Nasal Dorsum Swelling: A Rare Diagnosis of Necrobiotic Xanthogranuloma

Deepika Pratap¹, Shrinath D Kamath P², K Sajitha³, J Gayathri⁴

ABSTRACT

Necrobiotic xanthogranuloma (NXG) is a rare granulomatous disorder which presents as yellow plaques and nodules, commonly in the periorbital region. It is important to look for paraproteinemia by serum electrophoresis, as there is a 10% risk of developing multiple myeloma in these patients. This is the first report of NXG presenting as a nasal dorsum swelling. We wish to highlight the importance of histopathological diagnosis, serum electrophoresis and bone marrow biopsy in cases with similar clinical features.

Keywords: Midfacial degloving, Necrobiotic xanthogranuloma, Serum electrophoresis.

Clinical Rhinology An International Journal (2018): 10.5005/jp-journals-10013-1347

INTRODUCTION

Necrobiotic xanthogranuloma (NXG) is a rare, chronic, and progressive granulomatous disorder that manifests as yellowish plaques and nodules commonly in the periorbital region. Approximately 120 cases have been reported since it was first described in 1980.¹ To our knowledge, there have been no reports of NXG presenting as nasal dorsum swelling.

CASE DESCRIPTION

A 38-year-old lady presented with diffuse swelling over the dorsum of upper one-third of the nose since 3 years (Fig. 1). Swelling was firm and nontender, extending laterally up to frontal process of maxilla. Diagnostic nasal endoscopy showed no intranasal extension of the swelling. Computed tomography showed soft-tissue lesion over the nasal bones with no bony erosion (Fig. 2). Fine-needle aspiration cytology revealed spindle cells with no evidence of malignancy. She also had swelling over the eyelids, which were thought to be xanthelesma. Taking into consideration the age, sex, and extent of the lesion, excision of the tumor was done by midfacial degloving approach (Fig. 3). Routine hematological investigation done prior to surgery was normal. Histopathology showed extensive giant cells along with lymphoid follicles, focal entrapped skeletal muscle fibers, and degenerated collagen fibers (Fig. 4). A diagnosis of xanthomatous changes due to NXG was seen in the macrophages, foam cells, chronic inflammatory cells, and conspicuous touton. Serum electrophoresis was then performed which showed a diffuse increase in gamma globulin. Liver function test was normal and showed no A:G ratio reversal. Bone marrow biopsy done was normal. Patient was asymptomatic at 1 year of follow-up.

DISCUSSION

Necrobiotic xanthogranuloma was first described in 1980 by Kossard and Winkelmann, where they reported eight patients with xanthomatous plaques noted to have monoclonal gammopathy, predominantly immunoglobulin G (IgG) κ type.^{1,2} Necrobiotic xanthogranuloma most commonly involves the periorbital area (85% of cases). The other sites that have cutaneous involvement include the trunk and proximal extremities.³ Involvement of

^{1,2}Department of ENT, KS Hegde Medical Academy, Mangaluru, Karnataka, India

^{3,4}Department of Pathology, KS Hegde Medical Academy, Mangaluru, Karnataka, India

Corresponding Author: Deepika Pratap, Department of ENT, KS Hegde Medical Academy, Mangaluru, Karnataka, India, Phone: +447756305579, e-mail: pratap.deepika@gmail.com

How to cite this article: Pratap D, Kamath P SD, Sajitha K, et al. Nasal Dorsum Swelling: A Rare Diagnosis of Necrobiotic Xanthogranuloma. *Clin Rhinol An Int J* 2018;11(2 and 3):55–57.

Source of support: Nil

Conflict of interest: None

extracutaneous sites, such as the skeletal muscle, heart, lungs, kidneys, liver, spleen, intestines, and central nervous system, have been reported.^{4,5} Our patient had no systemic involvement of the disease.

A study done by Wood et al. on 17 patients with NXG found periorbital involvement in 11 patients. They reported systemic findings such as pulmonary involvement, facial palsy, mastoid

Fig. 1: Clinical view of diffuse swelling in the dorsum of the nose

Fig. 2: Computed tomography showing soft-tissue lesion over the nasal bones with no bony erosion

Fig. 4: H&E x400 showing diffuse inflammatory infiltrate comprising lymphocytes, plasma cells, foamy macrophages, touton giant cells (1) and degenerating collagen fibers (2). Inset (H&E x100) showing nodular lymphocytic infiltrate (3)

involvement, splenomegaly, hypertension, hyperlipidemia, melanoma, cirrhosis, and esophageal varices. Serum monoclonal gammopathy was seen in 12 patients, and 3 had multiple myeloma.⁶

Although the exact etiology of NXG is not well established, various hypotheses have been proposed. Paraproteins may act as autoantibodies and bind to lipid forming a complex that deposits in various parts of the body. Paraproteins may also function as a lipoprotein, which allows it to bind to lipoprotein receptors of monocytes and cause xanthoma formation.⁷ However, several cases without paraproteinemia have been reported. An infectious etiology has also been proposed, as *Borrelia* species has been identified in skin biopsy specimens.⁸

Histopathology is the main stay of diagnosing NXG. It is characterized by large areas of necrobiosis, touton and foreign body giant cells, lymphoid follicles, and bands of macrophages and foam cells in the dermis and subcutaneous tissue.⁹ The presence of cholesterol clefts in the areas of necrobiosis helps in differentiating NXG from necrobiosis lipoidica diabetorum that has similar microscopic findings. The combination of typical clinical and pathologic findings helps in differentiating it from other similar conditions.⁶ A serum electrophoresis should be done to look for paraproteinemia. Most (80–90%) of the cases demonstrate monoclonal gammopathy and may be asymptomatic.

Fig. 3: Intraoperative photograph of tumor over the dorsum of nose

IgG κ is the most common monoclonal gammopathy identified. However, 10% of cases can develop multiple myeloma.^{10,11} Other conditions that can be found with NXG include Hodgkin's and non-Hodgkin's lymphoma, chronic lymphocytic leukemia, myelodysplastic syndrome, macroglobulinemia, cryoglobulinemia, and amyloidosis.¹²

Various modalities of treatment have been tried for NXG, ranging from watch-and-wait in asymptomatic patients to intralesional corticosteroids, surgery, radiation, plasmapheresis, and systemic and cytotoxic agents such as chlorambucil, azathioprine, melphalan, interferon α -2b, cyclophosphamide, methotrexate, hydroxychloroquine, nitrogen mustard, and high-dose steroids.^{6,10,13} Stem cell transplantation following high-dose chemotherapy has also been tried with the patient remaining disease free on 2 years of follow-up.¹⁴ The response to treatment has been variable with recurrences in many cases.

Patients require lifelong monitoring for the development of paraproteinemia and lymphoproliferative diseases. There have been no randomized clinical trials to establish the treatment protocol or the frequency of monitoring required. In asymptomatic individuals, annual monitoring can be done and frequency increased if new symptoms or laboratory abnormalities develop.

In conclusion, NXG is a rare granulomatous disease whose pathophysiology and treatment still remains understood. More studies are needed to investigate the significance of serum monoclonal gammopathy in patients with NXG.

REFERENCES

1. Chen KY, Leslie W, Mahon B, et al. A patient with necrobiotic xanthogranuloma presenting with an anterior mediastinal mass, plasma cell dyscrasia, and a lymphoproliferative disorder. *Clin Adv Hematol Oncol* 2011;9(9):696–700.
2. Kossard S, Winkelmann RK. Necrobiotic xanthogranuloma with paraproteinemia. *J Am Acad Dermatol* 1980;3(3):257–270. DOI: 10.1016/s0190-9622(80)80189-7.
3. Mehregan DA, Winkelmann RK. Necrobiotic xanthogranuloma. *Arch Dermatol* 1992;128(1):94–100. DOI: 10.1001/archderm.1992.01680110104016.
4. Winkelmann RK, Litzow MR, Umbert IJ, et al. Giant cell granulomatous pulmonary and myocardial lesions in necrobiotic xanthogranuloma with paraproteinemia. *Mayo Clin Proc* 1997;72(11):1028–1033. DOI: 10.4065/72.11.1028.

5. Shah KC, Poonnoose SI, George R, et al. Necrobiotic xanthogranuloma with cutaneous and cerebral manifestations. *J Neurosurg* 2004;100(6):1111. DOI: 10.3171/jns.2004.100.6.1111.
6. Wood AJ, Wagner MV, Abbott JJ, et al. Necrobiotic xanthogranuloma: a review of 17 cases with emphasis on clinical and pathologic correlation. *Arch Dermatol* 2009;145(3):279–284. DOI: 10.1001/archdermatol.2008.583.
7. Balagula Y, Straus DJ, Pulitzer MP, et al. Necrobiotic xanthogranuloma associated with immunoglobulin m paraproteinemia in a patient with waldenström macroglobulinemia. *J Clin Oncol* 2011;29(11):305. DOI: 10.1200/JCO.2010.32.4921.
8. Zelger B, Eisendle K, Mensing C. Detection of spirochetal micro-organisms by focus-floating microscopy in necrobiotic xanthogranuloma. *J Am Acad Dermatol* 2007;57(6):1026. DOI: 10.1016/j.jaad.2007.05.016.
9. Spicknall KE, Mehregan DA. Necrobiotic xanthogranuloma. *Int J Dermatol* 2009;48(1):1. DOI: 10.1111/j.1365-4632.2009.03912.x.
10. Ugurlu S, Bartley GB, Gibson LE. Necrobiotic xanthogranuloma: long-term outcome of ocular and systemic involvement. *Am J Ophthalmol* 2000;129(5):651–657. DOI: 10.1016/s0002-9394(99)00469-9.
11. Fernandez MM, Prieto MA, Gonzalez IR, et al. Necrobiotic xanthogranuloma associated with myeloma. *J Eur Acad Dermatol Venereol* 2004;18(3):328–331. DOI: 10.1111/j.1468-3083.2004.00906.x.
12. Fernández-Herrera J, Pedraz J. Necrobiotic xanthogranuloma. *Semin Cutan Med Surg* 2007;26(2):108–113. DOI: 10.1016/j.sder.2007.02.008.
13. Vieira V, Del Pozo J, Martinez W, et al. Necrobiotic xanthogranuloma associated with lymphoplasmacytic lymphoma. Palliative treatment with carbon dioxide laser. *Eur J Dermatol* 2005;15(3):182–185.
14. Goede JS, Misselwitz B, Taverna C, et al. Necrobiotic xanthogranuloma successfully treated with autologous stem cell transplantation. *Ann Hematol* 2007;86(4):303–306. DOI: 10.1007/s00277-006-0231-0.