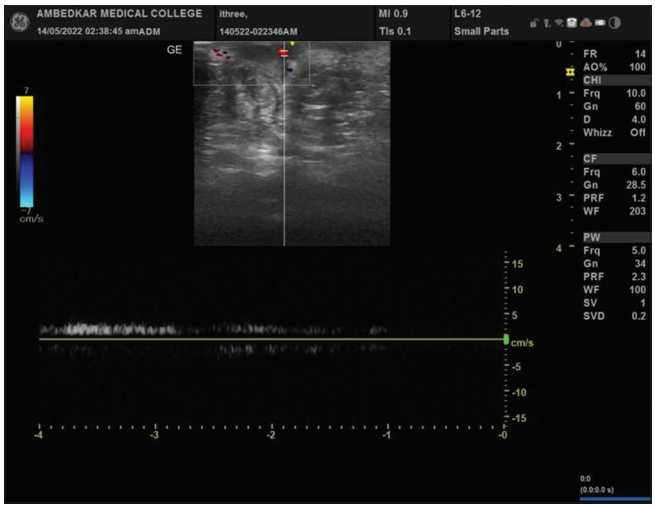

Figure 3: Doppler showing monophasic blood flow

and ethiodized oil with oral propanolol for pediatric hemangioma. Lignocaine 2% jelly was applied as a local anesthetic for 10 minutes prior to the injection. 15 IU of bleomycin was reconstituted with 5 mL of 0.9% sodium chloride and given intra-lesional using an insulin syringe at a dose of 0.3 mg per kilogram of body weight. Injection was stopped just at the point of blanching to prevent skin necrosis. Local digital pressure was applied for 15 minutes to prevent immediate washout of the sclerosant. Post-operative pain was minimal and was managed with paracetamol. Anti-inflammatory drugs can interfere with the process of sclerosis. A proximal vessel block was not required as it was a slow-flow lesion. Moreover, it is not possible for lesions involving the labia minora. After 3 months, 90% resolution was noticed [Figure 4]. A Doppler ultrasound was repeated before second sitting, but the sonologist could not distinguish between the normal labia and the vulval malformation. However, a small amount of bulge was clinically visible on the labia. A second dose resulted in complete resolution at 6 months [Figure 5].

VMs may occur sporadically. Familial cases have shown mutations in the gene that encodes for the tyrosine kinase domain of the endothelial cell receptor *TIE2* (tunica interna endothelial cell kinase) and somatic mutations in the angiopoietin receptor gene *TEK* (endothelial tyrosine kinase).[6] They are also associated with specific genetic syndromes such as capillary malformations – arteriovenous malformation syndrome (CM-AVM), Klippel–Trénaunay–Weber syndrome (KTWS), hereditary hemorrhagic telangiectasia (HHHT), Rendu–Osler–Weber syndrome, and hereditary benign telangiectasia (HBT).[7] Vulvar VMs are usually asymptomatic and may go unnoticed for years. A thorough history and clinical examination aided by imaging studies helps in early diagnosis and further management. MRI is useful in determining the extent of involvement, while ultrasonography studies

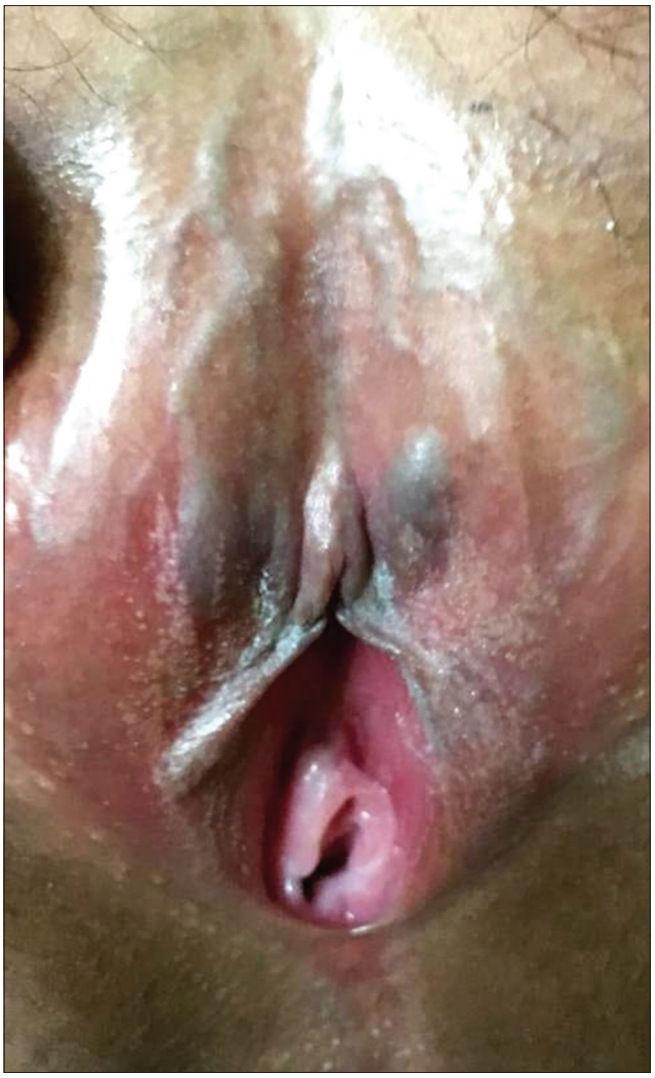

Figure 4: 90% resolution after 3 months of sclerotherapy

reveal hypo-echoic and heterogeneous lesions that help in diagnosis. Color Doppler analysis indicates a monophasic or biphasic flow, thus respectively differentiating between low- and high-flow malformations.[2] In the current case, a hypo-echoic lesion was visible on ultrasonography, with a monophasic blood flow pattern on color Doppler, consistent with low-flow VM.

The treatment of large and diffuse arteriovenous malformations is embolization, while sclerotherapy is preferred for small localized lesions. Surgical excision is curative.[4] As these are benign lesions, treatment is usually reserved for symptomatic and progressive cases.[2] The peri-clitoral location of the lesion in our patient prompted us to use sclerotherapy. Sclerotherapy is injecting a sclerosing agent into the lesion directly, which destroys the endothelium and subsequent obliteration of the lumen.[3]

Bleomycin is a chemotherapeutic agent with a specific sclerosing effect on the vascular endothelium. The

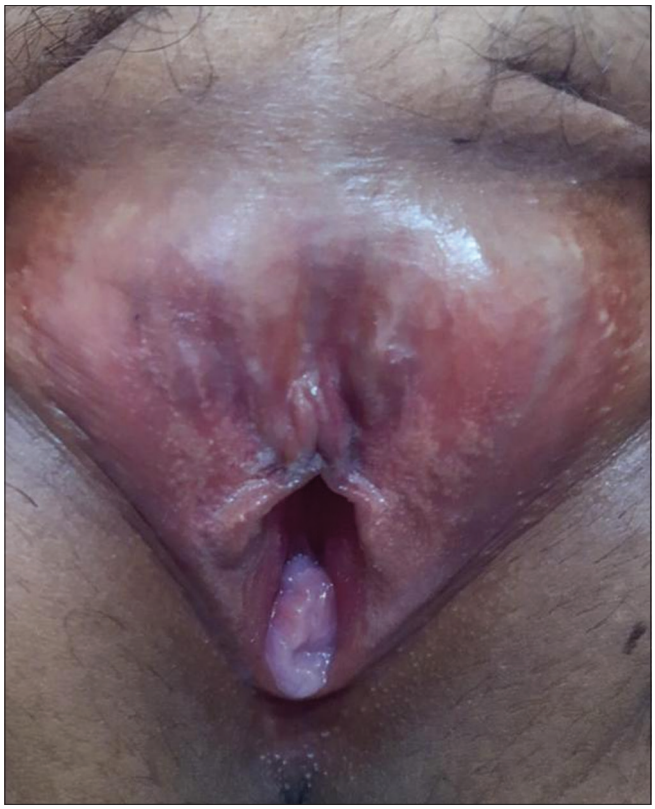

Figure 5: Complete resolution after 6 months of sclerotherapy

efficacy of bleomycin is comparable with other sclerosing agents, such as ethanol, but with fewer complications. Its adverse effects include injection site pain, tissue necrosis, pulmonary fibrosis, and idiosyncratic reactions. However, pulmonary fibrosis as a complication after sclerotherapy has never been reported as it is dose-dependent.[8,9] Recurrence with bleomycin has also been reported in the literature. Nevesny *et al*.[9] reported three cases of recurrences after 2 years of percutaneous sclerotherapy with bleomycin. Abdelaty *et al*.[10] detected recurrences in three cases out of the 50 cases of slow-flow VMs which were managed with intralesional injection of bleomycin. However, Mohan AT *et al*.[11] managed 32 cases of low-flow vascular malformations in children with intra-lesional bleomycin and reported no recurrences.

Pain following the procedure was treated with oral analgesics. The patient had minimal discomfort, and the lesions resolved after two sittings of sclerotherapy 3 months apart.

In conclusion, vulvar VM presents as vulvar swellings in the pre-menarcheal group. Sclerotherapy is a quick and effective treatment modality for managing VM with minimal adverse events and recovery time.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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