Lethal midline granuloma

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ABSTRACT

Lethal midline granuloma is a relatively rare disease characterized by destruction and mutilation of the nose and other structures of respiratory passages. The nonspecificity of symptoms obscures the correct diagnosis and is responsible for the delay in treatment which can be detrimental as this grave disease calls for urgent intervention. We present a case report of this disease in a 35 year old male who gave a short two month history of the clinical symptoms.

Key words: Angiocentric, lethal midline granuloma, lymphoma

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INTRODUCTION

Lethal midline granuloma is a midfacial necrotizing lesion that is characterized by destructive, mucosal lesions of the upper aero digestive tract. The patients complain of rhinorrhea, epistaxis, nasal stuffiness, obstruction and pain. The underlying mucosa is thickened and the patient usually develops extensive midfacial destructive lesions, perforated nasal septum and erosion of the nasal bone. The disease is localized to the upper aerodigestive tract at presentation but dissemination to distant sites may occur. Constitutional symptoms may develop. Also referred to as polymorphic reticulosis, midline malignant reticulosis, Stewart s granuloma most of the lethal midline granulomas are NK/T cell lymphomas.[1] Here, we present a case of NK cell lymphoma in a 35 year old male who came with a short clinical history.

CASE REPORT

A 35 year old male presented with swelling in the right side of neck since two months. This was followed by pain and swelling on the dorsum of nose since one and a half months. The pain radiated to right half of the face and head. Patient also gave a two week history of stuffiness in the right nostril and bilateral nasal mucopurulent discharge. There was also a three day history of fever.

On examination, there was a 5 × 4 cm swelling on the right side of dorsum of nose and nasolabial fold. The area was inflamed, erythematous and tender [Figure 1]. A crust covered mass was seen filling the right sided nasal cavity. There was no history of epistaxis, anosmia, post nasal drip, oral bleed or cough. Right sided cervical and submandibular lymphadenopathy was observed.

All routine hematological and biochemical investigations were normal. HIV, HBsAg and VDRL were negative. A contrast enhanced computed tomogram (CECT) of the paransal sinuses showed right sided superficial medial canthus related right sided nasolabial disease with metastatic right sided cervical and submandibular lymphadenopathy. Nasal endoscopic smears were positive for *Pseudomonas aeruginosa*. FNAC of right cervical lymph nodes showed atypical cells against a necrotic background. A biopsy was taken from the mass and sent to National Institute of Pathology for histopathological examination. A diffuse lymphomatous infiltrate having an angiocentric and angiodestructive pattern was seen. The infiltrate was seen destroying the hair follicles [Figure 2a]. Extensive areas of necrosis were also seen [Figure 2b]. Plasma cells, histiocytes and eosinophils were present admixed with atypical lymphocytes [Figure 2c]. Immunohistochemistry was performed and the lymphomatous infiltrate was found to be positive for LCA [Figure 3a], CD56 [Figure 3b] and CD3 [Figure 3c]. Infiltrate was negative for CD20. No neutrophilic microabscesses or granuloma was seen. However fungal stains, Periodic Acid Schiff (PAS) were done to exclude fungal disease. Based on these findings a diagnosis of extranodal

NK cell lymphoma was made. The patient was referred to the oncology department for further management and he received four cycles of multidrug chemotherapy (CHOP regimen). This was followed by external beam irradiation (36Gy). At six months follow-up the mass had slightly reduced in size, there was relief from episodes of epistaxis and nasal discharge had stopped.

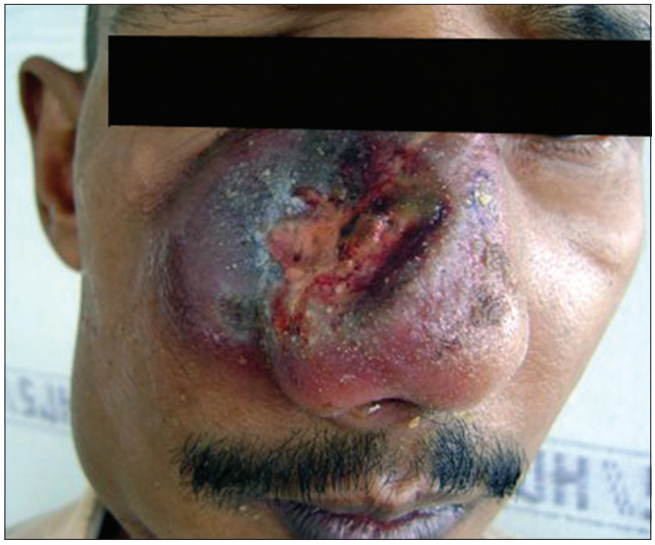

Figure 1: Clinical photo showing swelling and erythema on the dorsum of the nose

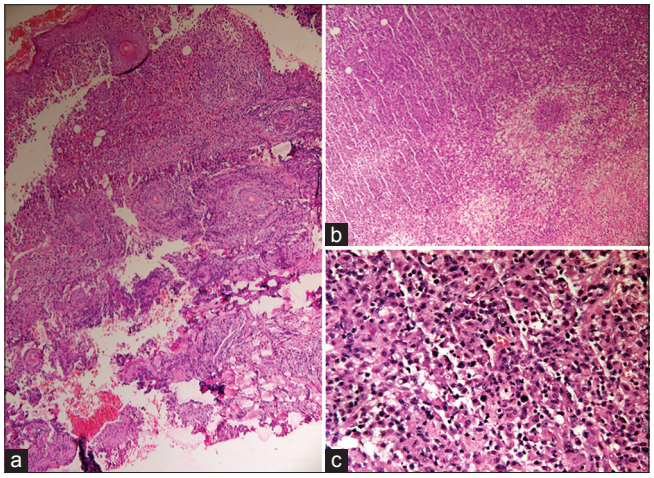

Figure 2: (a) Photomicrograph showing diffuse lymphomatous infiltrate destroying the hair follicles (H and E, ×40); (b) Photomicrograph showing extensive areas of necrosis (H and E, ×100); (c) Photomicrograph showing atypical lymphocytes admixed with histiocytes, plasma cells and eosinophils (H and E, ×400)

DISCUSSION

Malignant lymphomas of the sinonasal region and nasopharynx are majorly of Non-Hodgkins lymphoma type and fall either into NK/T cell type, B cell type or peripheral T cell type. The most common of the so called “nasal type” in which the nasal cavity is the prototype site of involvement are the extranodal NK/T cell lymphomas.[2] In the past this entity has been referred to as lethal midline granuloma, polymorphic reticulosis, malignant midline reticulosis.[3] The term “lethal midline granuloma” was first described by McBride in 1897.[4,5] Macroscopically the lesions usually look like necrotic granulomas and are characterized by ulceration and destruction of the nose and paranasal sinuses with erosion of soft tissues, bone and cartilage of the region. The patients show an aggressive and lethal course with rapid destruction of the nose and face (midline), therefore the term “lethal midline granuloma”. This disease occurs around the fourth decade and the male to female ratio is 8:1 to 2:1. The major symptoms are nasal stuffiness with or without nasal discharge. Oral or nasal ulcer with conjunctivitis may also occur. Perforation of the nasal septum with mutilation of the surrounding tissues eventually occurs.[6] This entity is associated with Epstein Barr virus. Morphologically it is characterized by extensive ulceration of mucosal sites with a lymphomatous infiltrate that is diffuse, but has an angiocentric and angiodestructive growth pattern.[3,5] These tumors have specific characteristics of NK cells. NK cells are active against tumor cells and cells infected with bacteria or viruses without prior sensitization. These develop from precursor cells that can differentiate into NK/T cells and this explains why some of the NK cells can also express T cell markers. The most common immunophenotype of the is CD56+, CD2+ and surface CD20-.[3,5-7] Seen mostly in adults in the Asian and Mexican region, skin involvement is seen in 10-20% of patients. Our patient was a 35 year old male with short history of right sided nasal obstruction and discharge with same sided tender erythematous lesion with lesional biopsy showing atypical lymphoid cells that were positive for CD56 on IHC.

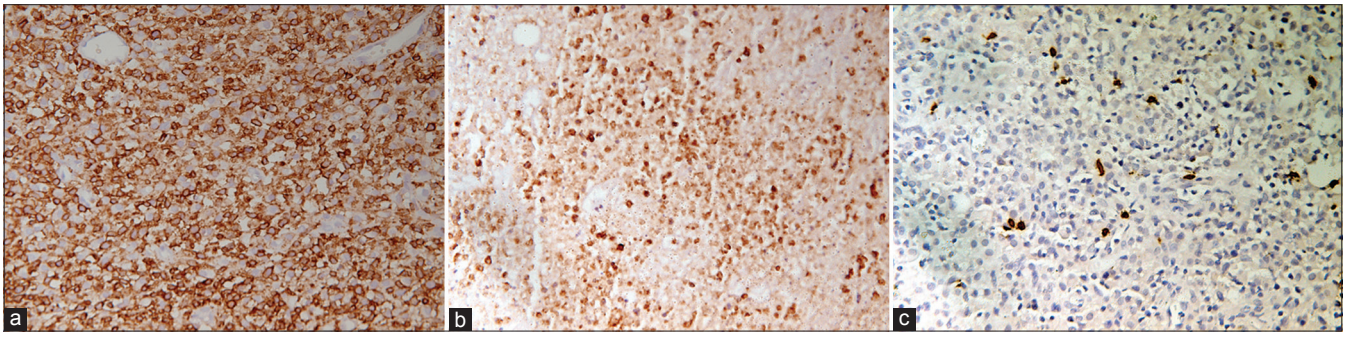

Figure 3: (a) IHC showing atypical lymphocytes positive for LCA (H and E, ×100), (b) IHC showing atypical lymphocytes positive for CD56 (H and E, ×100), (c) IHC showing atypical lymphocytes positive for CD3 (H and E, ×100)

Nasal NK/T cell lymphoma is an aggressive disease with a rapid downhill course. Untreated, this disease has a very high mortality reaching almost 100% due to septicemia, perforation into blood vessels or penetration into brain leading to abscess. Wang *et al*., reported combined chemotherapy followed by involved field external radiation beam to be beneficial in patients and prolonged progression free survival rates.[8] The confusing clinical picture of this disease calls for a high degree of suspicion for correct diagnosis and early intervention to prolong the patient survival.

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