

Sarcoidosis macrocheilitis: A clinical form of telangiectasia

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Abstract

Orofacial granulomatosis (OFG) is an uncommon chronic granulomatous condition with a multifactorial etiology and pathogenesis. Genetic, immunologic, allergic, and infectious mechanisms have been implicated. OFG is often used as a descriptor to encompass all entities with orofacial swelling and histologic evidence of non-caseating granulomas. Case report: A 53-year-old patient, admitted for management of macrocheilitis and lesion in the nose, whose exploration is in favor of lupus pernio.

Keywords: lupus pernio, granulomatous cheilitis, telangiectasia, orofacial granulomatosis

1. Introduction

Granulomatous macrocheilitis is a chronic inflammation of the lips, a rare disease of unknown etiology. It was first described by Miescher in 1945 ^[1]. It is characterized by an indurated edema of one or two lips, episodic at first and then permanent. Deep biopsies with numerous slice planes are necessary to reveal epithelioid and giganto-cellular granulomas without caseous necrosis ^[2]. There are many etiologies, particularly the oral manifestations of Crohn's disease ^[3, 4], acquired and hereditary angioedema leprosy, tuberculosis ^[5] sarcoidosis ^[6] Melkersson-Rosenthal syndrome, Miescher's Macrocheilitis, foods are also involved ^[1]. we report a patient with macrocheilitis and lupus pernio in the nose.

Case report

A 53 years-old woman, diabetic, consulted for redness in the nose, associated with a progressive increase in volume of the upper lip, without any notion of exaggeration of the lesions by the heat, asymptomatic with no notion of trauma, insect bite, drug or food allergy, as well as no history of system disease in the family. The dermatological examination had objectified a macro cheilitis non-cracking of the upper lip with an erythematous infiltrated macule surmounted by telangiectasias, sitting at the nose, with extension at the nasogenin sillion, the endo-nasal mucous membrane, the upper lip and the hard palate (figure 1-2). The oral hygiene was good, with no evidence of dental infection, there were no lymph nodes, and the systemic examination was normal. The dermoscopy had objectified an erythematous background, with polygonal telangiectasias, no lipoidic aspect on the vitro-pressure (figure 3). The blood test showed a normal formal blood count, with normal urine and blood calcium. Angiotensin-converting enzyme (ACE) test was normal. A biopsy of the lower lip showed non-castrating granulomas (figure 4-5-6). Tuberculin PCR on oral mucosal biopsy was negative. biopsy of the accessory salivary glands showed a chronic grade 1 of sialadenitis (according to Chisholm and Masson). An upper gastrointestinal endoscopy showed no abnormality eliminating Crohn's disease. Naso-fibroscopy showed a congestive aspect with extension of non-specific nasal

Telangiectasias. Chest X-ray and CT scan of the chest showed no lymphadenopathy or any evidence of tuberculosis or sarcoidosis. The ophthalmological examination and visual fields were without anomaly. The Skin lesion was diagnosed as lupus pernio secondary to sarcoidosis. The patient was treated by hydroxychloroquine 400mg/day, laser IPL PR 550, and laser Nd-yag. And with Improvement (figure 7).



Fig 1: A macro cheilitis non-cracking of the upper lip with an erythematous infiltrated macule surmounted by telangiectasias.



Fig 2: Macrocheilitis



Fig 3: polygonal telangiectasias en Dermoscopy.

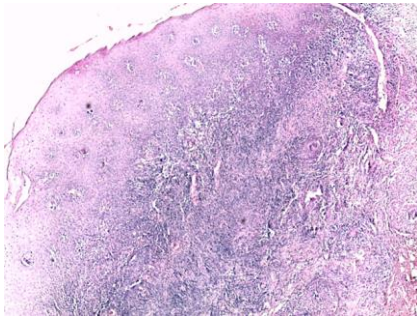


Fig 4: HES G x 50 -> Hyperplastic spongiotic mucosa, with a dense inflammatory infiltrate of the chorion with some epithelioid granulomas and gigantocellular.

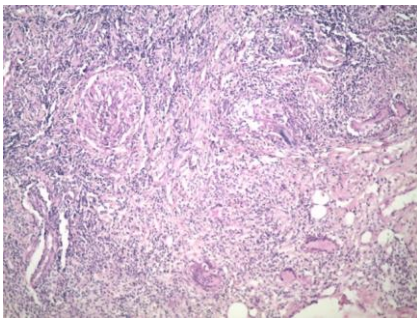


Fig 5: HES G x 100 -> Several more or less well defined gigantocellular epithelioid granulomas

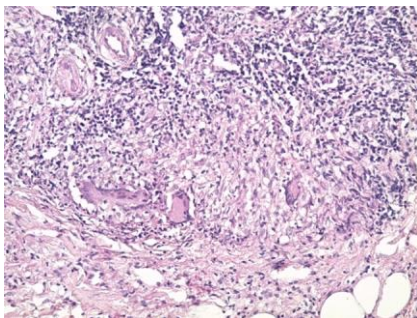


Fig 6: HES G x 200 -> Details of a gigantocellular epithelioid granule.



Fig 7: A good improvement after 10 months of treatment.

Discussion

Sarcoidosis is a chronic granulomatous disorder. It is a multi-systemic disorder affecting of the lungs, liver, of the liver, eyes, skin, and of the parotid glands [7]. The diagnosis is often made on the basis of skin lesions. The incidence of systemic sarcoidosis varies from 6 to 10 patients per 100,000 inhabitants; women are more affected than men. Sarcoidosis may have specific or non-specific cutaneous effects of the lesions. Non-specific lesions lack granulomas under the microscope erythema nodosum [8]. While specific lesions show granulomas of epithelioid cells that are histologically non caseinformal, and include papules, nodules, plaques, lupus pernio ...

Lupus pernio is a rare form of sarcoidosis, It affects both sexes with a predominance of the female sex. Clinically it show a large confluent nodules located on the face, including the nose, cheeks, naso-labial sillium and tongue. The presence of telangiectasias masking the lipid deposition at vitro pressure as well demonstrated by our patient [9]. Differential diagnosis was essentially granulomatous rosacea, lieshmaniasis, tuberculosis, crohn's disease, nasal NK type lymphoma, rhinoscleroma. The association of macrocheilosis with lupus pernio was to our knowledge rarely reported in lithature, this macho-silitis is probably caused by a reactive sarcoidotic granulomatous. The histology is non-specific it shows a naked sarcoidotic granule without caseous necrosis in the dermis, as it eliminates other diagnoses [10].

A paraclinical exploration is necessary to search for visceral sarcoidosis (usually asmtomatic) and to eliminate other differential diagnosis. Laboratory analysis should particularly assess blood and urine calcium levels as well as renal and hepatic functions. a complete blood count, C-reactive protein level and erythrocyte sedimentation rate are often elevated nonspecific findings. A high serum angiotensin-converting enzyme level, although not specific of sarcoidosis and increased in only 60% of patients with Sarcoidosis [9], can add support to the diagnosis, and is particularly useful as a monitor for the disease's progression and activity. Tuberculosis skin tests, and patch tests to research tuberculinic anergy [10, 11], chest X-ray or CT scan of the chest in search of lymphadenopathies hilar or media stinal or parenchymal involvement (radiological stages 1 to 4); ophthalmological examination for uveitis, electrocardiogram for the presence of rhythm and conduction disorder. The treatment is based on general corticosteroid therapy, particularly for severe visceral forms of sarcoidosis. On the other hand there is no indication to use it as first intention during isolated skin attacks unless they are a major aesthetic problem, because of their appearance, their multiplicity, their location on the face, their severity and their scalability [10]. Other treatments are indicated depending on the expected organ severity of the disease, including tetracyclines, hydroxychloroquine, methotrexate, mycophenolate mofetil, cyclophosphamide, azathioprine, cyclosporin, thalidomide, laser [11]....Our patient was treated with hydroxychloroquine, and laser with good improvement.

Conclusion

Granulomatous cheilitis is a chronic condition which is difficult to diagnose and treat. Its presence during Sarcoidosis is little described in the literature, the concern is mainly aesthetic but imposes an exhaustive assessment of

extension in the search for visceral localization which can be serious.

Declaration of interests

The authors declare that they do not have any conflicts of interest by relationship with this article.

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