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CASE REPORT

Lethal Midline Granuloma: A Diagnostc Dilemma

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ABSTRACT

Lethal midline granuloma (Stewart's syndrome, midline reticulosis) earlier was a name given to all progressive destructive lesions involving the nose, paranasal sinuses, hard palate, face, orbit, and upper airway. With the advent of immunohistochemistry, it is evident that it comprises a heterogenous group of disorders including non-Hodgkin lymphoma, Wegener's granulomatosis, and various granulomatous conditions which can often be encountered with dilemma of diagnosis.

Keywords: Angiocentric growth, Lethal midline granuloma, Midline destructive lesions, Nasal tuberculosis, T-cell non-Hodgkin lymphoma.

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CASE REPORT

A 39-year-old female presented with bilateral nasal obstruction and crusting off and on since 6 months and episodes of epistaxis for the same duration. There was associated low-grade fever since 1 month and facial swelling since 7 days, accompanied by anorexia and malaise. There was no history of cough and hemoptysis and no history suggestive of Koch's, syphilis, leprosy, or any other systemic disease.

On examination (Fig. 1), a diffuse edema was present over the left cheek and periorbital region. Nose had saddle deformity and was filled with foul-smelling crusts with a small septal perforation anteriorly (Fig. 2). Endoscopic examination revealed pink granulations near middle meatus with bony sequestrates. Orally, a midline 2×2 cm ulcer was present over the hard palate with intact underlying bone (Fig. 3). Ocular examination revealed periorbital edema with erythema over upper eyelid and conjunctival chemosis with normal ocular movements and vision.

Hematological investigations revealed leukocytosis with relative lymphocytosis and raised erythrocyte sedimentation rate. Urine microscopy (for casts or hematuria) and sputum analysis were normal. Biochemical tests for c-antineutrophil cytoplasmic antibodies (ANCA), p-ANCA, human immunodeficiency virus enzyme-linked immunosorbent assay were negative.

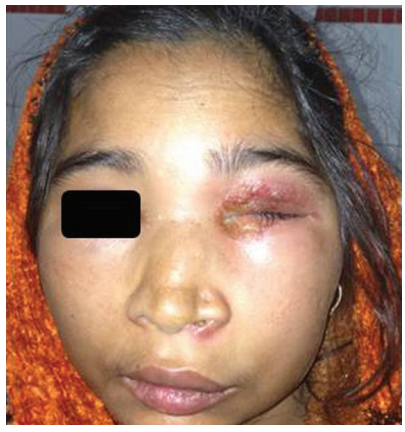

Fig. 1: Frontal view showing periorbital edema and saddle nose

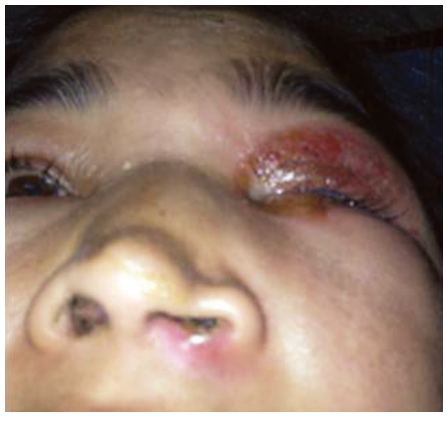

Fig. 2: Saddle nose and nasal crusting

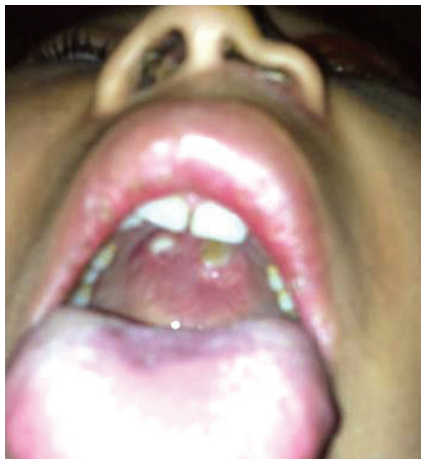

Fig. 3: Hard plate ulceration

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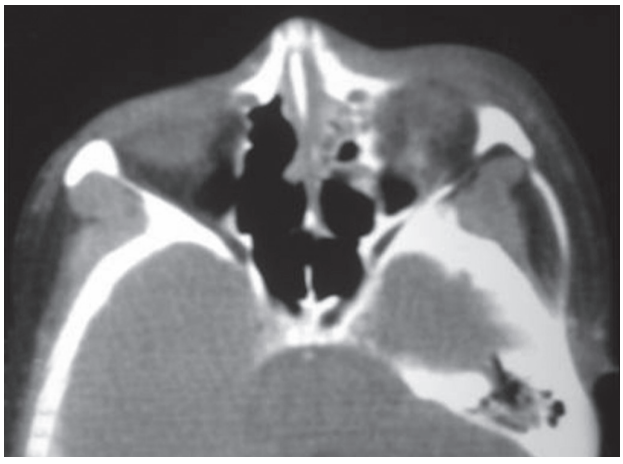

Fig. 4: Computed tomography scan nose and paranasal sinus axial section showing soft tissue thickening in left nasal cavity and maxillary sinus with bony erosion

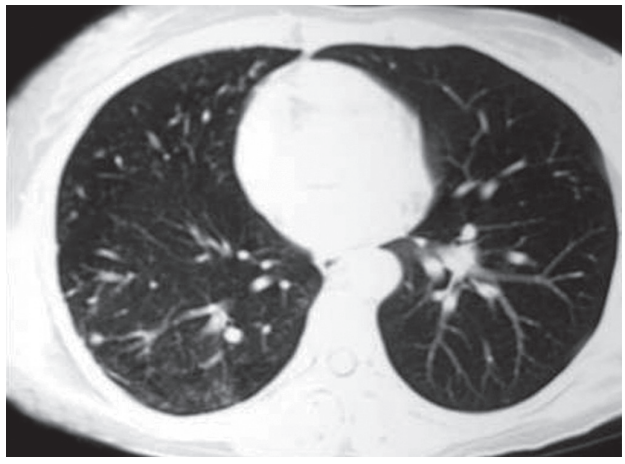

Fig. 5: High-resolution CT chest showing nonhomogenous opacities in perihilar region

Computed tomographic scans of nose and sinuses revealed soft tissue thickening in left nasal cavity and maxillary sinus with bony erosions and anteroinferior septal perforation. No orbital invasion was found (Fig. 4). High-resolution computed tomography of chest showed nonhomogenous opacities in right upper zone and perihilar region (Fig. 5).

Nasal biopsy showed granulation tissue along with fragments of necrotic bone. However, no granuloma was identified with negative fungal culture and acid fast bacilli staining.

Patient was started on symptomatic treatment and systemic antibiotics with anti-inflammatory drugs. And, on the basis of laboratory, radiological investigations, and histopathological examination, antitubercular therapy was started and patient was monitored.

Although, symptomatic improvement in symptoms was reported, the lesions and general condition deteriorated further rapidly over a week, prompting a repeat nasal biopsy.

Histopathology now showed an infiltrate comprising mixed large lymphoid cells and small lymphoid cells showing mitosis with a few B-cells in the background. The large lymphoid cells expressed T-cell markers (CD45 and RO) suggesting T-cell non-Hodgkin lymphoma. Thus, the patient was started on cyclophosphamide-based chemotherapy.

DISCUSSION

Extranodal natural killer (NK)/T-cell lymphoma,[1] nasal type, accounts for 7–10% of all non-Hodgkin lymphoma. It was first described by McBride[2] and subsequent comprehensive account of histopathological and clinical features by Stewart.[3] It is characterized histopathologically by angiocentric[4] and angiodestructive growth, by tumor cells that vary in size and may harbor Epstein Barr Virus (EBY) in a clonal episomal form and by an inflammatory infiltrate of plasma cells, histiocytes, and eosinophils and express T-cell markers on their surface. They are rare and mainly seen in Asia and Latin America. Elderly males above 50 years of age are usually affected with male: female = 8:1.

Differentials include trauma, infection (bacterial: Tuberculosis, syphilis, leprosy, rhinoscleroma, actinomycosis; fungal: Aspergillosis, mucormycosis), toxic (cocaine abuse, chromium salts), inflammatory (sarcoidosis, Wegener's granulomatosis, systemic lupus erythematosus), neoplastic (basal cell carcinoma, esthesioneuroblastoma, rhabdomysarcoma, lymphoma).

Nasal NK/T-cell lymphoma follows an aggressive and rapid downhill course with approximately 50% mortality.[5]

Multidrug chemotherapy followed by involved field radiotherapy appears to be the most effective treatment approach.[6]

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