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A case of prolonged restricted mouth opening-An approach to the diagnosis

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Abstract

Restriction of mouth opening can be due to a number of reasons; the connective tissue disorders are most rare of findings and are often ignored while marking differential diagnosis. Here we present an approach to diagnosis for a case of prolonged restricted mouth opening and unearthing a rare cause i.e. oral mucosal morphea, which is a localised form of scleroderma affecting only skin and subcutaneous tissue restricted only to oral mucosa as a cause of restricted mouth opening such a cause for this presentation has rarely been reported in the literature; nevertheless this should always be considered when there is a prolonged restriction in mouth opening without an obvious cause.

Keywords: Restricted mouth opening, morphea, scleroderma, oral submucous fibrosis.

Introduction to Oral mucosal morphea

Morphea, also known as localized scleroderma, is a clinically distinct inflammatory disease, primarily affecting the dermis and subcutaneous fat. It is characterized by excessive collagen deposition leading to thickening of the dermis and/or subcutaneous tissues,. The etiology of morphea remains elusive. Multiple mechanisms have been implicated, such as autoimmunity, infection, drugs, radiation and microchimerism. Clinically, various types of morphea are distinguished, such as plaque morphea, bullous morphea, deep morphea and linear morphea. The latter also encompasses morphea en coup de sabre and Parry-Romberg syndrome affecting the scalp and face with potentially devastating complications. Whilst in almost all cases the sclerosing process exclusively affects the skin, there are anecdotal cases in which associated mucosal involvement has been described .We here describe a unique variant of morphea affecting only the oral mucosa with significant local tissue damage and no evidence of cutaneous sclerosis

Morphea is a localized form of scleroderma which affects the skin and underlying structures it can be differentiated

from systemic sclerosis by absence of the sclerodactyly, Raynaud phenomenon, deep organ involvement and nailfold capillary changes[1]not much is clear regarding the aetiopathogenesis of this disorder but there seems to be an imbalance in the production and destruction of collagen which ultimately leads to the thickening of the skin ultimately leading to a scar-like sclerosis[2]. A number of mechanisms have been implicated such as trauma,[6] infections (borrelia spp.), medications[7] (bisoprolol, bleomycin, peplomycin, bromocriptine, carbidopa etc.) , auto immunity[8,9] and microchimerism [1,3].Here we describe a unique variant of morphea affecting only oral mucosa no such case has been reported in the literature to the best of authors knowledge.

Case report

- A fourteen year old male presented to the department of oral and maxillofacial surgery PGIDS, Rohtak with a complaint of severely restricted mouth opening. (figure 1)On further questioning about his complaint he gave history of progressively decreasing mouth opening since the age of four. According to the patient there had been numerous episodes of painful blister formation in his mouth which subsequently healed with hardening of oral mucosa. There was no positive history of tobacco or areca nut usage though the personal hygiene of the patient was dire. The patient gave no positive history regarding the Raynaud's phenomenon or dry eyes or dry mouth the medical history of the patient was non suggestive. A through physical and local examination was done which revealed that there were focal areas of hypo pigmentation on the face severely restricted mouth opening of about 5mm the intraoral examination could not be performed due to lack of access but the maxillary and mandibular labial vestibules showed presence of thick fibrous bands with mucosal contractures the dentition was crowded with generalized gingival recession of the anterior teeth his saliva was thick and tenacious. General physical examination was unremarkable and ultrasonography of the abdomen reveled normal echo texture of the abdominal viscera. His symptoms and history raised a possibility of bechet's syndrome so a pathergy test was performed which was negative. Antinuclear antigen titers were obtained which were highly raised indicating toward the possibility of an autoimmune disorder; therefore a dermatology opinion was taken which highlighted the possibility of oral mucosal morphea. The panoramic radiograph of the patient revealed horizontal bone loss of the alveolar bone. The patient was scheduled for surgery after the completion of preanaesthetic checkup and bilateral release of the sub mucosal fibrous bands was done up to the retro molar trigone and full depth mucosal biopsy was made but due to long standing nature of disease only 15 mm of mouth opening was achieved therefore bilateral coronoidectomy was performed through intraoral approach and mouth opening of 26 mm(Figure 2) was obtained the raw mucosal surface was to be covered by buccal fat pad but it was also found to be fibrosed therefore it was covered instead with full thickness skin graft adapted with bolster dressing for 5 days(figure 3) thereafter active mouth opening exercises were started and intralesional corticosteroid therapy (kenacort 40mg/ml)on each side was started on weekly basis mouth opening improved to 29 mm on subsequent follow ups(Figure4, 5) and the oral prophylaxis was performed but the alveolar mucosa continues to show fibrosis ; the tongue which was observed upon mouth opening revealed limited range of motion and smooth shiny mucosa and rubbery hard consistency upon palpation.

Discussion

Scleroderma is a group of chronic autoimmune diseases with skin thickening as the hallmark of the disease.

Morphea or localised scleroderma is a rare fibrosing disorder of the skin and underlying tissues, with equal prevalence in adults and children, that is a distinct and separate disease entity from systemic sclerosis.[4]

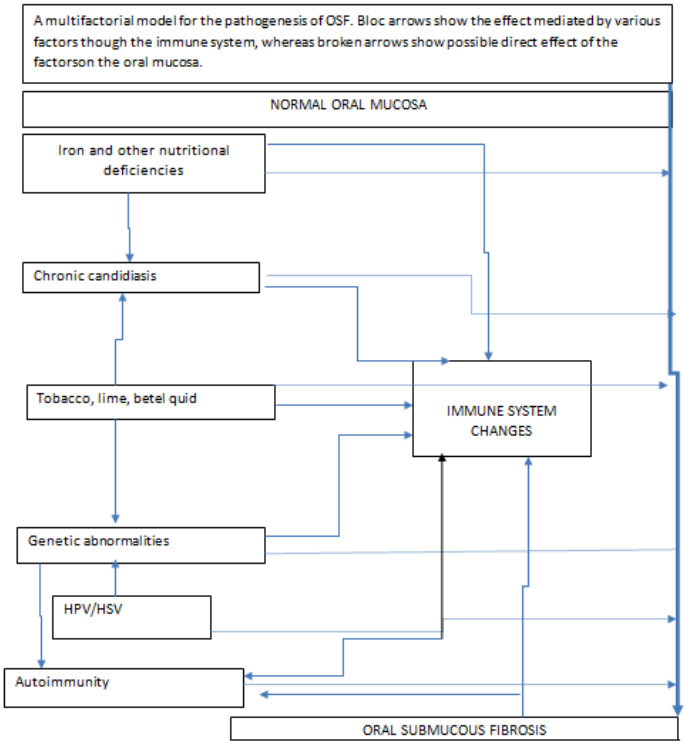
Morphea can cause sclerosis of all mesoderm-derived tissues and, rarely,

central nervous system[5]. Morphea has historically been classified based on clinical phenotype with plaque, generalized, bullous, linear, and deep subtypes.[2] Linear, generalized, and pansclerotic phenotypes are associated with the most morbidity; manifesting with joint contractures, facial disfigurement, psychological distress, arthralgia, and central nervous system involvement[1,2]. The underlying pathogenesis of the disease remains unknown, but is likely initiated by vascular injury that culminates in increased collagen production and decreased collagen destruction.[8,9]

The case reported above was unique in many ways the clinical, radiographic, serological and histopathological picture raised a few possible differential diagnosis like bechet's syndrome, oral sub mucous fibrosis, and oral mucosal morphea. The course of disease suggested bechet's syndrome which was ruled out by negative pathergy test but still the diagnostic dilemma prevailed between the possibility of autoimmune oral submucous fibrosis and oral mucosal morphea. Oral submucous fibrosis is a multifactorial disease[10] with proposed pathogenesis as shown in flowchart below where an array of factors contribute together or synergistically to cause submucous fibrosis; though we could not rule out possible HSV/HPV infection ,any underlying genetic abnormality which could have triggered the immune response to cause submucous fibrosis in this case , and the involvement of oral mucosa with submucous fibrosis is unknown in morphea but there have been a few case reports that have reported the occurrence of submucous fibrosis with generalized morphea[11] We label this condition as oral mucosal morphea as there is no history of areca nut or tobacco use, no signs and symptoms of nutritional deficiency, early onset of the condition highly raised ASA titers, and histopathological picture pointing towards morphea(figure 6) .This case is the first reported case which there is exclusive oral mucosal involvement with associated submucous fibrosis and no cutaneous or systemic evidence of sclerosis.

Conclusions

Restriction of mouth opening for a prolonged period of time should be diagnosed though a systematic approach. the autoimmune disorders and collagen disorders like morphea must be considered in the differential diagnosis and should be ruled out where obvious etiology of the condition is not apparent.



References

1. Nicole Fett, Victoria P. Werth. Update on morphea. J Am Acad Dermatol 2011;64:217-28.

2. Peterson LS, Nelson AM, Su WP, Mason T, O'Fallon WM, GabrielSE. The epidemiology of morphea (localized scleroderma) in Olmsted County 1960-1993. J Rheumatol 1997;24:73-80.
3. Laetsch B, Hofer T, Lombriser N, Lautenschlager S: Irradiation-induced morphea: Xrays as triggers of autoimmunity. Dermatology 2011; 223: 9–12.
4. Mayes MD: Classification and epidemiology of scleroderma. Sem Cut Med Surg 1998; 17:22–26.
5. Menni S, Marzano AV, Passoni E: Neurologic abnormalities in two patients with facial hemiatrophy and sclerosis coexisting with morphea. Pediatr Dermatol 1997; 14: 113–116.
6. Torrelo A, Suarez J, Colmenero I, Azorin D, Perera A, Zambrano A. Deep morphea after vaccination in two young children. Pediatr Dermatol 2006;23:484-7.
7. Peroni A, Zini A, Braga V, Colato C, Adami S, Girolomoni G. Drug induced morphea: report of a case induced by balicatib and review of the literature. J Am Acad Dermatol 2008;59:125-9.
8. Pandey JP, LeRoy EC. Human cytomegalovirus and the vasculopathies of autoimmune diseases (especially scleroderma), allograft rejection, and coronary restenosis. Arthritis Rheum 1998;41:10-5.
9. Takehara K, Sato S. Localized scleroderma is an autoimmune disorder. Rheumatology (Oxford) 2005;44:274-9.
10. Radhakrishna Pillai, Prabha Balaram, Kannan Sankara Reddiar, Pathogenesis of Oral Submucous Fibrosis. Cancer 1992; 69:2011-2020.
11. Sarvjit Kaur Virdi, Amrinder Jit Kanwar. Generalized morphea, lichen sclerosis et atrophicus associated with oral submucosal fibrosis in an adult male Indian J Dermatol Venereol Leprol | January-February 2009 | Vol 75 | Issue 1.