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Case Report

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Mediofacial malignant granuloma: a case report and review of the literature

Abstract

We report a case of malignant midfacial granuloma or idiopathic midfacial granulomatosis in a 50-year-old male farmer with no previous history. He was admitted to the department in an emergency context. The history was reportedly 18 months old. He presented a symptomatology made of nasal obstruction, fever, headache and rhinorrhea with an altered general state. He consulted several health facilities. The evolution was marked by the aggravation of the symptomatology. The examination at admission noted an alteration of the general state. Examination of the ENT sphere revealed ulcerative-necrotic lesions with a destructive nasal origin, with bone sequestration, midface edema, and extension to the sinuses and hard palate. The CT scan revealed bony and cartilaginous lysis, irregular thickening of the nasal pyramid, continuity of the nasal septum, and isodensity of the maxillary, ethmoidal, and left sphenoidal sinus walls without total filling. Deep biopsies were performed under general anesthesia. Histology was consistent with a granulomatous reaction with progressive tissue destruction. Mycobacterium tuberculosis was absent. Medical treatment was initiated. The evolution was marked by the extension of the lesions, malnutrition and the appearance of generalized oedema. The patient died on the 45th day of his hospitalization in a septic shock situation.

Conclusion: Mediofacial granulomatosis is a difficult lesion to diagnose and manage. It requires careful histological exploration in order to differentiate it from other mediofacial destructive lesions. The evolution is most often towards extension and lethal complications.

Keywords: granulomatosis, mediofacial, management, literature review

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Introduction

Nasal NK/T (T/Natural Killer) lymphoma was first described in 1933 as a malignant midfacial granuloma1. Mediofacial granulomas or processes are characterized by a double definition: Clinically by the presence of extensive ulcero-necrotic lesions located in the upper airways evolving towards the destruction of the mediofacial region; and histologically by the presence of an epithelio-giganto-cellular granuloma. The etiologies are multiple: infectious, inflammatory and tumoral; which poses diagnostic difficulties and delays in management.[1] It still has a poor prognosis even if current treatments allow long remissions. The evolution is often difficult to predict and it is necessary to look for other prognostic markers such as cytogenetics.[2]

We report a case of late diagnosed mid-facial malignant granuloma correlated with the literature review.

Observation

He is a 50 year old man, farmer, with no particular history. He was referred to us by a regional hospital in an emergency context. The history goes back 18 months with the occurrence of nasal obstruction, fever, headache, rhinorrhea and dysphagia a little more recent. He consulted in several health facilities. The evolution was marked by the aggravation of the symptomatology. Thus, he was referred to the department for treatment.

The examination at admission noted an alteration of the general condition (WHO index = 3). The examination of the ENT sphere (Oto-Rhino-Laryngology) revealed crusts, perforating ulcerative lesions of the nasal septum and destructive lesions of the turbinates and the nasal floor connecting the nasal cavity and the oral cavity. There was edema of the lips with a fall of the incisors and expulsion of the bone splinters (Figures 1&2). The ganglionic areas were free.

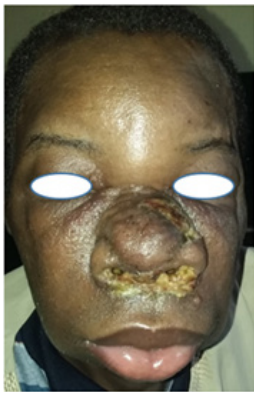

Figure 1 Patient with ulcero-necrotic lesions of the nasal cavity and nasal pyramid, facial edema.

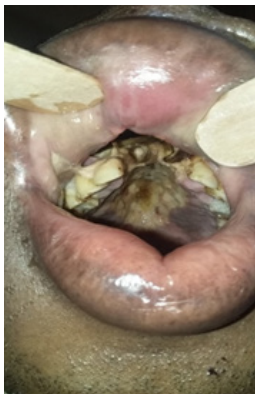

Figure 2 Patient with ulcerative-necrotic lesions of the midline of the bony palate with dental avulsions and lip infiltrations.

The CT scan of the facial mass showed bone and cartilage lysis, irregular thickening of the nasal pyramid, continuity of the nasal septum, and isodensity of the maxillary, ethmoidal and left sphenoidal sinus walls without total filling (Figures 3&4). The chest radiograph was normal.

Figure 3 The CT scan of the facial mass showed bone and cartilage lysis, irregular thickening of the nasal pyramid, continuity of the nasal septum.

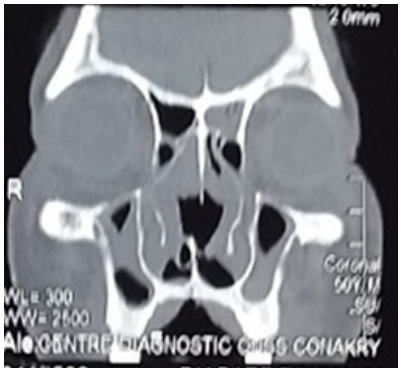

Figure 4 The CT scan of isodensity of the maxillary, ethmoidal and left sphenoidal sinus walls without total filling.

PCR (Polymerase Chain Reaction) using the Gene Xpert apparatus did not identify Mycobacterium tuberculosis DNA. Syphilis serology was negative. Swabbing for cytobacteriological examination and multiple biopsies under general anesthesia were performed. Cytobacteriological examination revealed gram-positive cocci sensitive to Erythromycin and Clindamycin. Histology revealed a destructive granulomatous reaction. The bone tissue was the site of osteonecrosis, with residual lamellae showing inflammatory, polymorphic lymphocytic infiltrate without cytonuclear atypia. There were local proliferations of fibrous tissue consisting of dense collagenous bundles that formed thick wefts. Capillaries were rare. The fibrous tissue was the site of fibrinoid-type necrosis. The blood count revealed an anemia of 8g/dl (normocytic normochromic), leukocytosis and thrombocytopenia. The myelogram noted a bictopenia of peripheral cause.

The patient was treated with ceftriaxone (2g/24h for 7 days); Metronidazole (500mg/8h for 6 days); Betamethazone (8mg/24h for 10 days). We proceeded to decalcification; nasopharyngeal disinfection with saline 0.9%; mouthwash; placement of a nasogastric feeding tube and administration of an antianemic by the tube. Transfusion of a blood bag. Then the antibiotic therapy was secondarily adapted to the result of the antibiogram. The evolution was marked by continuous extension of the lesions, anasarca and cachexia. The patient died on the 45th day of his hospitalization in septic shock.

Discussion

Granulomatous lesions of the upper airways are rare. Patients consult for a long time for a non-specific rhinosinus symptomatology making difficult the diagnosis of the disease at an early stage.[3] Indeed, this was the case in our patient. Endonasal biopsies are most often performed after the extension of the lesions; anatomopathological examinations allow at this stage to specify the diagnosis. The clinical signs and the various paraclinical examinations cannot yet clearly establish the pathogenesis of upper airway granulomatosis.[4,5] The literature presents a great diversity of terminology to name this pathology, in addition there is also a clinical similarity between midfacial granulomatosis and other destructive midfacial lesions.[6] Midfacial granulomatosis, described as enigmatic by some authors, has several names: Robert Woods in 1921, "malignant granuloma of the nose", Stewart in 1933, "progressive lethal granulomatous ulceration of the nose", Tsokos in 1980, "idiopathic destructive disease of the midline.[7,8] The lesions in our patient were identical to those described by Stewart and repeated by Valerio[4] and other authors.[2,9–11] These authors described the course of the disease by the appearance of a non-specific rhinosinusitis, followed by the development of an ulcerous lesion with purulent discharge. The patient's general condition remains surprisingly good throughout the course of the disease. Death occurs after a long period of evolution; it can be due to cachexia, hemorrhage, meningitis or intercurrent infection (often of opportunistic nature) as it was the case in our observation. Wegener's granulomatosis is a rhinogenic form of polyarthritis. Essentially, granulomatosis is necrotizing giant cell disease that usually presents first in an upper airway location with subsequent development of polyarteritic-type arteritis affecting the pulmonary and systemic vasculature. Death of the patient usually occurs because of renal failure caused by the renal vascular involvement. According to Friedmann[12] Wegener's granulomatosis, Stewart's granulomatosis, lymphoid granulomatosis, and the spectrum of diseases intermediate between them are variants of a single disorder. Authors suggest that further immunohisto chemical examination, flow cytometry or molecular testing should be performed in front of such lesions to exclude the possibility of a NK/T cell lymphoma, as many processes previously thought to be of mysterious origin have been found to be lymphomas (especially nasosinus natural killer cell or NK/T cell lymphomas).[13–16] In our case, the biopsy fragments examined showed no cellular atypia that might require further histological examination. However, in the face of such an analysis, we would not rule out repeat biopsies in order to further support the diagnosis. This was different from the case presented by Karima,[2] where granulomatosis was the clinical manifestation of tertiary syphilis. According to most authors, midfacial granulomatosis is predominant in men, as was the case in our observation, but in the study conducted by Alves[17] a female predominance was reported (5 women for 2 men). Can we then look for a hormonal factor that may play a role in the occurrence of midfacial granulomatosis? Given the extension of the lesions and their super infection, a broad-spectrum antibiotic therapy was instituted, local care and corticotherapy in our patient. Despite this treatment, the lesions continued to spread. Several treatments have been proposed by other authors with contradictory results: local or intralesional corticosteroids, dermabrasion, surgery, electrosurgery and cryotherapy. Therapeutic success with systemic corticosteroids, dapsone, clofazimine, antimalarials and PUVA therapy (photochemotherapy) have also been reported.[4,7,18] Friedmann[12] presented a case of granulomatosis that responded well to corticosteroid therapy and in addition had benefited from methotrexate treatment and Cald-Well-Luc; death occurred following extension of the lesions into the middle cranial fossa 6 years later.

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Treatment of this condition is difficult and recurrence is common, even after surgical excision. Survival rates at five years and more after high doses of irradiation are reported by some teams.[12,19] In our context, the insufficiency of the technical platform could explain the limits in the application of certain therapies.

Conclusion

Mediofacial granulomatosis is a lesion of difficult diagnosis and management. It requires a meticulous anatomopathological exploration in order to differentiate it from other mediofacial destructive lesions. The evolution is most often towards extension and lethal complications. Biopsies should be repeated for any midface granulomatosis that does not progress favorably under treatment.

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Conflicts of interests

Authors declare no conflict of interests.

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