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Necrobiotic Xanthogranuloma With Non- Progressive Hematological Dyscrasia, A Case Report

Jhonatan Q. Ossa[1]
Felipe Jaramillo-Ayerbe[2]
Juan Paulo Cardona[3]

[1]Dermatologist, Internal Medicine profesor, Universidad de Caldas, Manizales, Colombia.
[2]Dermatopathologist, Chief of Dermatology program, Universidad de Caldas
[3]Clinical Oncologist, Internal Medicine professor, Universidad de Caldas.

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***Corresponding author:**

Jhonatan Q. Ossa

Dermatologist
Internal Medicine profesor
Universidad de Caldas
Manizales
Colombia
Email: jhonaqo84@gmail.com

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ABSTRACT

Necrobiotic Xanthogranuloma (NXG) is a rare systemic and progressive histiocytosis of unknownaetiologyin which 80% of the cases is considered a marker of paraproteinemia, a monoclonal gammopathy IgG sometimes related with Multiple Myeloma (MM), it has also been associated with other progressive and non-progressive hematologic dyscrasias. Although its location in the periorbital area is distinctive, its course can be systemic and generalized. In the beginning, the diagnosis is difficult and its clinical-pathological correlation becomes necessary. Its prognosis is generally good, but currently there is no standardized or effective treatment;surgery is an option yet with high tendency for recurrence. We present aXNG clinical case associated with autoimmune idiopathic thrombocytopenia, which has remained stable for more than 15 years, although previous reports have shown this association more with adult orbital xanthogranulomathan NXG itself, the clinical significance is yet to be knownaccording to this spectral, infrequent and enigmatic condition.

KEYWORDS

Necrobiotic, Xanthogranuloma, Histiocytoses, Periorbital, Thrombocytopenia

PRESENTATION OF CASE

A 62-year-old man with history of idiopathic, long term thrombocytopeny (over 15 years ago) without history of major bleeding, managed for several years with low doses of prednisolone achieving stability of his platelet count. He had lesions of 17 years of evolution located in the periorbital areas, at the beginning xanthomatous, ill-defined plaques on the skin of both eyelids. He underwent plasticsurgery with recurrence within the following year after the procedure (Figure 1A) and with emergence in the subsequent years of nodular lesions in upper eyelids and indurated tumoral plaques in lower eyelids with smooth, shiny, telangiectatic , yellowish surface without epidermal changes (Figure 1B). The clinic history and the histology were reviewed at the Universidad de Caldas skin cancer clinic; histological findings consisted of an extensive fibrosing process with hypocellular areas, with degenerated collagen and foamy histiocytes and multinucleated giant cells Touton type (Figure 2A & 2B)characteristic of Necrobiotic Xanthogranuloma (NXG). The patient was sent to clinic oncology consultation for screening. Protein electrophoresis failed to show any clonal chain, also a bone marrow aspiration was performed with haematopoietic stem cell within normal limits. He also had a blood chemistry and a lipid profile without alterations. The patient remains asymptomatic but more prominent lesions of tumoral aspects in lower eyelids skin and with an important restriction of his field of vision.

DISCUSSION

Described by Kossard and Winkelman in 1980, NXG is a rare Histiocytosis of non-Langerhans cells, with about only 100 cases brought up to the date. Currently, according thelast revised classification of histiocytoses and neoplasms of the macrophage-dendritic cell lineages of 2016, they place it inside the so called group “C” or of the cutaneous and mucocutaneous histiocytosis none related to juvenile xanthogranuloma family [1-3]. It is also described as one of the four entities that compose the periorbital xanthogranulomatous disease, together with the

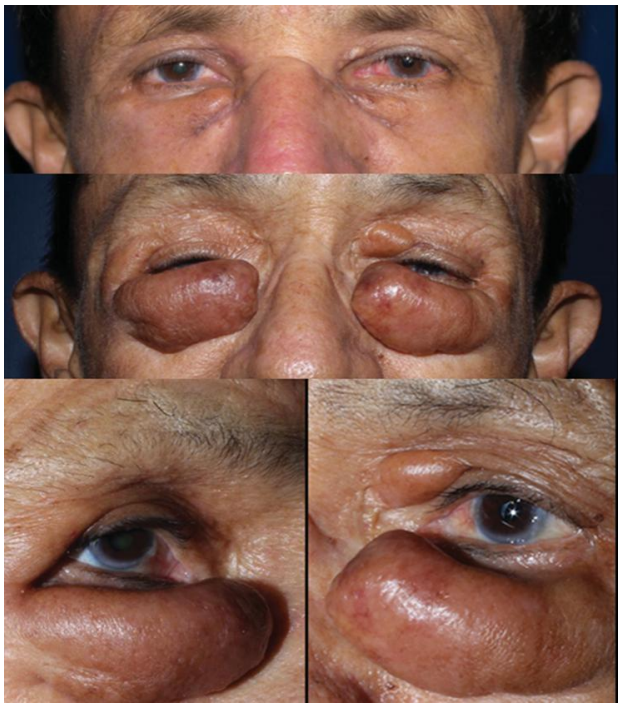

Figure 1: A. Recurrent xanthomatous plaques several years ago. **B.** Xanthogranulomatous periorbital Nodules and tumors.

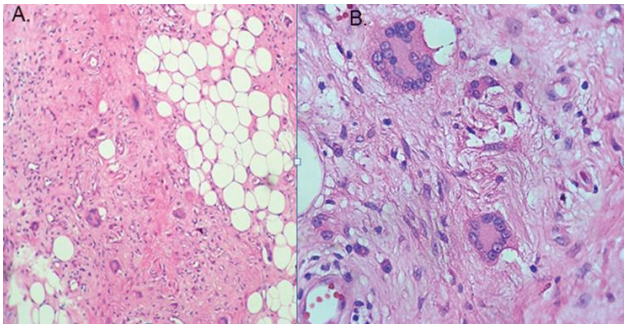

Figure 2: Extensive fibrosing process with hypocellular areas, with degenerated collagen, foamy histiocytes and Touton type multinucleated giant cells. H & E **A.** 10X. **B.** 40X

adult- onset asthmaand orbitalxanthogranulomas, Erdheim-Chester disease and the adult orbital xanthogranuloma (AOXG) [4,5,9].

It is believed that it originates from a stimulant agent that induces histiocytic proliferation that correspond to the free macrophages of the monocyte-macrophage system, nevertheless, up to the date, the above mentioned trigger agent has not been identified [1,3,4,9].

NXG affects both genders equally, commonly between the fifth and sixth decades of life, its location is preferably periocular (80%) and the lesions are described as asymptomatic nodules and papules that slowly turn into a big yellowish reddish tumoral plaque that compromise the dermis and the subcutaneous tissue; telangiectasias, ulceration and itching are also described. Except the last 2, our case presented most of these manifestations. Locations have been described with less frequency in thorax, limbs and at level of cardiac muscle in postmortem patients (myocarditis of giant cells). Its usual location can lead to the following complications: restriction of the field of vision, limitation of the ocular motility by infiltration of its own muscles, swelling like epiescleritis, keratitis; changes in the anatomy such as ptosis (eyelid), proptosis, and even complete blindness in one case -1,4,5,8,9].

From 80% to almost 100% of the cases of NXG are associated to paraproteinemia (mostly monoclonal gammopathy type IgG kappa or lambda); also it has been associated with hematologic dyscrasias of benign and malignant behavior, which in our particular case showed an Idiopathic Thrombocytopenia (ITP) controlled appropriately with low doses of prednisolone, as the only comorbidity; which has been associated in the case reports more to the AOXG(12 patients in total) than to the NXG itself. However, some authors argue that both are spectral of the same entity, being the NXG more severe and locally aggressive than AOXG [3,4,8].

Associations of NXG with malignancy include: Multiple Myeloma (MM), plasma cell leukemia, Waldenström's macroglobulinemia (WM), or cryoglobulinemia; in many cases, cutaneous lesions can precede in several years, even in decades (2-4 years average) those hematological manifestations [1,3,4]. It has also been described the "pure" NXG non-associated with other entities, [2,6]. In our case protein electrophoresis and bone marrow aspiration were normal and not progressive hematological dyscrasia (ITP) clinical significance is yet to be known.

There is no stipulated treatment for NXG, therefore there are multiple therapeutic schemes used in this pathology, with highly variable results, such as: surgical excision (with a recurrence up to 42%), systemic and intralesional corticosteroids; for steroid resistant lesions:methotrexate, azathioprine, cyclophosphamide, hydroxychloroquine, Interferon alpha-2a, radiotherapy, PUVA therapy, laser; Immunoglobulin plusextracorporeal photopheresis have been used recentlywith successful results in twoNXGsdisfiguring variants; all those schemes were not valued in randomized studies only in small series or individual cases and thus with low evidence power [3,4,7,9].

The prognosis of the XGN is generally good; with a survival rate to 10-15 years close to 90 % [1,3,4,6]. In our patient we opted for surgical management with close clinical and hematological follow-up.

CONCLUSION

We present a typical case ofdifficult to treat palpebral NXG associated with idiopathic thrombocytopenia, as far as we know, there is not a clear nor predictive physiopathological association between these 2 entities; nevertheless, the full distinction of this entity inside the xanthodermatosis by the dermatologist and its epidemiological nexus premonitory of appearance of hematologic dyscrasia, entail a multidisciplinary, periodical clinic follow-up and to an early recognition of the potential both local and systemic complications.

CONFLICTS OF INTEREST: The authors declare that they have no competing interests.

SOURCE OF FUNDING: Any study sponsors had no involvement.

ETHICAL APPROVAL: Written informed consent was obtained from the patient for publication of this case report and accompanying images. As these was a report of an interesting case and was not a trail or an observational research there was no need for an ethical approval.

CONSENT: Written informed consent was obtained from the patient for publication of this case report and accompanying images.

The authors use the CARE guidelines as a guiding framework.

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