

Granulomatosis of Wegener: Revealed by a Total Necrosis of the Nasal Pyramid

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Abstract: The granulomatosis of Wegener is a necrotizing, rare systematic vasculitis, of a not yet established origin. It must be evoked in front of a clinical set of three, dominated by the ENT signs, a total necrosis of the nasal pyramid as in our case is not usual. Therapeutic protocols are especially based upon immunosuppressors.

Keywords: Granulomatosis of Wegener, necrosis of the nasal Pyramid.

INTRODUCTION

The granulomatosis of Wegener is a necrotizing systematic vascularite which affects arterioles and veins; of a not yet established origin. It occurs mostly in middle-aged adults [1], the diagnosis is often late, aggravating even more the prognosis [2].

We take stock of this pathology through our patient's case, in whom it was revealed by an unusual pattern, a total necrosis of the nasal pyramid.

CASE REPORT

We report a case of a 53-year-old patient, admitted in the emergency department for a necrosis of his nasal pyramid, evolving since about ten days (Fig 1).

The interrogation did not reveal history of use of necrotizing products or drug addiction (cocaine), the maxillofacial examination had revealed an aseptic necrosis of the nasal pyramid spread to the labial philtrum, along with an infiltrated aspect of the rest of the mucocutaneous areas of the face.

A biology assessment had been requested urgently, it had objectified a renal insufficiency (creatinine at 45 mg / l), associated with to an inflammatory syndrome (CRP at 217 mg / l)

A facial CT scan had been made showing a thickened aspect of all the mucosa of the ENT sphere, without associated disorders, especially without bone lysis.

A cutaneous biopsy including a part of the facial artery had been realized immediately, and during which we had noticed a much reduced lumen of the artery with a weak arterial jet. An antibiotic treatment with cephalosporin 3rd generation and a corticosteroid therapy had been started urgently.

The evolution had been marked by the installation of an ischemic stroke suspected in front of the rough neurological deterioration and objectified in the brain scan.



Fig-1: Necrosis of the nasal pyramid extended to the upper lip and cheek with a mediobuccal edematous appearance

DISCUSSION

The granulomatosis of Wegener is a rare disease, the incidence of which is estimated to be from 2 to 12 cases / year / one million inhabitants and its prevalence is estimated at 24 in 157 cases / the year / millions of inhabitants [1].

Its pathogenesis is not yet well clarified, nevertheless, immune, infectious and environmental factors [1,2] are incriminated without an established link. Its diagnosis is often late, based on clinical and histological observations; In clinical terms, it must be associated with an ENT, pulmonary and renal triad.

The ENT signs are often the revealing signs of the disease dominated by the nasosinusal damage. The necrosis of the nasal pyramid and its collapse, as in our patient's case, is also found in atrophic polychondritis in 5 to 29 % of the cases in the literature [3].

The occurrence of the stroke in our patient's case is an unfortunate but rare complication, since the central disorder of the nervous system is very infrequent (from 6 to 13 % of the cases), happening a few months after the primary revelation, and it is revealing only in 1 in 2 % of the cases [4].

The paraclinical investigations include a biological impact assessment (inflammatory and renal report), immune system report in search of ANCA anti-PR3 and anti-MPO [1], a radiological report of upper and lower airways and others according to the signs of call. It is characterized by its variability and only allows a diagnostic assumption by gathering signs in favor of the clinical suspicion [5].

The biopsy is often performed at the ENT sphere, and its reading is inconclusive because of the often extensive necrosis, which justifies the utility of taking several samples for the certainty of the diagnosis. [2,3] as in the case of our patient where we were brought from the beginning to make several samples in which we observed two important findings of granulomatosis, it is the reduction of the vascular lumen of the facial artery and its weak arterial jet as well as the infiltrated appearance of peripheral tissues to necrosis.

The reserved prognosis of the disease and any possible complications justify an early therapeutic start [2]; the treatment is essentially based on corticosteroids and immunosuppressants in two phases of attack and maintenance of which the duration and the modalities are adapted according to the patients; the association of antibiotics in consultation with nephrologists can be of great use in the aim of avoiding the chronic carriage of germs (staphylococcus) and to optimize the prognosis in patients with multi-visceral damage [1].

CONCLUSION

Despite its scarcity, Wegener's granulomatosis is a life-threatening disease, from this point of view it must be mentioned whenever the clinician suspects it.

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