Case Report

Morbihan’s Syndrome - Report of a Case and Literature Review

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Abstract

Morbihan’s syndrome is a very uncommon entity with a characteristic clinical picture characterized by intense edema and erythema of the upper and middle third of the facial region. Chronic curse with persistent induration and swelling in this location provides patients with a characteristic rostral phenotype and symptoms related with limitations in the visual field and psychological status. We present a typical case of Morbihan’s syndrome reviewing literature reports.

INTRODUCTION

It is called Morbihan Syndrome (Morbus Morbihan), in reference to the French region in which the first cases were described, to an infrequent clinical picture characterized by intense edema and erythema of the upper and middle third of the facial region. Chronic curse with persistent induration and swelling in this location provides patients with a characteristic rostral phenotype.

CASE PRESENTATION

We present the case of a 45-year-old man with no clinical history of interest exhibiting the aforementioned signs during two years. As concomitant symptoms it referred difficulty to open the eyelids, with the corresponding consequences in the visual field. The swelling was accompanied by facial erythema and a waxy aspect of the skin in the upper hemifacies (Figure 1). After the correlation between the clinical picture and the nonspecific histological findings consisting of discrete accumulations of histiocytes without cohesion in the superficial dermis (Figure 2), the diagnosis of Morbihan syndrome was made. The patient was treated with corticoids, ciclosporin and isotretinoin without any improvement.

DISCUSSION AND CONCLUSION

Morbihan syndrome, often referred to as Morbihan disease, was described by Degos in 1957. Its name corresponds to the region of Brittany in which the first cases were described [1].

It is an uncommon but characteristic clinical condition (intense erythema and edema of the middle and upper third of the face, recurrent and chronic, with subsequent edema and swelling) (Figure 1) in which the histology only serves as a support in its confirmation [1,2-11]. Therefore, the careful examination of the patient and a meticulous anamnesis will be the diagnostic key although it is usually resorted to the taking of biopsies in order to rule out edematous pictures with facial manifestation with which, sometimes, a correct differential diagnosis must be established.

It is a picture described almost exclusively in Caucasian patients (it has been described in a black patient and one of an Indian race) [2], adults and in both sexes.

Among the symptoms most frequently reported by patients are the sensation of disfigurement and heaviness of the face and, in cases of eyelid involvement, as in our case, alterations in the visual field. The psychological consequences are usually present in most cases.

Etiology is unknown, having been formulated different hypotheses that have not been firmly demonstrated. In this sense we can talk about alterations of blood flow and lymphatic drainage as well as the same immunological type changes of an urticaria. What does seem to be true is that there is a functional insufficiency of the lymphatic drainage system, which is ultimately responsible for chronic and persistent facial edema [6-8].

The diagnosis is fundamentally clinical, supported by a concordant histology, although the absence of alterations in the analytical studies helps to rule out other entities. Among the most important differential diagnoses we highlight sarcoidosis, Melkerson-Rosenthal syndrome, collagen diseases such as lupus or dermatomyositis and even chronic or accentuated forms of spongiform dermatitis. Some nodular forms, less frequent, must be differentiated from the facial granuloma. Histology is useful in achieving this goal because, although the findings are not specific, they allow in many occasions to discard these other entities [1,2-10].

From the histological point of view, the findings, as we have said, are nonspecific, consisting of dermal edema (Figure 2) of different intensity associated with the presence of histiocytes,
sometimes difficult to identify. The immuno histo chemical positivity for CD 68 helps us to show the presence of them (Figure 3). In our case they are localized in relation to vascular or lymphatic structures, with frequent ectasia although in most of the cases described in the literature histiocytes form granulomas, generally well structured. This constitutes a peculiarity of the present case.

The presence of mast cells at the dermal level has also been described and it is attributed to the obstruction of lymphatic vessels or to the stimulus produced by the same fibrosis [1-3], although Giemsa staining was not able to demonstrate its presence in our case.

The treatment is, in general, unsatisfactory, having practiced multiple formulas with variable and generally poor results. We can mention radiotherapy, steroid corticosteroids, thalidomide, clofazimine and even antibiotics (broad spectrum, thinking of the possibility that it is a variety of rosacea) [11] none successful in their objectives. Lymphatic drainage (manual massage) helps to reduce residual edema. Some cases has he been treated with CO 2 laser blepharoplasty [12] and even surgical procedures [13].

The use of isotretinoin (dose from 20 to 40 mg daily for 3-6 months), only associated with ketotifen (1 mg twice daily) [14,15] has shown some improvement, although not substantial. The introduction of ketotifen in the therapeutic regimen is due to its interference in degranulation of mast cells, cells not identified in our case. Something similar occurs with triamcinolone, whose intralesional infiltration [16] has shown in some cases to have improved the clinical picture by acting on the function of mast cells [17].

Recently, a good response of the symptoms has been described with Tripterygium wilfordii [18], a result not yet generalized.

Finally, we find it curious to highlight the reference in the literature of a case of Morbihan syndrome complicated by dematitis a neglecta [19].

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REFERENCES


Figure 1 Erythema and aedema on the upper third of the facies.

Figure 2 There is some aedema in superficial dermis with collagen bundles separation. Left: HE stain. 40x. Right: HE stain. 100x.

Figure 3 CD68 Immunohistochemical stain demonstrates the presence of histiocytes (brown) in a edematous dermis. 200x.


