



**RECONSTRUCTION OF A TOTAL NOSE DESTRUCTION DUE TO IDIOPATHIC
MIDLINE GRANULOMA A RARE CASE REPORT**

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Abstract :

Idiopathic midline granuloma is a rare clinical condition, where diagnosing the lesion requires a high index of suspicion. Appropriate and timely management can cure the disease. The destruction that it leaves behind is a challenge for plastic surgeon to reconstruct. A 58 year old male with idiopathic midline granuloma of nose was treated with radiotherapy and later with bilateral forehead flaps.

Keyword : idiopathic midline granuloma, nose reconstruction

Introduction:

Midline granuloma includes a wide spectrum of bacterial, fungal and neoplastic diseases in addition to lesions of unknown origin. Idiopathic midline granuloma is characterised clinically by destructive lesions always localised to upper airway tract and pathologically by non specific acute and chronic inflammation with varying degrees of necrosis¹.

Malignant or atypical cells are invariably absent and no infectious agent can be identified by culture or special staining. Diagnosis of this disorder is important as it is uniformly fatal if left untreated and high dose of local radiotherapy can result in long term clinical remission². Subtotal nasal loss requires an aesthetic and functional restoration. With such a larger loss, reconstructive options are reduced to using forehead flaps and free flaps. Forehead flaps providing the nearest colour match and quality of tissue remains the first choice of treatment.

Case history:

A 58 year old farmer presented with ulceration of nose for six months duration. There was subtotal loss of nose with loss of ala, nasal tip, columella, dorsum, septal and alar cartilages. The exposed nasal cavity was filled with maggots. The infra orbital regions were swollen. He did not have any respiratory difficulty. He was investigated elsewhere and diagnosed to have nasal carcinoma based on biopsy report.



ULCERATION OF NOSE- ANTERIOR VIEW

WIDE LOCAL EXCISION



**IMMEDIATE POST RADIOTHER-
APY - LATERAL VIEW**



ULCERATION OF NOSE - LATERAL VIEW

Patient was admitted and evaluated. Under an-
aesthesia maggots were removed and wide
local excision done. Biopsy report stated it to
be idiopathic midline granuloma.



**IMMEDIATE POST RADIOTHER-
APY- ANTERIOR VIEW**



PLAING OF FOREHEAD FLAPS

He received radiotherapy. Later he was taken up for nose reconstruction. Two para median forehead flaps were planned. Lining of the nose was created with right side para median flap and cover of the nose was created with left side para median flap. Flaps settled well and the secondary defect settled well with split skin graft. In the next stage columella was reconstructed with local adjustment of the flaps.



FOREHEAD FLAPS - ANTERIOR VIEW



**FOREHEAD FLAPS LATERAL VIEW
RECONSTRUCTED NOSE - ANTE-
RIOR VIEW**



**RECONSTRUCTED NOSE - LATERAL
VIEW**

With a one and half year follow up the patient is disease free and has a near normal nose, which enables to continue his social life.

Discussion:

Idiopathic midline granuloma is a relentless progressive localised destructive inflammatory process. It predominantly involves the nose, paranasal sinuses and palate with erosion through contiguous structures, particularly the face³. The majority of patients present with pansinusitis and destructive lesions of the nasal septum or hard palate². A confusing variety of terms has been used since 1897, when McBride first described a case of rapid destruction of the face and nose⁴. This was followed in 1922, by Stewart's report of 10 cases of a chronic midfacial destructive process and came to be known as Stewart's syndrome or Stewart's granuloma⁵. In 1949, Williams popularized the unfortunate term lethal midline granuloma to designate inflammatory midline destructive lesions with no known etiologic factors⁵.

Friedman classified these non healing granulomas in two types

Stewart type- with out vasculitis. 2.Wegners type – with vasculitis⁶. It is also known as Stewart's granuloma, Non healing granuloma , Idiopathic Midline destructive disease, Malignant granuloma and Granuloma gangrenescens .

Both sexes may be affected and the mean age of onset is 35 years. Though the cause is unknown, it has been proposed that the disorder results from a fulminant inflammatory response to an unknown antigen⁷.

Histopathological findings are necrosis with atypical cellular exudates. The hallmark feature is the presence of nonspecific inflammation and necrosis with absence of granulomas and malignant cells³. Diagnosis is essentially one of elimination. There must be no evidence of generalised disease either inflammatory or neoplastic. A local infectious agent must be excluded by staining and culturing biopsy material for bacteria, mycobacteria and fungi. The presence of lymphoma and carcinoma must be sought by histopathological examination of multiple and deep tissue samples. It should be differentiated from polymorphic reticulosis and Wegener's granulomatosis¹.

Untreated the disease is uniformly fatal with death occurring usually after an extended illness from meningitis secondary to erosion of the meninges, haemorrhage or sepsis⁸. Radiotherapy plays the major role followed by reconstruction later.

Conclusion:

Idiopathic midline granuloma is a rare entity causing destruction of nose. There is a possibility to mistake the lesion as carcinoma even in histopathological examination. Hence awareness about the condition is necessary. Radiotherapy cures the disease and patient can be reconstructed and rehabilitated back to his normal life. With the twin objective of curing the disease and

an aesthetic reconstruction of nose being achieved this management provides great patient satisfaction.

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