An onerous knot untied: Morbihan disease- Report of two cases

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Introduction

Morbihan disease or rosaceous lymphedema is characterized by persistent erythematous lymphedema of upper two-thirds of face.[1] It is believed to be a complication of rosacea and may occur at any stage of the disease. Most of the patients remain asymptomatic, however, few may have disfigurement of facial contours, with subsequent visual field narrowing. The disease has variable therapeutic response with refractoriness to treatment. We present two challenging cases of Morbihan which were treated effectively.

Case report

- First case was a 46 year old male who presented with firm to hard swelling and erythema around forehead, eyes, lips, nose and cheeks since 2 years associated with difficulty in eye opening. Telangiectasia were present over nose and finger insinuation test was positive [Fig. A]. Second case was a 40 year male who presented with a firm swelling around bilateral eyes and lips since 3 months [Fig. B].
- Dermoscopy revealed multiple ill defined erythematous areas with follicular prominences with dilated blood vessels in eye lids[Fig. C]. On histopathology moderate inflammatory infiltrate composed of lymphocytes and mast cells was present around the blood vessels with pandermal edema and presence of telangiectatic capillaries with no granulomas in both the biopsies[. Immunohistochemistry with CD117 was positive suggestive of presence of mast cells[Fig.D-G]. Rest of the laboratory investigations were within normal limits. A diagnosis of Morbihan disease was made in both the cases. First patient was started on 20 mg isotretinoin with antihistamines with 50% improvement after 2 months and second case was started on oral prednisolone 40 mg with ketotifen. Later prednisolone was slowly tapered and patient was started on isotretinoin with 70%-80% improvement at 6 months of treatment [Fig. H,I].

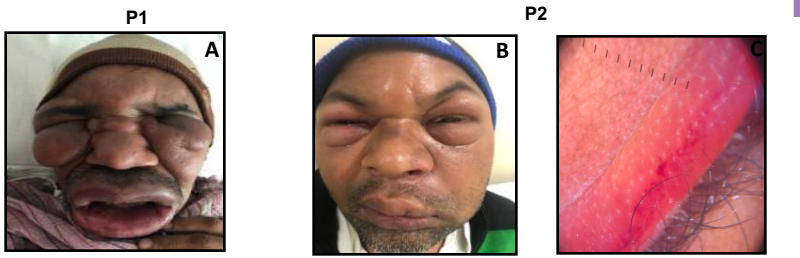
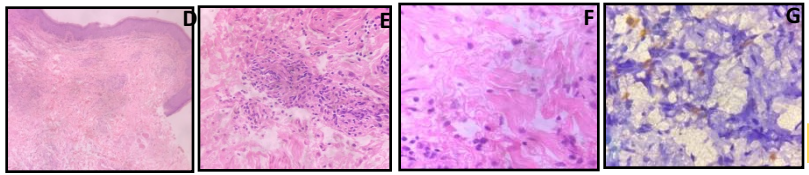


Figure [A,B]: Pretreatment photograph of patient 1 and 2 (P1 and P2) showing woody edema of upper two- thirds of face [C]: Dermoscopy P2: Follicular prominences with dilated vessels present



Figure[D-G]: D: Well defined collection of inflammatory infiltrate present in dermis (100X)
E: Infiltrate composed of lymphocytes and mast cells present around blood vessels (400X)
F:Magnified view showing mast cells (400X)
G: CD117 staining positive (brown) for mast cells (1000X)

Figure[H,I]: Post treatment: P1- Decrease in edema by 50% at 2 months and P2- Decrease in erythema and edema by 70-80% at 6 months

Discussion

Morbihan disease is generally regarded as a late-stage complication of rosacea. It presents as chronic persistent erythema and edema of the upper one- third of the face, with accentuation in the periorbital tissues, forehead, glabella, nose, and cheeks.[2] Involvement of lips was present in both the cases. Morbihan disease need to be differentiated from angioedema and other conditions presenting as pseudoangioedema like granulomatous cheilitis, dermatomyositis, DRESS syndrome, Allergic contact dermatitis. On histopathology, morbihan is characterized by perivascular dermal edema with a lymphohistiocytic periadenexal infiltrate containing numerous mast cells and dilation of lymphatic vessels. Granulomas are rarely present, and sebaceous gland hyperplasia can be observed in patients who have or had associated rosacea.[3]

Conclusions

Dermatopathology is extremely important to differentiate morbihan from other mimicking conditions so that early and appropriate treatment can be started for the same.

References

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