

LETHAL MIDLINE GRANULOMA IMPORTANCE OF EARLY DIAGNOSIS: A CASE REPORT

Asem El-Omari, MD*

ABSTRACT

Non-Hodgkin's lymphomas of the sinonasal region are uncommon. They are included in the so-called non-healing midline granuloma syndrome, which is characterized by slowly progressive deep midfacial destruction of soft tissue, cartilage, and bone. Many terms have been used in the literature such as malignant reticulosis or polymorphic reticulosis. A case of high-grade lymphoma is reported, and it is diagnosed in a very late stage as it was mistaken for inflammatory disease and treated medically for several months as a dacryocystitis. When appropriately diagnosed a combined radio- and chemotherapy was offered to the patient, but follow up of patient revealed that he was died at home after 3 weeks of treatment.

Key words: Lethal midline granuloma, Non-Hodgkin's lymphoma, Epstein Barr virus, T-cell lymphoma.

JRMS June 2004; 11(1): 44-45

Introduction

Angiocentric sinonasal T-cell lymphoma is a non-Hodgkin lymphoma; rare in Western countries but common in Asia and China. It has an association with Epstein-Barr virus⁽¹⁾. It is a confusing terminology previously described to include Wegener's Granulomatosis, Polymorphic reticulosis, idiopathic midline destructive diseases, and non-Hodgkin lymphomas now separated into Wegener's granulomatosis and angiocentric T-cell lymphoma⁽²⁾.

The so-called Lethal midline granuloma or 'rhinitis' gangrenosa progressiva (RGP) is however, heterogeneous in its pathogenesis, as it consists of at least two different groups, i.e, inflammatory and neoplastic diseases. The former group comprises Wegener's granulomatosis characterized by necrotizing granuloma, vasculitis and associated glomerulonephritis, whereas the latter includes malignant granuloma and true lymphoma which have been considered to be malignancies of the reticuloendothelial system, designated as 'reticulum cell sarcoma' or midline malignant reticulosis⁽³⁾.

Such extranodal lymphomas originating from the nasal cavities, paranasal sinuses, and hard palate had been included in the so-called non-healing lethal midline granuloma syndrome, which is characterized by slowly progressive deep mid-facial destruction of soft tissue, cartilage, and bones. These lymphomas are difficult to diagnose because the morphology reveals a polymorphous and necrotic backgrounds containing normal looking

inflammatory cells intermingled with large typical lymphoid cells⁽⁴⁾. Additionally, diagnostic confusion may result from the variety of pathologic terms that have been applied to this lesion over years including polymorphic reticulosis, lethal midline granuloma, and midline malignant reticulosis⁽⁵⁾. Angiocentric lymphomas also have been reported in other extranodal sites, such as the skin, soft tissue, testis, upper respiratory tract, and gastrointestinal tract⁽⁶⁾.

Still there is a controversy whether Idiopathic midline destructive disease remains a valid entity because some authors considering that such cases are unrecognized malignant lymphoma or Wegener's disease⁽⁷⁾.

Case Report

A-34-year old man referred from Ophthalmology Department at King Hussein Medical Center for ENT opinion because he developed a swelling of upper and lower right eyelids 10 days after a dacryocystorhinostomy. The anterior rhinoscopy did not show any obvious pathology, while the endoscopic nasal examination showed a soft tissue mass irregular in shape with ugly necrotic appearance in the most posterior part of the right nasal cavity. A biopsy was taken in the clinic, and an urgent coronal CT scan of the sinuses, right orbit, and brain was also requested. The CT reported a soft tissue density mass lesion involving mainly the right maxillary sinus causing bony erosion and complete obstruction of the right maxillary meatus with some fluid collection within the sinus. The mass occupying the nasal

*Correspondence should be addressed to Dr. A. El-Omari, Al-Sakha'a Medical Center, Amman-Jordan, P.O. Box 239 Amman 11947 Jordan
Manuscript received May 9, 2001. Accepted February 6, 2002

cavity posteriorly and extending upward involving right ethmoid air cells. The mass was also eroding the lamina papyracea and extending to the right orbit coming in touch with the medial rectus muscle, inferior rectus muscle and superior oblique muscle. Furthermore, the mass was pushing the globe anterolaterally causing mild exophthalmus. There was also some extension to the right frontal sinus with complete obliteration. There was no evidence of brain involvement, and neck, chest CT scans, and abdominal ultrasound were normal. The nasal biopsy taken in the clinic reported a high-grade lymphoma. Bone marrow aspiration, CBC and blood chemistry were normal.

As the eye swelling deteriorated, so a decision was taken to debulk the tumor in order to decrease the pressure symptoms. Under general anesthesia, an endoscopic examination showed that the right nasal cavity and antrum were filled with a dirty cheesy material, which was removed totally by a Caldwell-Luc approach, and the right frontal sinus was then opened and cleaned up through a Howarth incision. All removed materials were sent to the laboratory for another histopathological study, which showed an extensive necrosis and areas of viable tumor cells forming sheets and composed of large lymphocytes with plenty of clear cytoplasm. The tumor cells were invading the blood vessels and there was a destruction of the included bone. These features are those of high-grade lymphoma.

During his stay in hospital, the patient was on I.V. antibiotics (Rocephen one gram twice daily) and Decadron 4 mg three times daily. The patient was then referred to the oncologist who started him on both chemotherapy and radiotherapy but unfortunately the patient died at home after three weeks of treatment.

Discussion

Malignant lymphomas involving sinonasal tract are unusual in Western populations but much more prevalent in Asian countries⁽³⁾. In the United States, they represent approximately 1.5% of all lymphomas. A higher incidence however has been reported in Asian and South American Countries, especially Peru in which primary non-Hodgkin's lymphoma accounts approximately 6.7-8% of all lymphomas. Other dissimilarities between Western and Eastern cases of sinonasal tract lymphomas include primarily the finding of the EBV-positive T-cell phenotype in Asian countries and EBV-negative lymphomas of B-cell origin in the United States and European countries⁽⁵⁾. Primary nasal lymphomas are a diagnostic problem to the pathologist because of the confusion of terms used to describe this particular

disease⁽³⁾. Clinically lethal midline granuloma can readily be differentiated from Wegener's granulomatosis by an apparent chronic, inflammatory response. There is a remarkably little systemic disturbance and no evidence of pulmonary or renal involvement; death eventually following intercurrent infection or cachexia⁽⁹⁾.

The usual presenting symptoms are nasal obstruction, discharge, epistaxis; sinusitis, deformity and pain. The case reported here was diagnosed in a very late stage, as the patient was treated for 6 months for an infection of the lacrimal sac, before a senior ophthalmologist listed him for a DCR operation after which ENT examination confirmed the diagnosis by taking the biopsy from the sinonasal tract.

Sinonasal lymphomas appear to carry a poor prognosis, and although the lesion of the reported case in this study remained localized with midline destruction for long periods where it showed a very aggressive pattern.

References

1. **Randall DA.** The nose and sinuses. Lee KJ, editor. Essential otolaryngology, head and neck surgery, 7th edition, 1998; p: 781.
2. **Kmucha S, Randall DA.** Haward patrick boey, highlights and pearls. Lee KJ, editor. Essential otolaryngology, head and neck surgery, 7th edition, 1998; p: 1136.
3. **Ishii Y, Yamanaka N, Ogawa K, et al.** Nasal T-cell lymphoma as a type of so-called "lethal midline granuloma". *American Cancer Society* 1982; 50: .2336-23 44.
4. **Yang Y, Gau JP, Chang SM, et al.** Malignant lymphoma of sinonasal region, including cases of polymorphic reticulosis. *Chin Med J* 1997; 60: 236-244.
5. **Abbondanzo SL, Wenig BM.** Non-Hodgkin's lymphoma of the sinonasal tract. *Cancer* 1995; 75(6): 1281-1291.
6. **Jaffe ES, Chan JK, Su IJ, et al.** Report of the workshop on nasal and related extranodal angiocentric natural killer cell lymphomas, definitions, differential diagnosis and epidemiology. *The American Journal of Surgical Pathology* 1996; 20(1): 103-111.
7. **Barker THW, Hosni AA.** Pathology in focus. Idiopathic midline destructive disease does it exists? *The Journal of Laryngology and Otology* 1998; 112: 307-309.
8. **Arber DA, Weiss LM, Albuja PF, et al.** Nasal lymphoma in Peru. *The American Journal of Surgical Pathology* 1993; 17(4): 392-399.
9. **Chessman AD.** Non-healing Granuloma and tumors of the nose and sinuses. Kerr AG, Groves J editor. Scott's Brown's Otolaryngology, 5th edition. 1988; 294-297