

Wegener's Granulomatosis Revealed by Nasal Pyramid Necrosis: A Case Report

Abdelghafour JAIFI^{1*}, Yassine Bennaoui¹, Mohamed Salah Koussay HATTAB¹, Samir MAIDAME¹, Houssam GHAZOU¹, Zakaria AZIZ¹, Nadia MANSOURI¹

¹Maxillofacial Surgery Department, University Hospital Mohammed VI of Marrakech, Morocco

DOI: [10.36347/sjmcr.2024.v12i06.033](https://doi.org/10.36347/sjmcr.2024.v12i06.033)

| Received: 30.04.2024 | Accepted: 04.06.2024 | Published: 12.06.2024

*Corresponding author: Abdelghafour JAIFI

Maxillofacial Surgery Department, University Hospital Mohammed VI of Marrakech, Morocco

Abstract

Case Report

Wegener's granulomatosis, a rare disease, remains poorly understood. Its pathogenesis involves suspected immune, infectious, and environmental factors, but no definitive relationships have been established. Diagnosis is often delayed and relies on clinical, biological, and histological findings, necessitating an association with an ENT (ear, nose, and throat), pulmonary, and renal triad. ENT symptoms, particularly naso-sinus issues, are the most prominent indicators of the disease. To support the diagnosis, biological and immunological tests are essential. Biopsies, especially from the ENT region, are recommended when possible and safe. Histological confirmation is crucial, and renal biopsies provide valuable prognostic information, particularly in cases with renal manifestations. Treatment primarily involves corticosteroids and immunosuppressants. New biotherapies, such as rituximab, anti-TNF- α , and abatacept, are currently under investigation and show promise for significantly improving patient management in the future. Despite early treatment being crucial due to the disease's severe prognosis and potential complications, over 85% of cases achieve remission. Nonetheless, the high relapse rate remains a significant highlight the need for ongoing research and development of more effective long-term treatment strategies to manage and mitigate relapses in this complex and challenging disease.

Keywords: Wegener granulomatosis, Necrosis, Nasal pyramid, Vascular stroke, Recurrence.

Copyright © 2024 The Author(s): This is an open-access article distributed under the terms of the Creative Commons Attribution 4.0 International License (CC BY-NC 4.0) which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use provided the original author and source are credited.

INTRODUCTION

Wegener's granulomatosis is a necrotizing systemic vasculitis affecting arterioles and venules, of as yet unknown origin, occurring mainly in middle-aged adults. Diagnosis is often delayed, further worsening the prognosis.

We report a case of Wegener's granulomatosis revealed by a non-typical presentation: total necrosis of the nasal pyramid.

CASE REPORT

53-year-old patient admitted to the maxillofacial surgery emergency department of the Mohammed VI University Hospital, Marrakech, Morocco, with necrosis of the nasal pyramid that had been evolving for ten days. On examination, there was no evidence of use of necrotizing products or drug addiction (cocaine). Clinical examination revealed necrosis of the nasal pyramid extending to the labial philtrum,

with infiltration of the rest of the mucocutaneous skin of the face (Fig. 1).

- An emergency blood test showed chronic renal failure with creatinine at 45 mg/l, associated with an inflammatory syndrome with a CRP of 217 mg/l. Facial CT showed a thickened aspect of the entire mucosa of the ENT sphere, with partial lysis of the nasal cartilage framework without bone lysis. The clinical approach was to complete an immunological investigation with anti-PR3 and anti-MPO ANCA antibodies, which came back positive, and a skin biopsy encompassing part of the facial artery, during which we noted a very reduced lumen of the artery with a weak arterial flow. Histological analysis showed ischemic necrosis with amicrobial abscess formation and polymorphic granulomatosis associating polynuclears, lymphocytes and multiloculated giant cells.
- The diagnosis of Wegener's granulomatosis was confirmed, and an antibiotic treatment based on a 3-generation cephalosporin and corticosteroids was started as a matter of urgency.

Citation: Abdelghafour JAIFI, Yassine Bennaoui, Mohamed Salah Koussay HATTAB, Samir MAIDAME, Houssam GHAZOU, Zakaria AZIZ, Nadia MANSOURI. Wegener's Granulomatosis Revealed by Nasal Pyramid Necrosis: A Case Report. Sch J Med Case Rep, 2024 Jun 12(6): 1102-1104.

- The evolution was marked by the development of an ischemic stroke responsible for right hemiparesis with good therapeutic response. It should be noted

that a pulmonary exploration was carried out without any notable findings.



Fig. 1: Necrosis of the nasal pyramid extending to the philtrum

DISCUSSION

- Wegener's granulomatosis is a rare disease with an estimated incidence of 2 to 12 cases/year/million of population and an estimated prevalence of 24 to 157 cases/year/million of population. Its pathogenesis has yet to be fully elucidated, although immune, infectious and environmental factors have been incriminated, with no established relationship (Pagnoux & Teixeira, 2007).
- Diagnosis is often carried out at a late stage, based on clinical-biological and histological findings. Clinically, it must be associated with an ENT, pulmonary and renal triad. ENT signs are often the most revealing signs of the disease, dominated by naso-sinus symptoms. (AYACHE *et al.*, 2001) Necrosis of the nasal pyramid and its collapse, as in our patient's case, is also found in 5 to 29% of cases of polycondritis atrophicans in the literature. The occurrence of stroke in our patient is an unfortunate but rare complication, since central nervous system involvement is a very rare manifestation within a few months of primary revelation, and is revelatory in only 1-2% of cases. (Costentin *et al.*, 2001) (Raissouni *et al.*, 2011) (Silvera *et al.*, 2007)
- Biological and immunological tests are necessary to support the diagnosis, looking for anti-PR3 and anti-MPO ANCA antibodies. Biopsies are often taken in the ENT region, but histological confirmation should always be obtained when it is possible and safe to biopsy the affected organs. Renal biopsy is particularly useful in cases of renal manifestations, as it also provides prognostic information. (1.

Pagnoux C, Teixeira L. Ganolomatose de Wegener.... - Google Scholar, n.d.)

- Treatment is essentially based on corticosteroids and immunosuppressants in two phases of attack and maintenance, the duration and modalities of which are adapted to each patient. The combination of antibiotics must be discussed with the nephrologist.
- New biotherapies such as rituximab, anti-TNF- α or abatacept are currently being studied. Some of them seem quite promising, and could significantly change patient management in the years to come. For the time being, their indication should remain limited to certain refractory forms of the disease (Pagnoux & Teixeira, 2007).
- The reserved prognosis of the disease and possible complications justify an early start to treatment. To date, remission has been achieved in over 85% of cases, but the high relapse rate remains a cause for concern: around half of all patients relapse within five years of diagnosis (Pagnoux & Teixeira, 2007).

CONCLUSION

Despite its rarity, Wegener's granulomatosis is a disease with a reserved vital prognosis, and should therefore be considered whenever clinically suspected.

REFERENCE

- Ayache, S., Smail, A., Stramandinoli, E., Pichon, J., & Strunski, V. (2001). A propos d'un cas de forme pharyngée de maladie de Wegener. *Annales d'oto-Laryngologie et de Chirurgie Cervico-Faciale*, 118(3), 187–192. <https://pascal->

francis.inist.fr/vibad/index.php?action=getRecordDetail&idt=1037927

- Costentin, B., Dehesdin, D., Marie, J. P., Scarcella-Lecler, V., & Andrieu-Guitrancourt, J. (2001). Granulomatose de Wegener et localisations ORL: Analyse rétrospective de 16 patients. *Annales d'oto-Laryngologie et de Chirurgie Cervico-Faciale*, 118(5), 306–314. <https://pascal-francis.inist.fr/vibad/index.php?action=getRecordDetail&idt=14186310>
- Pagnoux, C., & Teixeira, L. (2007). Granulomatose de Wegener. *La Presse Médicale*, 36(5), 860–874.
- Raissouni, L., Aitbenhaddou, E., Ahbeddou, N., El Alaoui, K. T., Slimani, C., Benomar, A., & Yahyaoui, M. (2011). Accident vasculaire cérébral révélant une granulomatose. *Revue Neurologique*, 167(1), 35–38.
- Silvera, S., Vignaux, O., & Legmann, P. (2007). Imagerie des atteintes ORL et cérébrales de la granulomatose de Wegener. *La Presse Médicale*, 36(5), 913–921.