STUDY OF DIAGNOSIS AND MANAGEMENT OF MALIGNANT NEOPLASMS OF THE NOSE AND PARANASAL SINUSES: OUR EXPERIENCE

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ABSTRACT

INTRODUCTION

Malignancies (M) of the Nose (N) and Para nasal sinuses (PNS) are uncommon to find in ENT practice. They pose a challenge to the surgeon in terms of early diagnosis and management that includes multiple treatment modalities. Such malignancies are presented from our institute out of other diseases of the Para nasal sinuses, their clinical, radiological and Histo-pathological features with a special emphasis on their management. All the patients were followed up for recurrence.

AIM

To make a clinical study of malignant neoplasms of nose and Para nasal sinuses with regard to their, clinical presentation, radiological assessment, Histopathological studies and treatment modalities.

MATERIALS AND METHODS

Patients attending the ENT outpatient department of a Tertiary Level Teaching Hospital of Government Ear, Nose & Throat Hospital, Koti/Osmania Medical College, Hyderabad, Telangana State from January 2014 to June 2015 with various diseases of nose and Para Nasal sinuses were screened and malignant tumors were identified, investigated and treated appropriately. Among these few cases are included in the present study. The patients are kept on a follow-up period of 6 to 9 months.

INCLUSION CRITERIA

All patients with Histopathological confirmation of malignant neoplasms of nose and Para nasal sinuses irrespective of sex were considered for study. All patients who were immunologically competent only were included in the study.

EXCLUSION CRITERIA

Pediatric malignancies are excluded. All immune-compromised patients are excluded from the study.

CONCLUSION

The Sino-nasal malignancies are not uncommon in ENT practice. Elderly males are commonly affected. Exact aetiology is obscure There is no evidence that chronic sinusitis predisposes to cancer. The incidence of chronic sinusitis in patients with Sino-nasal malignancies is the same as that in the general population. The challenge lies in diagnosing the malignancy as early as possible followed by selecting an appropriate treatment option and adequate follow up.

KEYWORDS

Rare(R), Malignancies(M), Nose(N), Paranasal sinuses (PNS), Management(M), Diagnostic Nasal Endoscopy (DNE).

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INTRODUCTION: Tumours of the Para-nasal sinuses and the nasal cavity account for 0.2-0.8% of all carcinoma in the body.^[1] As few as 1 in 1,00,000 individuals are affected per year.^[2] Maxillary sinus is the most common site for development of malignancy. The most common malignancy is squamous cell carcinoma, which occurs twice as frequently in men as in women. The average age of presentation is 60 years it rarely occurs in individuals under 40 years of age.

Submission 05-01-2016, Peer Review 20-01-2016, Acceptance 28-01-2016, Published 24-02-2016. Corresponding Author: Dr. N. Venkatram Reddy, H. No. 17-1-391/T/Plot 12, Saraswathi Nagar, Saidabad, Hyderabad-500059. E-mail: venkatramreddyn@yahoo.com DOI: 10.18410/jebmh/2016/135 Ethmoidal sinuses account for less than 15% of the malignancies, while frontal and sphenoid sinus malignancies are so uncommon that they account for only 1% of sinus malignancies. When malignancy is suspected under30 years of age, then a sarcoma should be considered.

AIM OF THE STUDY: To study a series of cases of malignant neoplasms of nose and Para nasal sinuses with regard to their:

- 1. Clinical presentation.
- 2. Radiological assessment.
- 3. Histo-pathological studies.
- 4. Treatment modalities.

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MATERIALS AND METHODS:

- Histopathologically confirmed cases of malignancies of nose and Para nasal sinuses are studied with regard to their clinical presentation, radiology, histopathology and treatment protocols.
- The present study is done at the Tertiary Level Teaching Hospital of Government Ear, Nose & Throat Hospital, Koti/Osmania Medical College, Hyderabad, Telangana State for 18 months' duration. i.e. from January 2014 to June 2015.

Inclusion Criteria:

- 1. All Histopathologically confirmed cases of malignant neoplasms of nose and Para nasal sinuses irrespective of sex were considered for study.
- 2. All patients who were immunologically competent.

Exclusion Criteria:

- 1. Paediatric malignancies are excluded.
- 2. All immunocompromised patients are excluded from the study.

A 45 year old female patient came to our outpatient department with the chief complaints of nasal obstruction on the left side since 4 months and facial pain on the left side since 3 months.

Diagnostic nasal endoscopy (DNE) revealed a pink fleshy mass seen in the left nasal cavity arising from the left middle meatus. Examination of the oral cavity revealed a smooth bulge measuring 2× 2 centimetres seen on the soft palate on the left side, which was firm in consistency, with no tenderness on palpation. CT scan showed a heterogeneous soft tissue density in the left maxillary sinus and the nasal cavity with erosion of the floor of the orbit and inferior wall of the maxilla and extending into the left infratemporal & Pterygo-palatine fossa. Biopsy was taken with the help of an endoscope from the mass in the left nasal cavity. Histopathology revealed low grade Adenoid cystic carcinoma of the left maxillary sinus. Staging: T_{4a} N₀ M₀. Treatment: Left total maxillectomy followed by obturator placement was done. The patient has been under regular follow up without any recurrence. Follow-up: This patient was followed up once in two months for 1 year and there is no evidence of recurrence as evidenced in the Diagnostic Nasal Endoscopy.



Fig. 1: Diagnostic nasal endoscopy showing the mass in the left nasal cavity



Fig. 2: CT Scan of the paranasal sinuses: Heterogeneous soft tissue density in the left maxillary sinus

Fig. 3: Left total maxillectomy

A 60 year old male patient presented with the chief complaints of swelling over the left cheek since 8 months and diminished vision since 2 months. On examination there was a single 8×7 cm oval swelling with smooth surface and ill-defined margins over the left cheek below the left eye. Skin over the swelling was normal. On palpation, it is non tender & hard in consistency.

Diagnostic nasal endoscopy revealed purulent discharge in the left middle meatus. CT scan revealed a soft tissue density in the left maxillary sinus extending superiorly into the left orbit and anteriorly into the subcutaneous tissue of the left cheek with erosion of the floor of the orbit and anterior wall of the maxilla on the left side. Biopsy: Biopsy was taken from the proliferative mass filling the left maxillary following a middle meatal antrum antrostomy. Histopathological examination and immunohistochemistry (LCA, CD 20) was suggestive of Diffused Large B Cell Lymphoma of the left maxillary sinus. Treatment: The patient was sent for radiotherapy. Follow-up: After completion of radiotherapy, patient was followed up once in two months for 1 year and there is no evidence of recurrence as evidenced in the Diagnostic Nasal Endoscopy.



Fig. 4: CT Scan of the paranasal sinuses: Soft tissue density in the left maxillary sinus

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Fig. 5: Histopathological picture of diffused large B Cell Lymphoma

A 38-year-old male patient came with the chief complaints of nasal obstruction on the left side since 6 months. Diagnostic nasal endoscopy revealed a pinkish polypoidal mass in the left nasal cavity, not bleeding on touch. CT Scan was suggestive of a homogenous soft tissue density seen in the left maxillary sinus, extending into the Ethmoidal sinus, the orbit, the orbital apex and superior orbital fissure and erosion of the pterygoid plates and extending medially into the left nasal cavity and erosion of the hard palate inferiorly. Biopsy was taken under endoscopic guidance and the Histopathological examination and immunohistochemistry (cytokeratin, S100 and Ki 67) revealed myoepithelial carcinoma of the left maxillary sinus. Staging: T_{4b} N₀ M₀. Treatment: The patient was sent for radiotherapy. The patient has been under regular follow up for 6 months and there was no recurrence. Follow-up: This patient was followed up once in two months for 1 year and there is no evidence of recurrence as evidenced in the Diagnostic Nasal Endoscopy.



Fig. 6: CT Scan: homogenous soft tissue density in the left maxillary sinus

A 20 year old female patient came to our outpatient department with the chief complaints of nasal obstruction on the left side since 5 months. Diagnostic nasal endoscopy shows a pinkish red polypoidal mass in the left nasal cavity, not bleeding on touch. CT scan revealed a homogenous soft tissue density seen in the left Ethmoidal sinus extending into the left anterior nasal cavity. Biopsy was taken under endoscopic guidance and the histopathological examination and immunohistochemistry: cytokeratin, S100 and Ki 67 – positive and Synaptophysin negative) revealed Neuro endocrine tumour Grade II (Atypical Carcinoid) of the Ethmoidal sinuses. Treatment: Left medial maxillectomy by lateral rhinotomy approach and tumour resection was performed. The patient was sent for post-operative radiotherapy. Follow up: The patient has been under regular follow up for 6 months and there was no recurrence.



Fig. 7: Diagnostic nasal endoscopy: Pinkish red polypoidal mass in the left nasal cavity



Fig. 8: Left medial maxillectomy

A 32 year old female patient came with the chief complaints of a forehead swelling on the left side extending up to the left upper eyelid with blurring of vision since 4 months. On palpation, a single firm swelling was noted extending from the left supraorbital rim up to the upper eyelid. The CT scan of the Para nasal sinuses revealed a nonhomogenous soft tissue density occupying the frontal and Ethmoidal sinuses with intracranial and intra-orbital extension. Diagnostic nasal endoscopic examination revealed a proliferative mass in the left nasal cavity which did not bleed on touch. Biopsy was taken and the tissue sample was sent for histopathological examination which was suggestive of a fronto-ethmoidal squamous cell carcinoma. Staging: T4a N0 M0. Treatment: Enbloc tumour resection was done via a Lynch Howarth incision. The defect thus formed was filled with abdominal fat harvested from the periumbilical region. Follow-up: This patient was followed up once in two months for 18 months and there is no evidence of recurrence as evidenced in the Diagnostic Nasal Endoscopy.



Fig. 9: The CT scan: Anon-homogeneous soft tissue density occupying the frontal and Ethmoidal sinuses

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H&E Poorly differentiated sinonasal carcinoma



IHC cytokeratin- Poorly differentiated sinonasa l carcinoma



IHC synaptophysin - Poorly differentiated sinonasal carcinoma

Fig. 10



Fig. 11: Intra operative Photographs

A 70 year old male patient came with the chief complaints of nasal obstruction on the right side since 6 months, nasal bleeding from the right side and blurring of vision in the right eye since 5 months. Diagnostic nasal endoscopy revealed a pinkish red proliferative mass occupying the entire right nasal cavity; bleeding on touch. Ophthalmological examination revealed proptosis on the right side with restricted movements of the eyeball. The patient could perceive only hand movements. CT scan revealed an ill-defined soft tissue density lesion in the right maxillary sinus with destruction of its medial, superior & lateral bony walls and extension into the right orbit, frontal, Ethmoidal & sphenoid sinuses and the nasal cavity. Biopsy was taken under endoscopic guidance and the histopathological examination and immunohistochemistry (cytokeratin, S100, CD34 negative; Ki 67 – positive [50%] and Vimentin positive) revealed spindle cell sarcoma of the right maxillary sinus. The patient refused surgery and thus had to be sent for radiotherapy after explaining about the poor prognosis. Follow-up: This patient was followed up once in two months for 6 months and lost on follow up thereafter.



Fig. 12: CT Scan: An ill-defined soft tissue density lesion in the right maxillary sinus

A 60 year old female patient came with the chief complaints of pain and swelling over the left side of the nose since 1 year. Examination of the face revealed a single 4×3 cm swelling lateral to the nasal bridge on the left side, with ill-defined margins and smooth surface. On palpation, it is tender and hard in consistency .Skin over the swelling is erythematous and not pinchable. Diagnostic nasal endoscopy showed a pinkish grey polypoidal mass in the left nasal cavity, not bleeding on touch. CT Scan: A homogenous soft tissue density seen in the left Ethmoidal sinus with extension into the skin and subcutaneous tissue and extending into the left nasal cavity with erosion of the medial wall of the orbit. Biopsy: Biopsy was taken under endoscopic guidance and the histopathological examination revealed Adenoid Cystic Carcinoma - intermediate grade of the left Ethmoidal sinuses. Staging: T_{4a} N₀ M₀, Treatment: Tumour resection along with excision of the overlying infiltrated skin was done. The defect thus formed was reconstructed with a midline forehead flap and the defect was closed by primary repair. Follow-up: This patient was followed up once in two months for 8 months and had a small recurrence and subjected for local excision and sent for radiotherapy and the patient is under regular follow up till date.



Fig. 13: CT Scan: A homogenous soft Tissue density the left ethmoid sinus



Fig. 14: Intra operative photographs

DISCUSSION: The Sino-nasal malignancies are a rare clinical entity. Males are more commonly affected than females with a majority of them presenting at an older age. Although the exact aetiology is obscure certain occupations actually have a predilection for developing malignancies of nose and Para nasal sinuses. Wood workers - Acheson et al,^[3] reported higher rates of nasal cavity and sinus adenocarcinoma in the High Wycombe area than elsewhere and proposed that this was caused by exposure to wood dust, it is thought that biologically active compounds in wood dust impair mucociliary clearance and predispose to carcinogenesis.^[4]

Other potential carcinogens include Radium dial painters, certain hydrocarbons, Mustard gas, Nickel smelting ^[5] and Chromium manufacturing.

There is no evidence that chronic sinusitis predisposes to cancer. The incidence of chronic sinusitis in patients with Sino nasal malignancies is the same as that in the general population. ^{[6].} According to study at University of Minnesota Hospital and Veterans administration Hospital, the most common presenting signs and symptoms of nose and Para nasal sinus malignancies are nasal obstruction, swelling of cheek, pain or discomfort over cheek, long history of chronic sinusitis, epistaxis and ophthalmologic symptoms. For diagnosis of the malignancy, Computerized Tomography is the most accurate non- invasive test available. It has a diagnostic accuracy of 98% and the radiation is only one third of the staging and grading of the tumour. The overall survival over 5 years ranges from 31% to 46%. [7,8] Adenocarcinoma has a better survival over 5 years (65%) than squamous cell carcinoma (35%).^[9]

Approximately 60-70% of Sino nasal malignancies (SNM) occur in the maxillary sinus and 20-30% occurs in the nasal cavity itself. An estimated 10-15% occurs in the Ethmoidal air cells (sinuses), with the remaining minority of neoplasms found in the frontal and sphenoid sinuses.^[10,11,12]

Risk factors for Sino nasal malignancies (SNM) have been extensively investigated. They are complicated,

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multifactorial, and somewhat controversial. The idea that squamous cell carcinoma (SCC) and adenocarcinoma in this area are associated with exposure to nickel dust, mustard gas, thorotrast, isopropyl oil, chromium, or dichlorodiethyl sulphide is well established. Wood dust exposure, in particular, is found to increase the risk of SCC 21 times and the risk of adenocarcinoma 874 times.^[13] Many of these products are found in the furniture-making industry, the leather industry, and the textile industry. A careful social and employment history should be asked of all patients presenting with symptoms concerning for Sino-nasal malignancies (SNM).^[14, 15]

Viral infections and their relationship to malignancy is an interesting area that has not received sufficient investigation. Preliminary studies show that epidermal growth factor receptor (EGFR) and transforming growth factor-alpha (TGF-alpha) in elevated levels of expression may be associated with early events in inverting papilloma (IP) carcinogenesis. Human papilloma (HPV) and Epstein-Barr virus infection may also be an early event in a multistep process of malignant transformation of inverting papilloma (IP).^[16,17]

Analysis of the study cases: During the study period of 18 months from January 2104 to June 2015 our OP was around 3, 60,000. In our study the commonest age group was 31 to 40 years (2 cases) followed by 51 to 60 years (2 cases) and 1 case each in the age group of 11 to 20, 41 to 50 and 61 to 70 years.

Age in years	No. of cases
0-10	0
11-20	1
21-30	0
31-40	2
41-50	1
51-60	2
61-70	1
Table 1	

In our series of 7 cases of Sino-nasal malignancies compiled over 18months we encountered 2 cases of Adenoid Cystic Carcinoma, 1 case each of diffuse B Cell Lymphoma, Neuroendocrine tumour, Myoepithelial carcinoma, Squamous cell carcinoma (fronto-ethmoidal), Spindle cell carcinoma. As per literature squamous cell carcinoma is commonest Sino- nasal malignancies, but in our limited series we reported adenoid cystic carcinoma (2 cases) as common.

Type of Malignancy	No. of cases	
1. Adeno cystic carcinoma	02	
2. B Cell Lymphoma	01	
3. Neuroendocrine tumour	01	
4. Myoepithelial carcinoma	01	
5. Squamous cell carcinoma	01	
6. Spindle cell carcinoma	01	
Table 2		

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In Hyderabad we have Govt. Regional Cancer Institute where the common malignancies of nose and Para nasal sinuses are squamous cell carcinomas and very rarely these cases are reported in this institute.

CONCLUSION: The malignancies of the nose and Para nasal sinuses are an intriguing group of disorders due to the variations in their clinical presentation, histopathological diagnosis, staging and grading. The type of treatment modality also varies depending upon the aforementioned factors. The challenge lies in diagnosing the malignancy as early as possible followed by selecting an appropriate treatment option and adequate follow up.

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