Mutilating Lesions of Face

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Abstract The destructive lesions affect midline facial areas causing extensive destruction. These lesions are challenging till date due to peculiarity in presentation and non availability of any single objective test. An attempt has been done to evaluate these different clinical presentations of lesions presenting at a single centre and the challenges faced by us at a tertiary government setup. Prospective study done on 15 patients presenting in department during period of 2006–2016. After a detailed history, hematological and biochemical markers, ESR, radiographical tests, pus culture, C ANCA, CECT PNS and histopathological evaluation were done. 12 out of 15 patients were male; Most common age group was above 45 years of age. All cases had ulceration of nose, 9 had cartilaginous destruction and 3 had osseocartilaginous destruction. 5 cases of mucormycosis had hyperglycemia with 2 having ketonuria and 4 had nerve palsies. All patients needed multiple biopsies. 3 cases of Wegners and 4 cases were suspected of IMDD and immunohistochemistry was advised. C ANCA was negative in Wegners cases as they were in limited form. Mutilating diseases of face usually present at an advanced stage with diverse etiology, are slowly progressive and require detailed evaluation. Due to lack of availability of IHC markers and C-ANCA at every centre it is difficult to diagnose them. The treatment is challenging due to incomplete response and frequent relapses. These cases require proper work up plan and perseverance leads to a final diagnosis.

Keywords Midline destructive lesion · IMDD · Wegners granulomatosis

Introduction

Since Hippocrates midline destruction of nose has been documented in literature. The destructive pattern of nose can range from auto-rhinectomy to idiopathic mid line granuloma or granulomatous lesion. Stewart in 1933 reported a case series of chronic midfacial destructive diseases which were later termed as Stewart granuloma [1]. Since then many clinicians have done research over these destructive lesions. This study is an attempt to evaluate the destructive pattern of the disease and available treatment modalities at a tertiary referral centre.

Materials and Methods

This study is comprised of 15 patients, who presented to ENT department with distortion and destruction of nose and mid face during 2006–2016. All the patients underwent detailed history and systemic examination. Blood parameters including hemoglobin levels, blood sugar, renal and liver function tests, urine examination, ESR, radiographical tests, C ANCA tests and CECT PNS were done. Pus collected from the discharging lesion was sent to culture and sensitivity. Punch biopsy from the ulcerating lesion site was subjected to histopathological evaluation. To rule out leprosy, split skin tests were also undertaken. Nasal polyposis, inverted papilloma, benign nasal masses, Malignant cases (squamous, adenocarcinoma, keratoacanthoma etc.) were excluded from the study.

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Aims

The aim of this study was to evaluate

- Presentation of the midline destructive lesions
- Extent of destruction caused by such lesions
- Etiopathologic work up required in these destructive lesions
- Treatment outcome observed in patients after giving first line treatment

Observations

Total 15 patients could fit into the study criteria for this study. Amongst all these, 3 were females and 12 were males. Male to female ratio is 4:1.

Age Group Profile

Youngest case was of a 16 year old male while eldest case was of a 65 year old female. All the patients belonged to rural areas and low socioeconomic strata (Table 1).

Duration of the Disease

Two patients presented with brief duration of 20 days, 4 patients presented after a month of the appearance of the lesion, while 2 cases had a long standing history of 1 year and one had longer history of 2 years.

Symptoms

Nasal obstruction was commonest complaint present in all the patients followed by nasal bleeding and facial edema (Table 2).

Presenting Signs

Ulceration present over nose/face was universal feature seen in all these patients followed by facial/infraorbital edema (Table 3).

Laterality: 10 cases had bilateral involvement while 5 had unilateral affliction of the disease.

An overview of all the patients included in the study is presented here for the complete analysis (Table 4).

- All patients had raised ESR, low hemoglobin (below 9 gm/dl), VDRL and HIV negative.

After detailed history, the patients were subjected to relevant blood investigations (Table 5).

All the patients had raised ESR and hemoglobin levels below 9 gm/dl.

Table 1 Age group distribution

Age group (years)	Number of patients
Less than 20	2
21–30	1
31–40	3
41–50	4
51–60	4
More than 60	1

One patient had bilateral infiltration in chest radiographs and X-ray PNS in all the cases had non specific mucosal thickening and haziness in the maxillary sinuses in all the cases.

CT scan: out of 15 cases, 9 patients underwent CECT PNS. Prominent feature noted was erosion of the bony walls of the maxillary sinus and enhancement correlating to clinical presentation of the lesion.

BIOPSY: all cases needed repeat biopsies. On the basis of biopsy, confirmed diagnosis could not be made for all the cases.

Mucormycosis—5; Wegners—3; Rhinoscleroma—2; Requiring further evaluation—4 (For these cases, further confirmatory investigations like immunohistochemistry were advised which are not available in our institute).

We had mortality of 2 patients during the study period and 8 were later lost to follow up.

Discussion

The diseases affecting face and sinonasal tract are very varied in their clinical spectrum. Various etiologies contribute to mutilating lesions of nose ranging from granulomatous disorders to auto immune diseases. Differentials considered for the mutilating lesions were: (Table 6).

The midline destructive lesions are primarily a diagnosis of exclusion. Even the signs and symptoms are non specific except for the destruction caused by them. Literature review showed every researcher doing Meta analysis taking 2–3 patients at a particular centre, but our study is unique in the fact that 15 cases were taken in the study at a single centre. This excluded those patients with midline destructive lesions who refused for hospitalisation.

In this study, majority of the population were elderly people (age more than 45 years). Male to female ratio was 4:1. Male predominance was significantly noted. All the patients belonged to rural areas and low socioeconomic strata. Majority of the patients were labourer by occupation. All these contribute a major role in etiopathogenesis of the destructive lesion. Poor hygiene, negligence towards

Table 2 Symptoms at the time of presentation

Symptoms	Number of cases
Nasal obstruction	15
Nasal bleeding	9
Facial swelling	9
Diminished vision	7
Pain	6

Table 3 Signs present at the time of examination

Presenting signs	Number of cases
Necrosis of nose	15
Swelling over face/infraorbital area	9
Cranial nerve palsy	4
Nasal septal perforation	8
Palatal ulcer	8
Dorsal cartilaginous destruction	9
Both dorsal bony and cartilaginous destruction	3

health and delay in seeking medical attention makes early diagnosis of the patient difficult.

The presentation was insidious in onset in all the patients, with earliest presentation seen was of 20 days and longest seen was of ulceration since 2 years. The presenting complaints varied with different symptoms like nasal obstruction, foul smelling discharge, ulceration over nose or palate, bleeding from nose and facial edema. Ulceration involving the nose was the universal feature affecting all the patients. Second most common symptom observed was nasal bleeding and facial edema.

10 cases had bilateral involvement of the lesion, most of them being due to progressing extensive destructive pattern of the disease while 5 cases had unilateral affection of the disease. 4 cases had cartilaginous destruction primarily in the necrosed ulcer area while 2 had extensive destruction of both bony and cartilaginous part.

A universal and important finding observed was raised ESR and anemia in all the cases. Chief non specific features that are commonly seen in vasculitis and granulomatous diseases are:

- Low hemoglobin levels (less than 9 gm/dl),
- neutrophilia,
- lymphocytosis and
- raised ESR

VDRL and HIV were universally negative in all the patients taken in the study.

The patients with extensive destruction of nose and huge swelling over dorsum of the nose were mouth breathers. But none of the patients had any adventitious respiratory sounds or any evident respiratory distress.

Radiologically all patients had normal chest radiographs except 1 patient whose radiograph had shown bilateral infiltration. This patient had presented with joint pain and abdominal pain. She was already under the treatment for Koch's abdomen. Palatal ulceration was present along with nasal regurgitation of fluids and denasal speech. On further workup, Mantoux test and sputum for AFB came to be negative. ESR was raised above 80. Radiographs of paranasal sinuses and chest both showed features of bilateral sinusitis and infiltrations respectively. Serum creatinine was 0.64 and biopsy was taken from palatal ulcer as well as endoscopic nasal biopsy. Histopathology suggested the lesion to be chronic non specific inflammatory tissue with features of necrotising vasculitis. Wegners granulomatosis was considered to be diagnosis but the patient had partial response to Cyclophosphamide and steroids. Unfortunately she was lost to follow up.

Second case is of a 35 year old farmer presented in dermatology with presenting complaint of ulceration of face and left forearm since 6 years and burning sensation and ulceration over hands since 15 days. Ulceration which was initially erythematous later on became pustular. Burning sensation developed in the arms and skin started turning blackish. After 6 months, he developed erythematous lesions over dorsum of the nose later on spreading over to cheek areas and face. Patient was treated for rhinoscleroma elsewhere but didn't get any relief. On presentation to our centre, he had developed extensive ulcerated lesion over tip, ala of nose destroying part of nasal septum, columellar skin and pus present in both the nasal cavities (Fig. 1). After ruling out leprosy by split thickness grafts and examination, he was referred to our department for autoimmune disorder. Endoscopic nasal biopsy suggested hyperplastic epidermis and subcutaneous tissue showing dense infiltration by chronic inflammatory cells mainly lymphocytes and plasma cells. Fine granulomas were also seen composed of epithelial macrophages and multinucleate giant cells with blood vessel proliferation and features of vasculitis suggesting towards Wegners Granulomatosis. C ANCA came to be negative in this case.

Available literature suggests that C ANCA test is positive in 95% of patients with vasculitis. But the sensitivity falls to 60% with localized diseases affecting respiratory diseases and sinonasal tract. In our patient who had ulceration over nose and hands, joint pain we suspected limited Wegners granulomatosis. Though CANCA came negative, but the patient responded well to cyclophosphamide and steroid. In these cases, endoscopic tissue

Table 4 Overview of the patients studied

Case number	Patient	History	Physical examination	Diagnostic tests	Final diagnosis
1	21 Y/F	Pain in throat and abdomen, Hyponasal speech Nasal regurgitation	Palatal swelling with perforation and necrosis, Dorsal cartilaginous destruction Bilateral Nasopharyngeal inflammation	X-ray chest: bilateral infiltration X-ray PNS: sinusitis Pus culture: klebsiella Biopsy: chronic non specific inflammatory tissue with vasculitis changes C ANCA: negative	Wegners
2	16 Y/M	Swelling of nose Discharge since 6 months Nasal bleeding	Ulcerated bilateral ala periorbital edema,, gingivolabial nasal fistula, dorsal bony and cartilaginous destruction, palatal perforation	X-ray PNS: bilateral maxillary sinus hazy Pus culture: <i>E. coli</i> Biopsy: chronic non specific granulation tissue	IMDD
3	45 Y/M	Pain and swelling upper jaw 6 months Bleeding on/off	Facial edema, bilateral maxilla swelling periorbital edema, nasal septal perforation, Palatal perforation and necrosis, extensive bony and cartilaginous destruction	X-ray PNS: hazy maxillary sinus Biopsy: intense mononuclear infiltration With degeneration	IMDD/N-KTL
4	65 Y/F	Nasal obstruction and bleeding since 1 month Blindness since 5 days	Ulcer left ala Black debris nose gross destruction of cartilaginous nasal structure; nasal septal perforation eye: no perception of light	Blood sugar –310 Ketone bodies + X-ray PNS– maxillary haziness CT: destruction medial maxillary wall Biopsy: chronic inflammation with broad nonseptate hyphae	Mucormycosis
5	42 Y/M	Swelling cheek 1 month Mucopurulent discharge Facial edema	Ulceration left nose Atrophied middle and inferior turbinates Nasal septal perforation; Diffuse left facial sided edema, Dorsal bony and cartilaginous framework destruction of nose	Pus culture: <i>E. coli</i> X-ray PNS: sinusitis CT: extra osseous enhancement of premaxilla and medial canthus of eye Biopsy: chronic inflammatory and necrotic tissue with areas of monomorphic lymphoid cell population, IHC markers: CD2, CD 3, CD4, CD8, CD 56, CD 20 (test results could not be obtained)	N-KTL
6	35 Y/M	Burning sensation and pustule left hand—4 year Blackening and ulceration of nose—2 year	Ulceration over cheek destruction of bilateral ala, nasal septal perforation, Erythema of lower two-thirds of nose, dorsal cartilaginous framework, Necrosis of upper lip, palatal perforation	CT: Bilateral maxillary enhancement with destruction Biopsy: palisading necrotizing vasculitis C ANCA negative	Wegners
7	55 Y/M	Facial swelling and nasal bleeding—20 days	Infraorbital anaesthesia Necrosis and black debris in bilateral nose cranial nerve 2, 3, 4, 5, 6, 7 palatal perforation, dorsal cartilaginous destruction	Blood sugar 290 X-ray PNS: thickening of sinus mucosa Biopsy: fungal granuloma	Mucormycosis
8	55 Y/M	Facial edema and ulceration—1 month Nasal bleeding	Necrosis of infra orbital area and right nose, black debris, cranial nerves 2, 3, 4, 5, 6, 7 palsy, dorsal cartilaginous destruction of nose	Blood sugar 240 X-ray PNS: sinusitis CT: erosion of medial maxillary wall Biopsy: chronic granulomatous disease with fungal hyphae	Mucormycosis

Table 4 continued

Case number	Patient	History	Physical examination	Diagnostic tests	Final diagnosis
9	54 Y/M	Nasal bleeding—1 month Facial edema—20 days Periorbital edema—10 days diminished vision—7 days	Palatal perforation, Atrophied bilateral middle and inferior turbinates, necrosed dorsal cartilaginous framework, nasal septal perforation, Conjunctival cheimosis, cranial nerves 2, 3, 4, 5, 6, 7 palsy	Blood sugar –290 Ketone bodies + Fungal culture: mucormycosis CT: destruction of middle and inferior turbinates with edema, destruction of medial orbital wall and premaxilla, No intracranial extension Biopsy: granulation tissue suggestive of mucormycosis	Mucormycosis
10	60 Y/M	Nasal blockage blackish discharge Infra orbital margin discoloured	Necrosis and erosion of left middle and inferior turbinates, dorsal cartilaginous destruction, 2, 3, 4, 5, 6, 7 cranial nerve palsy	Blood sugar 220 X-ray PNS: maxillary mucosa thickening CT: involvement of left eye with maxillary sinus thickening Biopsy: granulomatosis broad nonseptate hyphae suggestive of fungal origin	Mucormycosis
11	34 Y/F	Nasal obstruction—1 year	Hard nodular swelling over both ala, Slough and blockage of both nasal cavities, Palate retracted	X-ray PNS: maxillary sinus hazy Biopsy: rhinoscleroma	Rhinoscleroma
12	34 Y/M	Facial swelling—1 year Nasal congestion and discharge	Ulcerated bilateral nasal mucosa with mucopurulent discharge, Gross destruction of endonasal dorsal cartilaginous framework, nasal septal perforation	X-ray PNS: bilateral maxillary sinus hazy Biopsy: chronic inflammatory tissue with vasculitis features C ANCA –	Wegners
13	18 Y/F	Swelling over nose—8 months Hyponasal speech	Woody hard swelling over dorsum of nose, bilaterally	X-ray PNS: maxillary sinus hazy Biopsy: Rhinoscleroma CT: extensive tissue destruction in anterior two-thirds of bilateral nasal cavity	Rhinoscleroma
14	48 Y/M	Nasal pain and obstruction—3 months Bleeding from nose—1 month	Palatal perforation Anterior septal perforation Foul smelling nasal discharge bilaterally, dorsal cartilaginous destruction	CT: infective pathology Biopsy: tubercular origin	Tubercular granulaoma
15	45 Y/M	Ulcer over nose and facial edema—9 months	Destruction of cartilaginous nasal framework, bilateral ala destroyed, septal perforation, Foul smelling discharge, palatal perforation, Infraorbital edema	C ANCA: – Biospy: chronic non specific inflammation CT: diffuse soft tissue over alveolus, bilateral nasal bones and maxillary sinuses. No bone erosion	IMDD

biopsy is very crucial for making a proper diagnosis [2]. Jennings et al. suggested that a negative nasal biopsy and negative C-ANCA does not rule out the possibility of Wegners granulomatosis [3]. Biopsy may be needed multiple times as mostly non specific inflammatory changes are seen.

For Wegners granulomatosis, differential diagnosis like polyarteritis nodosa (PAN), midline lethal granuloma, idiopathic midline destructive disease (IMDD), sarcoidosis and leprosy were also considered [4]. Midline destructive

lesions are separate clinical entity that primarily affect upper respiratory tract and sinonasal tract [5]. Leprosy was not considered as a final diagnosis for the patient with skin lesion and nasal destruction since the split thickness skin biopsies came to be negative and there was no recordable history of any exposure to the pathogen.

IMDD is itself a separate entity and it is very crucial to differentiate it from Wegners granulomatosis. Freidman differentiated between these two entities stating that Wegners shows a giant cell inflammatory granulomatous

Table 5 investigations carried out in the study

Investigations	Total patients	Positive results	Negative results
Hemoglobin level	15	All had < 9 gm/dl	
Lymphocytes	15	15 (raised > 70% polymorphs)	
Blood sugar	15	5 (raised; >210 gm/dl)	10
Blood urea	15	9 (raised; >45 mg/dl)	6 (wnl; 16–40 mg/dl)
Ketone bodies in urine	15	2	13
Serum creatinine	15	5 (>1.5 mg/dl)	10
ESR	15	All (raised)	–
VDRL	15	–	All
HIV	15	–	All
Pus culture		3	
Biopsy	15	11	3 (inconclusive)
C ANCA	2	–	2

Fig. 1 Patient with Wegners Granulomatosis on first visit (with hand lesions)**Table 6** Differential diagnosis of midline destructive diseases

Neoplastic	Non Hodgkins lymphoma Adenoid cystic carcinoma Lymphangioma Basal cell carcinoma Esthesioneuroblastoma
Autoimmune	Wegners granulomatosis Goodpastuers disease Relapsing polychondritis Systemic lupus erythematosus Churg strauss syndrome Sjogrens syndrome Idiopathic midline destructive disease
Traumatic	
Cocaine abuse	
Miscellaneous	

infiltration while IMDD has a more pleomorphic infiltrate with or without histiocytic cell predominance. Primarily the diagnosis of IMDD is that of exclusion [6]. It is a very extensive destructive disease with extensive systemic involvement. But there is early involvement of upper respiratory tract and sinonasal tract and we the otolaryngologists are the one to catch it earlier. The patient with IMDD presented with the lesion gradually progressing from the lateral wall to later extending to autorhinectomy (Fig. 2). For the three cases whose biopsy came to be inconclusive and clinically resembling to midline destructive lesions like

Fig. 2 Autorhinectomy seen in advanced stage of IMDD

idiopathic midline destructive disease (IMDD) or NK T cell lymphoma, following markers were advised: Immunophenotyping, CD 16, CD 56, NKTL, CD2, CD3, NKT cytotoxic granule associated RNA binding protein [TIA1]. But due to poor economic status and loss to follow these test result could not be obtained.

The prime limitation faced during the study was exhaustive work up, multiple biopsies, failed cytometry leading to IMDD diagnosis and lack of advanced tests at our centre like immunohistochemistry. As quoted by [7], IMDD is a diagnosis by elimination, these markers are quite directional in leading towards a final diagnosis. Another difference between wegners and IMDD is the

treatment response; Wegner's granulomatosis responds best to immunosuppressants and steroids while IMDD is highly responsive to radiation therapy [8–10].

Four patients presented with cranial nerve palsies of 2, 3, 4, 5, 6, 7 nerves at the time of presentation. These patients had extensive soft tissue destruction and discharging ulceration present. Such advanced lesions were seen associated with comorbidities of raised blood sugar levels, ketoacidosis and generalized deterioration of body built. We found a strong correlation between mucormycosis and raised blood sugar levels [11]. Poor diabetic control and low socioeconomic status are chiefly leading to more complication rates as earlier disease symptoms are neglected and medical attention is sought at a later stage. Out of total 15 cases, 5 were diagnosed to be of Mucormycosis; 4 being males and one female patient, all above 45 years of age. These patients presented with ulceration over dorsum of the nose extending to infraorbital region and black coloured debris, facial swelling, blackish nasal debris and nasal bleeding had altered cranial nerve 2, 3, 4, 5, 6, 7 function. Out of 5 patients, 2 had ketone bodies in the urine and all had raised blood sugar levels above 210 mg/dl. Biopsy report suggested chronic inflammation with broad non septate fungi. Treatment was started with injection amphotericin for mucormycosis, insulin and endoscopic debridement. 3 patients responded very well, but 2 ultimately died.

Paranasal sinuses radiographs showed universal haziness in maxillary sinuses and mucosal thickening. Sinus involvement is a very common finding with fungal infections affecting sinonasal tract [12].

We had two cases of rhinoscleroma (Fig. 3); both female patients with typical woody swelling over nose,

Fig. 3 Patient with Rhinoscleroma

bilateral ala and retracted palate. The biopsy was definitive of rhinoscleroma (Mikulicz cell). They responded very well with injection streptomycin, dressing with acryflavin and recanalization.

Conclusion

These mutilating diseases are a rare and infrequent clinical entity. They usually present at an advanced stage, have a diverse etiology, are slowly progressive and require very detailed and thorough evaluation. Confirmatory diagnosis cannot be formed in every case. No clear cut tests are available to make a final diagnosis. These patients also keep changing their treating physicians and it is very difficult to evaluate them. No response to the usual treatment should alert us to evaluate these group of rare diseases to be considered and special tests and workup should be done to establish the diagnosis. Not every diagnosis is having fatal outcome; if diagnosed early, mortality and morbidity can be reduced.

Compliance with Ethical Standards

Conflict of interest Both authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with human participants or animals performed by any of the authors.

Informed consent Informed consent was obtained from all individual participants included in the study.

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