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Uncommon chronic lymphedema leading to vulvar elephantiasis in an Indian woman: A case report

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Abstract

Elephantiasis, a chronic manifestation of lymphatic filariasis, primarily affects the limbs, scrotum, and trunk, with a lower incidence observed in females. Vulvar elephantiasis is an even rarer presentation, complicating diagnosis due to its uncommon site and the absence of identifiable parasites in tissue samples. Histopathological analysis alone is often inconclusive, as microfilariae are rarely seen in tissue sections. Therefore, a definitive diagnosis relies on identifying microfilariae in nocturnal blood samples or confirming seropositivity for filarial antigens, underscoring the importance of comprehensive diagnostic approaches in such rare cases.

Keywords: Labial; Hypertrophy; Enlarged vulva; Labial elephantiasis; Köbberling-dunnigan syndrome; Crohn's disease of the vulva.

Introduction

Vulvar elephantiasis is an extremely rare condition, often resulting from chronic filarial infection, which typically affects limbs but can also involve the scrotum and, more rarely, the vulva. The condition is marked by extreme hypertrophy of the affected tissue due to chronic lymphedema. Vulvar involvement is less common and presents diagnostic challenges due to its rarity and clinical overlap with other conditions, such as Bartholin's gland cysts, which can initially mask the diagnosis [1]. Chronic inflammation and lymphatic obstruction contribute significantly to the pathogenesis of elephantiasis, with females being affected less frequently than males due to anatomical and immunological factors [2].

Case presentation

A 37-year-old female presented to the Shalya Tantra OPD at Mahatma Gandhi Ayurveda College Hospital and Research Centre, Salod, Wardha, with complaints of extreme swelling in the vulvar region, accompanied by mild itching but no pain or tenderness. The patient also reported a disturbed menstrual cycle. Her history revealed no significant family or medical factors related to similar conditions. Physical examination showed marked labial hypertrophy, which prompted consideration of differential diagnoses including labial elephantiasis, Köbberling-Dunnigan syndrome, and Crohn's disease of the vulva. Laboratory and imaging evaluations, including ultrasonography, were conducted to assess lymphatic involvement and rule out alternative causes.

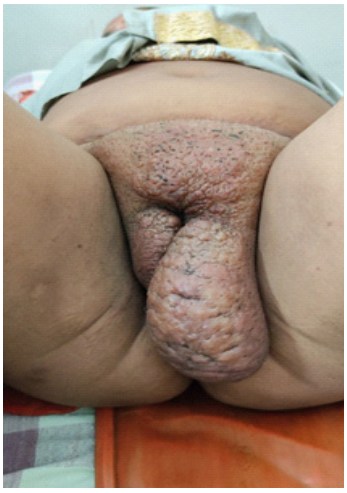

Figure 1: Hypertrophy of labia.

Discussion

Vulvar elephantiasis, particularly in cases with no direct history of filarial infection, requires an in-depth examination of inflammatory and lymphatic health factors. Risk factors for vulvar elephantiasis include chronic inflammatory conditions, immunosuppressive factors, and occasionally genetic predispositions [3]. This case highlights the clinical ambiguity of vulvar elephantiasis, as initial presentations often resemble benign cystic conditions, resulting in delayed diagnosis. The patient's occupation may have contributed to prolonged UV exposure, a known risk factor in some lymphoproliferative conditions [4]. Surgical management remains the primary treatment for symptomatic relief, as conservative approaches often yield limited improvement due to the fibrotic nature of the tissue [5].

Conclusion

This case underscores the importance of considering vulvar elephantiasis in differential diagnoses of labial hypertrophy, especially in patients with chronic swelling. Early and accurate diagnosis is vital for effective management, often necessitating surgical intervention for optimal outcomes.

Differential diagnosis: Labial elephantiasis, Köbberling-Dunnigan syndrome, Crohn's disease of the vulva.

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