

A rare case of elephantosis nostras verrucosa

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Abstract— Elephantiasis nostras verrucosa (ENV) is a rare condition that is characterised by hyperkeratosis, fibrosis and disfiguration of dermis. It is caused mostly by non infectious diseases such as surgery, trauma, tumors, and venous obstructions. There are very few report of ENV in Middle East region and her we present one case about it.

Index Terms— elephantosis nostras verrucosa, surgery, elephantiasis, venous stasis, depp venous thrombosis, skin, lymphedema.

1 INTRODUCTION

Elephantiasis nostras verrucosa (ENV) is a rare disease that is believed to be caused by chronic standing lymphedema, which lead to cutaneous hypertrophy. It can usually affect the lower because of the gravity-dependent blood flow. There are very few report of ENV in the middle east and her we report one case.

2 THE CASE

62 years old, Saudi men presented to our clinic with complain of sever swelling in the right and left foot that is slowly progressive over period of few years. The patient complain started with developing of lymphedema in both legs. He had a history of DVT in the right leg before that . Over time the patient foots start showing deformity and disfiguration. The patient denied any history of trauma, surgery radiation. He is a known cause of diabetes and he is currently using sitagliptin, glicazide and metformin. He is also taking Rivaroxaban for risk of DVT

On physical examination there was a severely enlarged left and right foot reaching the level of the mid leg with no odor. Diffuse thickening of both feet and leg with non pitting edema and cobblestone like skin surface. Posterior tibial and doraslis pedis pulse were not palpable.

Laboratory test showed normal CBC, liver and renal function. Except for glucose random 348mg/d but the patient is a known case of diabetes

Skin biopsy showed hyperkeratosis, parakeratosis and acanthosis of the epidermis. KOH and acid shiff was negative. leg x ray of the lower extremities showed diffuse soft tissue swelling (figure1-2).

According to these finding ENV was diagnosed. The patient was given acitretin to control the hyperkeratosis. Potassium permanganate. Unfortunalty the patient did not come back for follow up.

3 DISCUSION

Elephantiasis nostras verrucosa is a rare, disfiguring complication of longstanding chronic lymphedema The pathogenesis of ENV is still unclear. But it thought that first the lymphatic channels are damaged and blocked, and protein-rich fluid accumulates in the dermis and subcutaneous

tissues which will cause decreases oxygen tension and might decrease the immunity of the skin. After that, the poor immunity increases the skin's susceptibility to infection. Finally, there will be swelling, fibrosis, and disfiguration of the affected areasⁱ. Hence, a vicious cycle begins.

The presentation of Elephantiasis nostras verrucosa include a grossly enlargement and disfiguration of the lower extremities and the feet, with a cobblestone or mossy appearance. Because of the special nature of the edema the skin usually feels "woody," because the edema is nonpitting, and does not resolve upon elevation of the extremity. The initial signs of this condition include a cobblestone appearance which progresses to a verrucous and mossy appearance and over time will ulcerate and provide a favorable environment for the growth of bacteria or fungiⁱⁱ.

The diagnosis can be made based on the history and physical examination. Imaging studies are not necessary for the diagnosis. ENV carry a high risk of malignancy so skin biopsy is Important in those cases ⁱⁱⁱ. Characteristic histological findings include pseudoepitheliomatous hyperplasia, dilated lymphatic channels, widened tissue spaces, and extensive fibrous tissue hyperplasia within the dermis, subcutaneous tissue, and lymphatic vessel walls^{iv}.

Elephantiasis nostras verrucosa have a very long treatment course which is sometime unsuccessful because there is no standard treatment the clinician is left with a handful of case reports and literature reviews to guide management. the first step in the management should include management of any underlying cause of lymphedema. To reduce the lymphedema several methods are available such as, massage, compression stockings, medical bandages, mechanical massages^{v vi}.external sequential pneumatic compression devices reported a good Success rate^{vii}. More complex form of decongestive lymphatic therapy, includes skin hygiene, limb compression, and exercise is also effective.^{viii}

Pharmacological drugs can play a role in the treatment. Topical retinoid can be used to control the hyperkeratosis.^{ix x} Surgical debridement can be used in advance cases not responded well to conservative or medical treatment^{xi xii xiii xiv}. In the refractory cases with no good response to treatment limb amputation can be considered as last choice.

Early diagnosis and early intervention in the vicious cycle will have a better outcome.



Figure 1. X ray of the left and right foot showing diffuse soft tissue edema



Figure 2. X ray of the right leg AP view showing diffuse soft tissue edema

4 CONCLUSION

ENV have many possible etiologies and finding the underlying cause play a major role in the management plan. The key to diagnose EVN is history taking and physical examination. Laboratory tests and imaging studies may help in excluding other differential diagnosis. The treatment includes management of the underlying causes of lymphatic obstruction and prevention of the complications associated with lymphedema

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