

## Vulval Mixed Macrocystic and Microcystic Lymphatic Malformation in an Adolescent Girl Showing Cyclical Changes with Menstrual Cycles

Sir,

Lymphatic malformations (LM) are non-malignant tumours of lymphatics which can be primary (congenital) or secondary (acquired). Congenital LM usually present in early childhood (up to 2 years of age). There are very few cases of congenital LM of vulva presenting in adults.<sup>[1]</sup> We report a 19-year-old adolescent female who presented with a swelling in the vulva which was histopathologically diagnosed as mixed cystic LM.

A 19-year-old female presented to us with a history of swelling on the left side of vulva for the past 4 months which had a waxing and waning course. The swelling appeared after menstrual cycles 4 months prior to presentation, but gradually regressed to about half of the original size but again after 2 months it increased in size during pre-menstrual period [Figures 1 and 2]. The lesion was asymptomatic except for a dragging sensation. She had no history of trauma or ulceration over the genitalia or any abdominal pain or lump. There was no menstrual irregularity or menorrhagia. She was unmarried and sexually inactive. There were no associated medical, surgical, or gynaecological complaints. She had attained menarche at 16 years of age. On examination of the genitalia, there was a pendulous cystic lesion of size 6 cm × 3 cm × 3 cm arising from the left side at the

junction of labia majora and minora arising by a broad base. The swelling was of bluish tinge, translucent, cystic, and compressible [Figure 1]. Anterior to the base of the growth there were a few cystic to firm papular lesions. No healed scar was seen. About 3 mL of gelatinous fluid was aspirated. On ultrasound of the abdomen and pelvis, there was no organomegaly--the uterus and other pelvic organs were normal. Biopsy was taken from the base of the lesion which showed marked dilation of several lymphatics in papillary dermis and reticular dermis [Figure 3]. So, a final diagnosis of mixed macrocystic and microcystic LM of vulva was made. Immunohistochemistry for estrogen and progesterone receptors was positive. The patient came for follow-up before her next menstrual cycles and about 4 days following cessation of menstrual flow for two subsequent months. Decreased size of swelling was seen after menstrual cycles which increased again premenstrually. As the macrocystic part of LM was showing spontaneous regression, we planned to wait for 6 months and counselled her about the benign nature of the disease. The patient was advised regular follow-up.

Congenital lymphatic malformations (CLM) are due to developmental disorder of lymphatics which may be due to failure of foetal lymph vessels to involute and/or absence of lymphatics getting connected with the central lymphatic system. International Society for



Figure 1: Swelling on the left side of the vulva showing macrocystic and microcystic lymphatic malformations

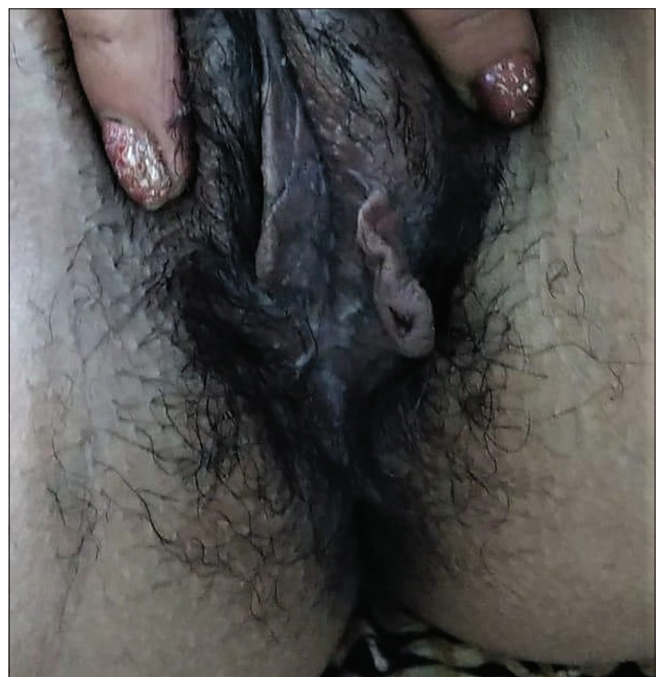
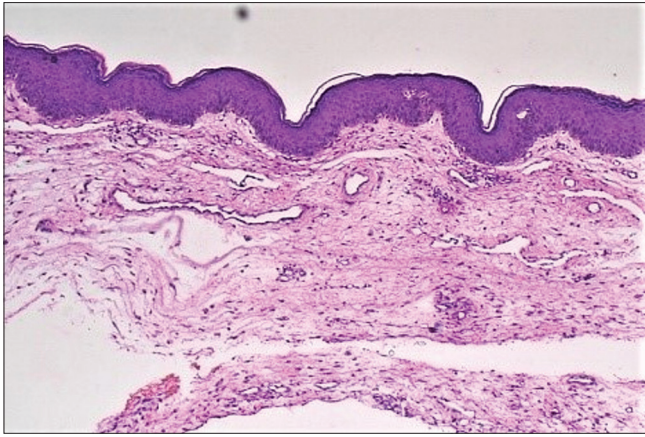


Figure 2: Regression of the size of macrocystic lymphatic malformations while the microcystic ones are persisting



**Figure 3: Histopathology scanner view showing marked dilation of several lymphatics in papillary and reticular dermis (H and E X 40)**

Study of Vascular Anomalies (ISVVA) classified cystic LM into three types: macrocystic LM, microcystic LM, and mixed cystic LM.<sup>[2]</sup> Macrocystic variant has cysts larger than 2 cm, microcystic type has no cysts or has small ones (<2 cm) and mixed variant has both large and small cysts. Mixed LM are the commonest. Macrocystic LM are least common, and clinically, the lesions appear as soft, translucent, bluish-coloured cystic masses which are compressible. Superficial small cysts of microcystic lesions may give a warty appearance. Less than ten cases of macrocystic lymphangioma of vulva have been reported.<sup>[1,3]</sup> Macrocystic LM are usually asymptomatic or become symptomatic due to the mass effect. Histopathologically, the dilated lymphatics are more in papillary and reticular dermis in microcystic and macrocystic variant, respectively.

Regression of macrocystic LM of internal organs has been reported following decreased progesterone levels in other studies.<sup>[4,5]</sup> Similarly, our patient had a premenstrual increase in the size of LM with regression later. The presence of estrogen and progesterone receptors on the lymphatics are responsible for this manifestation. Adult onset LM presenting in pregnancy further support the role of hormones in dilatation of lymphatics.

Alteration of balance of inflow and outflow leads to dilatation of lymphatics. In patients with intact outflow, a sudden increase in inflow leads to appearance of dilatation, which slowly resolves due to the normal outflow. This leads to spontaneous regression of lesions.<sup>[6]</sup> In our patient also with increased progesterone premenstrually, there was a sudden increase of inflow, which reduced with decreased hormone levels and unaffected outflow lead to spontaneous regression but the microcystic component persisted. The main therapeutic options include percutaneous drainage, surgical excision or radiofrequency ablation, and sclerotherapy.

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## Conflicts of interest

There are no conflicts of interest.

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