ARCHIVES OF HEADANDNECK SURGERY
ISSN 2595-2544

CASE REPORT

Lethal midline granuloma

Caio Paschoalin Trindade[1]*, Rogério Aparecido Dedivitis[1,2], Silvia Migueis Picado Petrarolha[1,3], Kaue Moura[1], Daniel Partezani[3]

Abstract

Lethal midline granuloma is a syndrome with ulcerative vegetative lesion destructing nose, paranasal sinus and palate. Nasal type T-NK cells Lymphoma is often related with this syndrome and without treatment the mortality tax is around 100%. There is association with systemic infection with Epstein Barr Virus. The treatment is the association of radiotherapy and chemotherapy. A 70-year-old male patient presented nose congestion and bleeding with fast growing of an ulcerative lesion in the nose. The computer tomography showed destruction of nose septum and right sinuses. The biopsy diagnosis was nasal type T-NK cells lymphoma.

Keywords: granuloma; lethal midline; lymphoma; T-Cell; nose neoplasms.

How to cite: Trindade CP, Dedivitis RA, Petrarolha SMP, Moura K, Partezani D. Lethal midline granuloma. Arch Head Neck Surg. 2020;49:e00082020. https://doi.org/10.4322/ahns.2020.0015

[1]Hospital Ana Costa, Departamento de Cirurgia de Cabeça e Pescoço, Santos, SP Brasil

[2]Universidade de São Paulo (USP), Faculdade de Medicina, Departamento de Cirurgia de Cabeça e Pescoço, São Paulo, SP, Brasil

[3]Hospital Santa Casa de Santos, Departamento de Cirurgia de Cabeça e Pescoço, Santos, SP, Brasil

Financial support: None.
Conflicts of interest: No conflicts of interest declared concerning the publication of this article.
Submitted: March 25, 2020. Accepted: April 14, 2020.

The study was carried out at Serviço de Cirurgia de Cabeça e Pescoço, Hospital Ana Costa, Santos, SP, Brasil.

Introduction

Lethal midline granuloma is a rare syndrome characterized by a midline ulcerative and vegetative lesion, manifesting in the nose, paranasal sinus and palate, destructing this structures. McBride first described it in 1897. It is more frequent among men, around forty years old[1], being quite rare in the US and Europe and more common in East Asia and Latin America[2,3].

T cell-NK lymphoma and Wegener's granuloma are involved in the majority of the cases[1,4,5]. This is a very aggressive, fast growing and lethal disease. The diagnosis is hard and usually needs several biopsies[1,2]. Wegener's granuloma is characterized by presence of glomerulitis and necrotizing vasculitis involving both arteries and veins. T cell-NK lymphoma has a large granular lymphocyte morphology[2]. It is often associated with systemic infection by Epstein Barr virus[3].

The symptoms are unspecific and the most usual is nasal congestion, however, infections, sinusitis, nose bleeding may occurs[3]. If untreated, it has a very high mortality rate in a short course[5].

Case report

A 70-year-old man presented progressive nasal congestion for six weeks and intermittent nose bleeding. After three weeks, an ulcerative lesion in right nasal cavity with fast growing happened and he lost six kilograms. The CT scan showed a mass in right nose cavity with septal nose destruction and secretion in right maxillary and ethmoid sinus – Figure 1. The biopsy showed nasal extranodal type T-NK lymphoma – Figure 2. The patient underwent

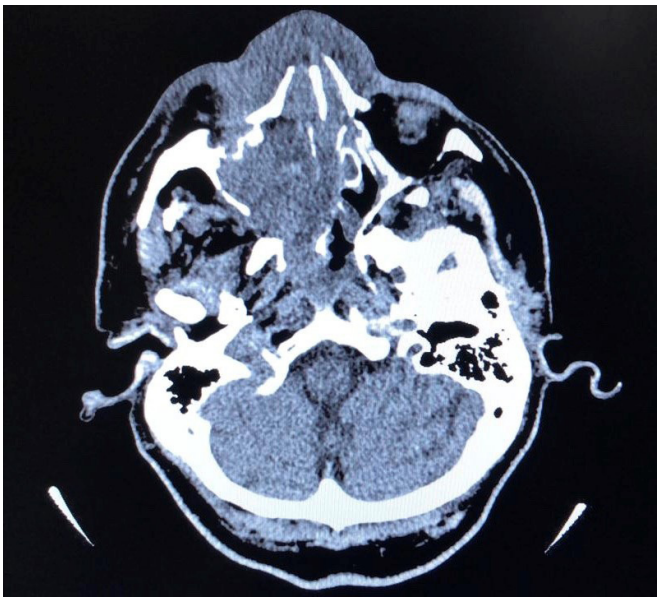

Figure 1. CT scan, axial image, showing a mass with destruction of nose septum and right nasal sinus.

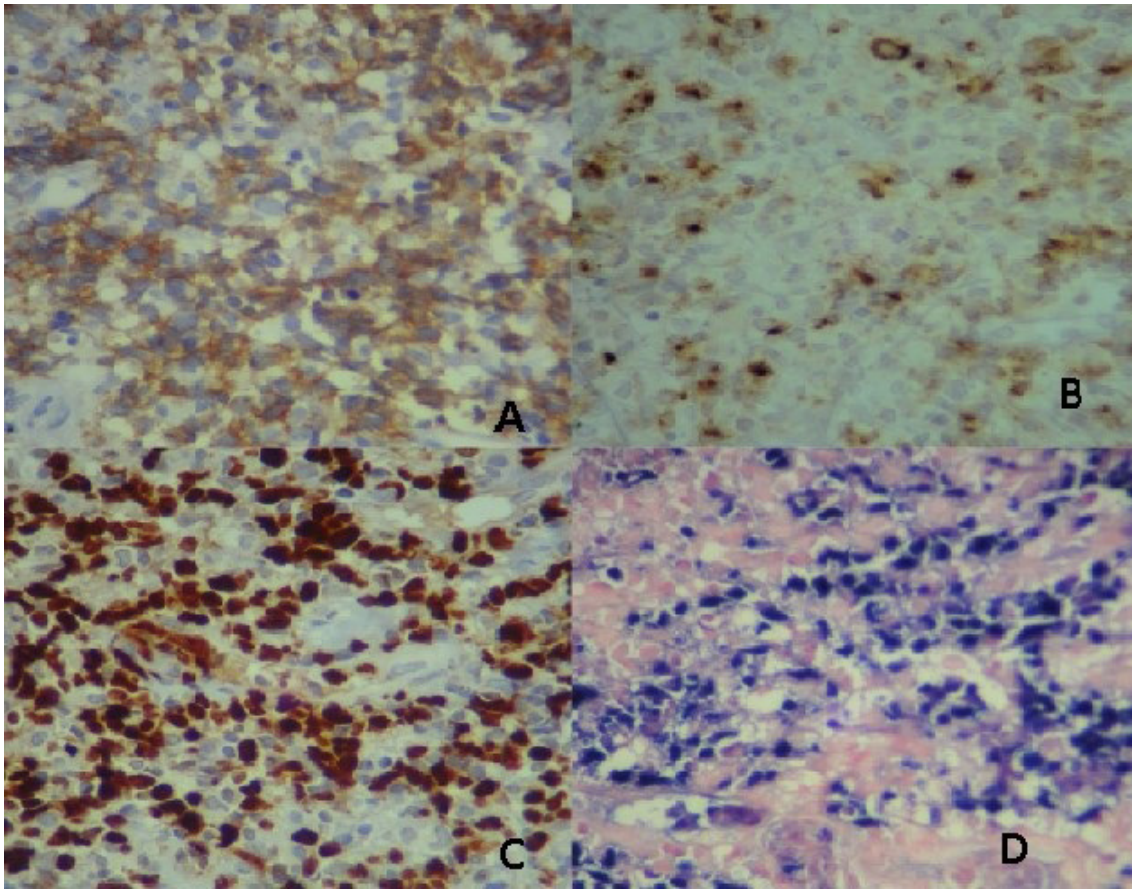

Figure 2. A - CD56 immunohistochemistry reaction on neoplasic lymphoid cells. **B** - Cytotoxic granules immunohistochemistry reaction on neoplasic lymphoid cells. **C** - Ki67 immunohistochemistry in neoplastic lymphoid cells core with high rate of proliferation. **D** - Hybridization reaction with EBV *in situ*.

***Correspondence**
Caio Paschoalin Trindade
Hospital Ana Costa, Departamento de Cirurgia de Cabeça e Pescoço
Av. Conselheiro Nébias, 444, 16º andar
CEP 11045-000, Santos (SP), Brasil
Tel.: +55 (13) 3223-5550
E-mail: caio7med9@hotmail.com

Authors information
CPT - Head and Neck Surgeon; RAD - Assistant Full Professor, Department of Head and Neck Surgery, University of São Paulo School of Medicine; SMPP - Master in Sciences, Head and Neck Surgeon; KM - Resident, Head and Neck Surgery; DP - Resident, Head and Neck Surgery.

chemoradiotherapy associated with total response, without lesion in the physical exam and computer tomography. There was no sign of the disease during second and fourth medical follow up. The patient felt strong headaches on the fifth month and was performed a brain computer tomography with detection of brain metastasis. A new chemotherapy session was performed, however, it had a low response. After 3 months of the diagnosis of brain metastasis the patient died.

Discussion

Nasal type T-NK lymphoma is a very aggressive lymphoid cancer. It is more usual in male than female, in a ratio from 8:1 to 4:1. Very rare in Europe, most cases related are in East Asia and Latin America and is involved with systemic infection of Epstein Barr virus[2,3].

Lethal midline granuloma has a high morbidity and mortality if unthread by septicemia. Initial symptoms are nasal changes as congestion and nasal bleeding[2,3]. It has a very fast and aggressive growing, evolution with an ulcerative granuloma lesion and destruction of the nose, paranasal sinus and palate. Our patient presented these symptoms with fast progressive growing and the quick biopsy and treatment prevent the ulcerative mass to further progress. Most cases of midline lethal granuloma are nasal type T-NK lymphoma[1-5] and Wegener's granuloma. Differential diagnosis includes nose epidermoid carcinoma and cocaine abuse[3].

A higher level of DNA of Epstein Barr virus on blood exams of patients with nasal type T-NK lymphoma is associated with aggressive growing, worst chemotherapy answer and reserved predicted status[3].

The treatment consists in chemotherapy associated with radiotherapy for local lesions. The treatment with chemotherapy only has a not satisfactory regression[1, 4, 5]. The survival time in these cases is around 10-12 months, however, in some cases with fast diagnosis and treatment the survival time is longer and reparative surgery is necessary[3].

References

1. Lessa M, Goto E, Voegels R, Koishi H, Sennes, L, Butugan O, Miniti A. Granuloma de linha média: revisão de 17 casos. @rq Int Otorrinolaringol. 2001;5(1):38-42.
2. Aozasa K, Zaki M. Epidemiology and pathogenesis of nasal NK/T-Cell; lymphoma: a mini-review. Scientific World Journal. 2011;11:422-8. **http://dx.doi.org/10.1100/tsw.2011.41**. PMid:21336457.
3. Silva M, Santos M, Romão J, Paiva A. Linfoma T/NK do tipo nasal: casuística dos últimos 15 anos. Rev Bras Cir Cabeça Pescoço. 2015;44(2):49-53.
4. Ribeiro BNF, Bahia PRV, Oliveira ALVSM, Marchon JL Jr. Síndrome do granuloma letal da linha média: um dilema diagnóstico. Rev Radiol Bras. 2012;45(6):353-5. **http://dx.doi.org/10.1590/S0100-39842012000600013**.
5. Mallya V, Singh A, Pahwa M. Lethal midline granuloma. Indian Dermatol J. 2013;4(1):37-9. PMid:23440011.