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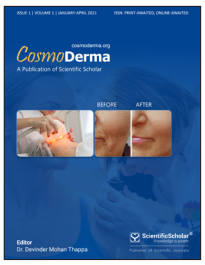

Spot the Diagnosis

Big lips and crooked smile in a young female

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Quick Response Code:

A 24-year-old female presented to us for the evaluation of persistent painless swelling of the upper lip and adjoining perioral region for eight months. She reported a gradual progression in the size and extent of the swelling. She denied any history of loose stools, abdominal cramps, difficulty breathing, or coexisting skin lesions. On further probing, she reported recurrent episodes of facial palsy in the past year. On clinical examination, diffuse erythematous, soft-to-firm swelling involving her upper lip, perioral region, and upper gingivae was evident [Figure 1a]. Longitudinal fissuring over the dorsum of the tongue was observed with grooves arranged transversely along the dorsum [Figure 1b]. There was the absence of forehead wrinkling over the right side, inability to lift the right brow, and deviation of the angle of the mouth toward the left, indicative of lower motor neuron facial nerve palsy [Figure 1a]. Routine hematological and biochemical investigations, fecal calprotectin, as well as a chest radiograph did not reveal any abnormality. The patient was treated with monthly intralesional triamcinolone acetonide 10 mg/mL injections along with weekly oral azithromycin 500 mg pulse therapy.

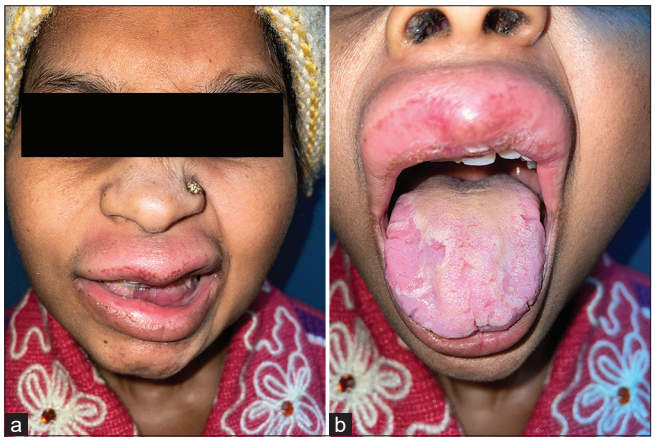

Figure 1: (a) Diffuse erythematous, soft-to-firm swelling involving the upper lip, perioral region, and lower gingivae. The absence of forehead wrinkling over the right side, inability to lift the right brow, and deviation of the angle of mouth toward the left indicate lower motor neuron facial nerve palsy. (b) Longitudinal fissuring over the dorsum of the tongue was observed with grooves arranged transversely along the dorsum.

WHAT IS THE DIAGNOSIS?

Answer:

Melkersson-Rosenthal syndrome (MRS).

DISCUSSION

The MRS is an idiopathic neuromucocutaneous syndrome, presenting with a typical clinical triad of the fissured tongue, orofacial swelling, and recurrent facial nerve paralysis.[1] The presence of two or all of these features is pivotal for a clinical diagnosis of MRS.[2] Granulomatous cheilitis (GC) is a chronic inflammatory condition causing persistent lip swelling and can occur in isolation or as part of MRS.[1] Monosymptomatic MRS, or Miescher's cheilitis, refers specifically to cases where only the lip swelling (GC) is present without the other symptoms of MRS.[1] Etiology remains elusive, with multiple factors currently implicated, such as viral infections, Down syndrome, inflammatory bowel disease, sarcoidosis, and mutations in fatty acid transporters.[3,4] Facial palsy is the initial presenting feature in 30–90% of the cases, with unilateral or bilateral involvement.[3] Accompanying orofacial swelling typically involves the eyelids, lips, tongue, buccal mucosa, and larynx.[3] The swelling is non-pitting and non-tender and may resolve partially or entirely with fibrosis, leading to disfigurement and functional impairment.[3] Biopsy is indicated in cases with persistent painful swelling non-responsive to standard treatments. It may show the presence of perivascular infiltrate of lymphocytes and plasma cells, late cases may show scattered non-caseating granulomas with Langhans giant cells. Lingua plicata or fissured tongue appears clinically as longitudinal grooves over the dorsum of the tongue, which should be by definition at least 2 mm deep and 15 mm long.[4] The complete triad of MRS is seldom fully apparent initially, with signs developing gradually over months to years.[4] A myriad of neurological manifestations, such as facial paresthesias, hypogeusia, tinnitus, visual disturbances, and cranial nerve palsies, may be present in MRS in addition to the triad.[1] Treatment modalities include oral steroids, intralesional steroids, immunosuppressive agents, and anti-tumor necrosis factor agents such as infliximab and tetracycline.[5] Recurrent facial palsies necessitate surgical management, which comprises standard mastoidectomy and decompression of the facial nerve.[5]

CONCLUSION

We present an exciting case manifesting the characteristic features of MRS in a young female, which can be misdiagnosed owing to the lack of awareness and the absence of all three classical features at presentation.

Ethical approval

The Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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