RESEARCH PAPER | Medical Science | Volume : 4 | Issue : 2 | Feb 2014 | ISSN - 2249-555X

Paranasal Sinus Malignancies – A 6 Year Tertiary Hospital Experience

KEYWORDS: Paranasal sinus malignancies, Squamous cell carcinoma, Maxillectomy, Chemoradiation

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ABSTRACT *AIMS AND OBJECTIVES: To illustrate our experience in the demographics, tumour characteristics and management of paranasal sinus malignancies during an 6-year time period.*

MATERIALS AND METHODS: This is a retrospective, observational study where clinical data of 41 patients diagnosed and treated for paranasal sinus malignancies were analysed.

RESULTS: The male:female ratio was 1.6:1 and the median age of presentation was 48 years. The most common presenting symptom was epistaxis (46.4%). The median time gap between the onset of symptoms and presentation was 3 months. The maxillary sinus was the most commonly affected site (68%) and the most common malignancy was squamous cell carcinoma(39%). 90% of the patients underwent surgery as part of a multimodality curative treatment plan or alone as curative treatment.

CONCLUSION: Advanced T stage, regional and distant metastasis of paranasal sinus malignancies are highly predictive of poor survival. Hence early diagnosis and treatment is of immense value in decreasing the morbidity and mortality due to these tumors.

INTRODUCTION

Sinonasal malignancies are rare tumours and comprise approximately <5% of all head and neck tumours[1]. These tumours are highly challenging to diagnose and manage. These tumours are often diagnosed late and have aggressive behaviour. The associated complicated anatomy of the nose and paranasal sinuses prevents complete exenteration of the disease resulting in poor outcome and high morbidity.[2]

Patients initially present with symptoms like nasal blockade, nasal discharge, headache, facial pain and pressure and hyposmia. These symptoms are seen commonly in benign nasal pathologies like allergic rhinosinusitis and chronic sinusitis. Patients are treated for these benign conditions, thereby delaying the diagnosis of the malignancy.[2]

MATERIALS AND METHODS

We retrospectively reviewed the charts of all patients treated for paranasal sinus (PNS) malignancies treated at a single institution for 6 years with a minimum 3-year follow-up. Gender, age, TNM stage, anatomic site, pathology, treatment and recurrence rates were reviewed.

Results:

There were 41 cases diagnosed with PNS malignancy during the above mentioned period. The male to female ratio was 1.6:1 and the median age of presentation was 48 years. The most common presenting complaints were epistaxis – 19 patients (46.4%) followed by nasal obstruction – 16 patients (39%), swelling of the cheek – 4 patients (10%) and swelling of the cheek and proptosis and nasal obstruction 2 patients (4.8%). The median time between the onset of symptoms and presentation was 3 months (range 1 – 36 months). All the patients were followed up for a minimum period of 3 years.

The maxillary sinus was the most commonly affected site (68%) followed by the maxilloethmoid complex (17%) sphenoid(7.3 %), sphenoethmoid complex(4.87%) and frontoethmoid complex(2.43%). The distribution of site of tumors and T staging was as follows:

(1) Maxillary sinus T1 – 2 (4.88%), T2 – 7 (17.07%), T3 – 5(12.2%), T4a – 10(24.39%) and T4b – 4(9.75%).
(2) Maxilloethmoid complex T2 - 1(2.44%), T3 – 4(9.75%) and T4a – 1(2.44%).
(3) Frontoethmoid complex T4a – 1(2.44%).
(4) Sphenoid T3 – 1(2.44%), T4a – 2(4.88%).
(5) Sphenoethmoid complex T4a – 2(4.88%).

The nodal staging was as follows:

(1) Maxillary sinus N0 – 18(43.9%), N1 – 5(12.2%), N2a – 2(4.88%), N2b – 2(4.88%) and N2c – 1(2.44%).
(2) Maxilloethmoid complex N0 – 4(9.75%), N1 – 1(2.44%) and N2c – 1(2.44%).
(3) Frontoethmoid complex N0 – 1(2.44%).
(4) Sphenoid N1 – 1(2.44%) N2a – 1(2.44%) N2b – 1(2.44%).
(5) Sphenoethmoid N0 – 2(4.88%)

One patient had Olfactory neuroblastoma Kadish stage 2 and not staged as per TNM staging. Two patients were diagnosed with haematolymphoid tumours- AML and Non-Hodgkins lymphoma and staged as per TNM classification. Nodal spread was seen in 36.6% of the patients. One patient had metastatic squamous cell carcinoma (SCC) of the sphenoethmoids from SCC of tongue. One patient of round cell sarcoma of the maxillary sinus developed distant metastasis in the the lower cervical vertebra and mastoid bone(Table 1). The staging of all tumors in the study is mentioned in Table 2. The most common malignancy was SCC(39%), followed by adenocystic carcinoma (17%). Other tumours including soft tissue tumours, haematolymphoid tumours, neuroectodermal tumours together comprised 44 % with <5% contribution from each of the individual histological types. 13 patients(31.7%) percent of the patients underwent surgery alone, 22 patients(53.65%) underwent surgery followed by radiation therapy, 1 patient(2.4%) underwent radiation thera-

Volume : 4 | Issue : 2 | Feb 2014 | ISSN - 2249-555X

py followed by surgery, 1 patient(2.4%) underwent radiation therapy alone, 2 patients(4.8%) underwent surgery followed by chemoradiation therapy. Chemotherapy alone was given for 2 patients(4.8%) the patients with AML and NHL.(Table 3) On follow up after 1 yr, 9 patients (22%) developed recurrent disease after treatment.

Most of these were SCC of stage III/IV who received only surgery as sole modality of treatment. These patients have taken radiotherapy/chemotherapy for the recurrence and on follow up for 1 year after the radiotherapy did not have further recurrences. Most of the recurrences occurred in the sited adjacent to the primary. Two patients had nodal recurrences after surgery and radiotherapy and underwent radical neck dissection.

DISCUSSION:
Etiology of the PNS malignancies is related to the occupation. Exposure to smoking, formaldehyde, asbestos, and leather dust has increased risk of developing sinonasal adenocarcinoma and squamous cell carcinoma. Exposure to wood increases the risk of developing adenocarcinoma particularly in the ethmoid sinuses. The role of tobacco and alcohol has not been proven in the oncogenesis of the sinonasal malignancies[3]. This is the drawback of this study that the exact nature of occupation of the patient was not mentioned in the charts.

SITE: Maxillary sinus was the most commonly involved sinus– 68.3%, followed by maxilloethmoidal complex - 17 %, Other studies in literature have shown the similar results regarding the site of involvement.[3,4] In view of the extensive local spread and delayed presentation, these tumours were not confined to one site, hence making it difficult to determine the exact site of origin.

HISTOLOGY: SCC was the most common histology (39%). Other studies have shown about 75-90% of the PNS malignancies were SCC.[3,4]

DIAGNOSIS: In view of the nonspecific initial symptoms, which mimic the commonly seen diseases like allergic rhinitis and sinusitis, the referral to a physician is delayed. Recurrent epistaxis is the commonest presenting symptom followed by nasal obstruction.[3] The specific signs and symptoms pertaining to the tumour depends on the location and extent and involvement of the adjacent structures like orbit, cranial nerves or invasion of skull base.[1,3,4]

STAGING: Majority of the patients were diagnosed in the advanced stages. Many studies have shown that majority of the patients of paranasal sinus malignancies present in advanced stages.[3,4] Regional spread is rare. It occurs through the lymphatics draining into the facial, parotid, submandibular, retropharyngeal and parapharyngeal nodes.[2] In our study, one patient even had metastatic squamous cell carcinoma to the sphenoethmoids from SCC of the tongue. One patient had olfactory neuroblastoma (Kadish stage B) with involvement of the sphenoid sinus and pterygopalatine fossa. One case of undifferentiated round cell sarcoma had metastasis to the lower cervical vertebra and mastoid.

TREATMENT: Ethmoid sinus: Early stage(T1-T2) tumors can be resected by endoscopic or external approach whereas locally advanced tumors can be resected by external approach or craniofacial resection.[3,4] Radiotherapy forms an important adjuvant therapy pre- or post –operatively.[3,4]

Maxillary sinus: Early stage (T1-T2) tumors can be resected by endoscopic maxillectomy or partial maxillectomy whereas advanced tumors (T3-T4) warrant a total/radical maxillectomy with or without orbital exenteration depending on orbital periosteum involvement with adjuvant radiation therapy.[3,4]

Lateral rhinotomy and midfacial degloving were the preferred approaches. Radiotherapy as a primary modality was considered in patients not willing or unfit for surgery. Surgery was followed by radiotherapy for recurrent tumours and those with positive margins. Hence surgery was the main modality of treatment for early staged tumours with radiotherapy reserved for recurrences. Combined modality in the form of surgery followed by radiotherapy is the treatment of choice for advanced tumours. The protocols followed in the management of these tumours were consistent with those seen in similar studies in literature.[4,5] Patients underwent modified radical neck dissection for T1, T2 tumours with positive nodes. We had two cases of chondrosarcoma of the maxillary sinus in stage IV who underwent total maxillectomy. Both of them had a parapharyngeal mass which was excised via transmandibular approach and histopathology reported as chondrosarcoma. One of the patient underwent radiotherapy following surgery for residual disease. One patient had olfactory neuroblastoma who underwent maxilloethmoidectomy and had residual disease in 1 month and was given radiotherapy and chemotherapy(CT)- cisplatin.

RECURRENCE: The recurrence rates were less(21%) compared to the previous studies which showed around 35-50 % recurrence within a median time of one year.[4,5] In view of the short period of follow up, the 5- year survival rates could not be calculated but the mortality rates were less compared with the previous studies.

CONCLUSION:
Most patients with paranasal sinus malignancies presented with locally advanced disease. Advanced T stage, regional and distant metastasis are highly predictive of poor survival. Hence early diagnosis and treatment is of immense value in decreasing the morbidity and mortality. Surgery is the main stay of treatment in early stages T1, T2 and surgery followed by chemoradiotherapy is the treatment of choice for advanced malignancies T3, T4 or radiotherapy can be given followed by salvage surgery for recurrence. Recurrence rate is high and typically occurs within the first year after treatment. Hence patients should be on regular follow up with nasal endoscopy and radiological imaging. Any recurrence or residual disease detected should be managed with radical surgery/ RT. The recurrence rates and mortality have decreased over a period of time probably due to increased efficacy of the diagnostic tools and treatment.

TABLE 1: HISTOPATHOLOGY	No:of patients	Maxillary	Maxilloethmoid	Frontoethmoid	Sphenoid	Sphenoethmoid
SCC	16	11	4	-	1	-
Adenocarcinoma	3	2	-	-	-	1
Adenocystic carcinoma	4	3	-	-	1	-
Rhabdomyosarcoma	3	3	-	-	-	-

Volume : 4 | Issue : 2 | Feb 2014 | ISSN - 2249-555X

Inflammatory myofibroblastic tumor	2	1	-	1	-	-
Chondrosarcoma	2	1	1	-	-	-
Acute myeloid leukaemia with sarcoma	1	1	-	-	-	-
Non Hodgkins Lymphoma	1	1	-	-	-	-
Olfactory neuroblastoma	1	-	1	-	-	-
Malignant melanoma	1	1	-	-	-	-
Sinonasal poorly differentiated carcinoma	4	3	-	-	1	-
Haemangiopericytoma	2	1	1	-	-	-
Secondary tumours	1	-	-	-	-	1
	41	28	7	1	3	2

TABLE 2: TUMOR STAGING	No. of patients	Maxillary	maxilloethmoid	frontoethmoid	sphenoid	sphenoethmoid
I	2	1	1	-	-	-
II	5	6	-	-	-	-
III	9	4	3	-	2	-
IV A	13	11	2	-	1	1
IV B	7	5	-	1	-	1
IV C	1	1	-	-	-	-
	40	28	6	1	3	2

TABLE 3: T STAGE	No. of patients	Surgery	Surgery -> Radiotherapy	Radiotherapy -> surgery	Radiotherapy	Chemotherapy	Surgery -> Radiotherapy -> Chemotherapy
T1	2	2	-	-	-	-	
T2	8	5	2	-	-	-	1
T3	10	1	7	-	1	1	-
T4a	16	4	10	1	-	1	-
T4b	4	1	3	-	-	-	-
	40	13	22	1	1	2	1

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