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Primary vulvar lymphangioma and VAIN in patient with systematic lupus erythematosus

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INTRODUCTION AND CASE PRESENTATION

Lymphangiomas are rare benign proliferations of the lymphatic system. Lymphangioma is considered either primary or congenital if it is present at birth or if it develops in early childhood or secondary or acquired if it is caused by impaired lymph flow [1, 2]. However, the primary vulvar lymphangioma occurs mostly in adulthood. Secondary lymphangioma is also termed acquired lymphangioma and lymphangiectasis. Vulvar involvement by lymphangioma is rare [3–5].

We present a new case of vulvar lymphangioma associated with vaginal intraepithelial neoplasia (VAIN) in a 32-year-old nullipara with diagnosis of systemic lupus erythematosus (SLE) from 2002. Patient was hospitalized multiple time because of aggravation of SLE (renal insufficiency, osteoporosis, neck of femur fracture, lumbar fracture, severe pneumonia). Since 2017 she has observed swollen labia minora of both sides, after punch biopsy of vulva final diagnosis of lymphangioma was given. In differential diagnosis, condyloma, lichen, vulvar intraepithelial neoplasia and vulvar cancer was excluded. For three years local treatment was prescribed without improvement. The patient complained of intolerance of vaginal globules or creams with estrogen, local steroids or hyaluronic acid. After such treatment she observed enlargement of labial oedema and irritation. Additionally in Pap smear result in 2019 low intra epithelial lesion (LSIL) occurred and in 2020 high grade squamous intra epithelial lesion (ASC-H) occurred. Human papilloma virus (HPV) DNA status was negative. Vaginal examination with speculum was difficult and painful because of adhesions of vaginal walls. Under general anesthesia cervical biopsy revealed no signs of intraepithelial dysplasia, p16 negative, however punch biopsy of vaginal walls (adhesions) revealed low intraepithelial lesion, p16 negative, but reactive changes cannot be excluded. Patient was married, however not sexually active because of painful intercourse, because of described lesions. She had regular cycles. The patient decided for surgical treatment of enlarged labia minora. During the procedure a labioplasty was performed with excision of vulvar adhesions and with cervical biopsy. In result vulvar lymphedema was described with no signs of cervical dysplasia, although with suspicion of VAIN II/low grade squamous intraepithelial lesion, p16 negative. Shortly after the procedure she was instructed to use vaginal dilators to prevent recurrence of adhesions. (Fig. 1) presents patients labia before and after surgery) After procedure vulvar pain and oedema was relieved. Patient self-esteem and mood improved, although in the first year after surgery she was still not sexually active because she felt strong anxiety related to penetration. Vaginal examination with speculum was accessible and not painful to the patient. The patient remains in follow-up for 3 years after surgery without recurrence of lymphangioma, she is sexually active, although still suffers from adhesion of upper part of vagina.

DISCUSSION

The etiologic factors for primary vulvar are not unknown. It is supposed to occur from local, closed, sequestrated lymphatic cisterns in the reticular dermis. They consist of muscular lining and connect with superficial lymphatics via dilated channels. These muscular lymphatic cisterns pulsate and transmit pressure, secondary to which dilatation of

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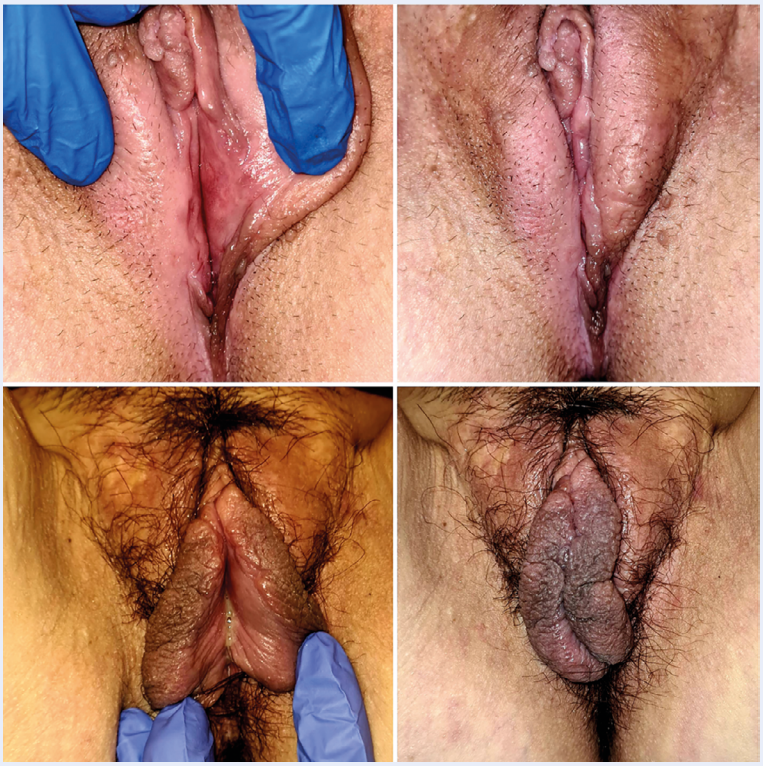

Figure 1. Appearance before and 6 months after surgery

lymphatics occurs in dermal papillae visible as vesicles externally. The probable etiology of secondary vulvar is the architectural disruption of previously normal channels. There is no standard treatment of choice described for the management of vulvar LC. Depending on the type and extent of lymphedema, the treatment options include procedures such as observation, abrasive modalities, superficial radiotherapy and surgical excision. Even after surgical treatment recurrence is common, up to 50% of patients [1–5].

Article information and declarations

Ethics statement

Written informed consent for publication was obtained from the patient.

Author contributions

Marta Balajewicz-Nowak — 80% study conception and design, manuscript preparation, Bartlomiej Galarowicz — 20% data collection.

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None.

Conflict of interest

Authors declare no conflict of interest.

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