Rare tumors of sinonasal track

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Abstract

Objective To characterize the clinical behavior of rare sinonasal malignancies.

Methods Clinical data from the cases of rare sinonasal malignancies at Gujarat Cancer and Research Institute during 2001–2007 were extracted. Data for histologic type of tumor, tumor stage and survival were analyzed.

Results Eighty-nine cases of the non-squamous cell malignancy were identified. The mean patient age was 54 years. In this study, we found minor salivary gland tumor in 31 patients, sarcoma in 19 patients, spindle cell carcinoma (SpCC) in 19 patients, undifferentiated carcinoma in 9 patients, lymphoma in 6 patients and melanoma in 3 patients. Adenoid cystic carcinoma exhibited the best survival rate (3 years survival rate was 77%), whereas melanoma and undifferentiated carcinoma exhibited poor survivals (1 year survival was 25% and 33%, respectively and 3 years survival rate is 0%).

Conclusions Adenoid cystic carcinoma is the most common squamous cell carcinoma (SCC) of the sinonasal track. Survival for the patients with undifferentiated carcinoma and melanoma involving the sinonasal track is poor.

Keywords Sino nasal · Rare tumor · Clinical features

Introduction

Malignant tumors of the nasal cavity and paranasal sinuses constitute <1% of all malignancies and 3% of upper aerodigestive tract malignancies [1]. Because of the overall rarity of these tumors, development of an understanding of their clinical behavior, response to therapy, and overall prognosis may be difficult. Malignancies of the maxillary sinus constitute the largest fraction of these paranasal sinus and nasal cavity malignancies. Although the squamous cell carcinoma (SCC) is the most common malignancy of the sinonasal track, several other histologic types of tumor may be encountered as well. These include minor salivary gland malignancies, sarcomas, melanomas, undifferentiated carcinomas lymphoma and carcinosarcoma. The minor salivary gland tumor includes adenocystic carcinoma, mucoepidermoid carcinoma (MEC), etc. sarcoma includes osteosarcoma, chondrosarcoma, spindle cell sarcoma, rhabdomyosarcoma, Ewing's sarcoma, etc. and lymphoma includes non-Hodgkin's and Hodgkin's lymphoma [2].

Adenocarcinoma accounts for approximately 15% of sinonasal cancer with male predominant and peak age incidence between 55 and 60 years. It can arise from any site but the ethmoid sinuses are most frequently involved. Radical surgery is the treatment of choice and 3 years survival rate is 82% [1]. One percent of head and neck malignant neoplasm and 10% of salivary gland neoplasms are adenoid cystic carcinomas (ACC). This tumor frequently occurs in the fifth decade of life, usually affecting women. The tumor grows slowly, but neural invasion, distant metastases and multiple recurrences are common. This behavior means that ACC patients have a limited prognosis, even after radical surgery and radiotherapy [3].

MEC of sinonasal track arises from minor salivary gland over palate. Histologically MEC is classified in to grade I (low grade), grade II (intermediate grade) and grade III (high grade). Treatment of low grade is surgery only while intermediate and high grade required postoperative radiotherapy [4].

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Table 1 Incidence, TNM staging

Histology	Total patients	Mean age with range in years	M:F	Mean duration and range of symptoms in months	TNM stage and No. of patients
Adenocarcinoma	12	44 (30–65)	1.5:1	5 (3–6)	Stage II: 3 Stage III: 4 Stage IVa: 3 Stage IVb: 2
Adenocystic carcinoma	13	50 (30–73)	1.1:1	6 (1–12)	Stage II: 4 Stage III: 4 Stage IVa: 0 Stage IVb: 2 Stage IVc: 3
MEC	6	45 (18–54)	1:1	6 (2–12)	Stage III: 3 Stage IVa: 0 Stage IVb: 3
Adenosquamous carcinoma	1	36	M	4 months	Stage III: 1
Undifferentiated carcinoma	9	45 (18–71)	0.8:1	3 (1–6)	Stage IVa: 7 Stage IVb: 2
Carcinosarcoma	19	54 (42–66)	1.3:1	3 (2–12)	Stage II: 3 Stage III: 5 Stage IVa: 5 Stage IVb: 6
Rhabdomyosarcoma	3	12 (3–24)	2:1	1 (1–2)	Stage IVa: 1 Stage IVb: 2
Ewing's sarcoma	4	10 (3–23)	3:1	3 (1–4)	Stage IVa: 1 Stage IVb: 3
Osteosarcoma	6	35 (4–45)	2:1	4 (1–24)	Stage III: 2 Stage IVa: 1 Stage IVb: 3
Chondrosarcoma	1	60	F	2 months	Stage IVc: 1
Osteoclastoma	1	45	F	24 months	Stage III: 1
Angiosarcoma	1	4	F	3 months	Stage IVa: 1
Esthesioneuroblastoma	2	38	F	5 (3–7)	Stage IVb: 2
Chordoma	1	34	M	6 months	Stage IVa: 1
Malignant melanoma	3	34 (28–50)	2:1	3 (2–5)	Stage IVa: 1 Stage IVb: 2
NHL	3	24 (18–34)	2:1	3 (1–6)	Stage I: 3
Large cell lymphoma	3	24 (6–34)	2:1	1 (1–2)	Stage I: 3
Plasma cell myeloma	1	24	F	11 months	Stage I: 1

Table 2 Metastatic pattern, treatment intent of patients with rare tumors of maxilla

Histology	Lymph node involvement	Distal metastasis	Curative Rx	Palliative Rx	No Rx or incomplete Rx
Adenocarcinoma	–	–	9	2	1
Adenocystic carcinoma	–	3 lung	8	4	1
MEC	–	–	3	3	–
Adenosquamous carcinoma	–	–	1	–	–
Undifferentiated carcinoma	–	–	0	7	2
Carcinosarcoma	–	–	11	5	3
Rhabdomyosarcoma	–	–	–	2	1
Ewing's sarcoma	–	–	4	–	–
Osteosarcoma	–	–	3	3	0
Chondrosarcoma	–	Multiple bone metastasis	–	1	–
Osteoclastoma	–	–	1	–	–
Angiosarcoma	–	–	1	–	–
Esthesioneuroblastoma	–	–	–	1	1
Chordoma	–	–	1	–	–
Malignant melanoma	–	–	1	2	–
NHL	–	1 patients CSF +ve 5 months after CT	2	–	1
Large Cell Lymphoma	–	–	2	–	1
Plasma cell myeloma	–	–	1	–	–

Rx: Treatment; NHL: Non-Hodgkin's lymphoma

Clinically sinonasal undifferentiated carcinoma (SNUC) is characterized by extensive tissue destruction and frequent involvement of orbit and anterior cranial fossa. Histologically, SNUC is composed of pleomorphic cell with high nuclear-cytoplasmic ratio, arranged in nests, sheets, and trabeculae. In the recent report of 25 patients with SNUC, the median survival was 18 months [1]. The mean age of the patients of spindle cell carcinoma (SpCC) was 72 years and the female-to-male ratio was 1:2 [5]. It is the biphasic tumor that has also been referred to by names such as pseudosarcoma, carcinosarcoma, metaplastic carcinoma, and collision tumor, generally, accepted treatment of SpCC is wide local excision with negative margins. Radiotherapy

might be useful for sinonasal track tumors, in case with positive margin, and in patients who can not tolerate surgical excision. Overall 3 years mortality rates for SpCC vary between 29 and 45% [6, 7].

Rhabdomyosarcoma is a malignant neoplasm of primitive mesenchyme exhibiting skeletal muscle differentiation. Oral rhabdomyosarcoma is rare and accounts for only 0.04% of all head and neck malignancies. Treatment modalities include surgery, chemotherapy and radiation [8]. Ewing's sarcoma is the second most common malignant bone tumor of childhood, yet it is a rare tumor of sinonasal track. Primary maxillary localization is unusual and occurs in only 1–2% of cases. The prognosis of Ewing's sarcoma has been

Table 3 Treatment adopted in GCRI and prognosis of rare tumors of maxilla

Histology	Treatment and No. of patients	Surgery performed	Chemotherapy	Death due to disease	With disease at last follow up	Follow up mean and rang in month
Adenocarcinoma	Sx + RT : 9 CT + RT : 2	Partial Maxillectomy-2 TM + TMF/STG-7	Cis + 5-FU every every 21 days 4–6 cycle	2	2	36 (6–44)
Adenocystic carcinoma	Sx + RT : 8 CT + RT : 4	Subtotal Maxillectomy-3 TM + TMF/STG-5	Cis + 5-FU every 21 day for 4–6 cycle	0	3	40 (24–48)
MEC	Sx + RT : 3 CT + RT : 3	TM + TMF/STG-3	Cis + 5-FU every every 21 days 4–6 cycle	0	3	24 (12–36)
Adenosquamous carcinoma	Sx + RT : 1	TM + TMF-1	-	0	0	38 months
Undifferentiated carcinoma	RT : 2 CT : 2 CT + RT : 3	-	Cis + 5-FU every 21 days 4–6 cycle	4	5	6 (3–12)
Carcinosarcoma	Sx : 2 Sx + RT : 7 RT : 2 CT + RT : 5	TM + TMF/STG-7 TM + Eye ball Exentartation 1 CFR-1	Cis + 5-FU every 21 days for 4–6 cycle	0	5	38 (6–40)
Rhabdomyo-sarcoma	CT + RT : 2	-	VAC every 21 days for 1 year	1	2	3 (1–6)
Ewing's sarcoma	CT + RT + CT: 4	-	VACM and I + E alternately every 3 weekly for 48 weeks	0	0	40 (12–59)
Osteosarcoma	Sx + RT + CT : 6	TM + TMF/STG-4 TM + Eye ball Exentartation 1 CFR + VRAM flap-1	Cis. + Adr. every 3 weekly for 6 cycle	1	2	18 (1–36)
Chondrosarcoma	CT : 1	-	VACM and I + E alternately every 3 weekly for 48 weeks	0	1	
Osteoclastoma	Sx + RT : 1	TM + TMF-1	-	0	0	21 months
Angiosarcoma	Sx + RT : 1	TM-1	-	0	0	24 months
Esthesioneuro-blastoma	RT : 1	-	-	0	2	12 months
Chordoma	Sx + RT : 1	CFR - 1	-	0	0	12 months
Malignant melanoma	Sx + RT : 1 CT + RT : 2	Lower partial maxillectomy-1	Dac. + Temo. every month for 6 cycles	1	2	16 (2–20)

Table 3 Treatment adopted in GCRI and prognosis of rare tumors of maxilla

Histology	Treatment and No. of patients	Surgery performed	Chemotherapy	Death due to disease	With disease at last follow up	Follow up mean and rang in month
NHL	CT : 2	-	CHOP every 3 weekly for 6 cycle	1	2	24 months
Large cell lymphoma	CT : 2	-	CHOP every 3 weekly for 6 cycle	1	2	12 (6–18)
Plasmacell myloma	RT : 1	-	-	0	0	37 months

Sx: Surgery; RT: Radiotherapy; CT: Chemotherapy; Ca: Carcinoma; NHL: Non-Hodgkin's lymphoma; Cis: Cisplatin; Adr: Adriamycin; Dac.: Dacarbazine; Tem: Temozolamide; CHOP: Cyclophosphamide, doxorubicine, vincristine, prednesolone; VACM: Vincristine, adriamycin, cyclophosphamide, mesna; I+E: Iphosphamide, etoposide; VAC: Vincristine, actionomycin-D, cyclophosphamide; TM: Total maxillectomy; TMF: Temporalis muscle flap; STG: Split thickness skin graft; CFR: Craniofacial resection, VRAM: Vertical rectus abdominis musculocutaneous flap

improving considerably since the introduction of combined modality treatment. The estimated overall survival at 4 years is 75% [2].

Osteosarcoma is a malignant mesenchymal sarcoma characterized by the direct formation of bone or osteoid by tumor cells [9]. The most effective single therapeutic measure appears to be radical resection. If palpable nodes are present, neck dissection does apparently improve survival [10]. CT scan was superior to conventional radiography, including panoramic radiography, for detecting the extent of bone destruction and new bone formation in the lesion, allowing more accurate evaluation of the osteosarcoma [11]. Neutron beam irradiation results in significant loco regional control [1]. Chondrosarcomas are extremely rare tumors of which approximately 10% are found in the maxillofacial region. They are slow growing tumors which can eventually metastasize to the lungs; the peak prevalence occurs during the third to fifth decades without a sex predilection. Surgical excision is the only effective treatment for chondrosarcoma as they are resistant to radiation and chemotherapy. However, some authors have advocated postoperative high-dose radiotherapy. In overall terms, the prognosis in these patients is not good since the 5-year survival rate is <40% [12]. Radiation-induced sarcoma is a rare complication of radiation therapy [13]. Angiosarcoma is rare tumor develop in young age and surgery and postoperative radiotherapy is the treatment [1]. Chordoma is extremely rare, locally aggressive tumor originates from embryonic remnants of the notochord. Surgery is primary treatment and if not resected completely recurrence is common. Postoperative radiotherapy is required for advanced tumor and incompletely resected tumor [1].

Malignant melanomas develop from malignant transformation of melanocytes. The malignant melanomas

developing from turbinates or maxillary sinuses are extremely rare. Symptoms of nasal melanoma are generally non-specific, and patients often fail to seek prompt medical attention. Diagnosis can be confirmed by the immunochemical profile of staining with anti-S100 and HMB-45 antibodies, even in the case of melanin-free pigment melanoma [14]. The prognosis for patients with oral malignant melanoma is extremely poor. Earlier recognition of this condition simplifies treatment and greatly improves the prognosis for these patients [15].

Extranodal non-Hodgkin's lymphoma of sinonasal track is rare. Radiation therapy is the primary modality of treatment for localized disease, especially for low grade disease. Combination chemotherapy with or without radiation is used for more advanced disease and for intermediate and high grade lymphoma. Plasmacytoma of the jaw bones and oral cavity, as in other anatomic sites, comprises three distinct entities: Multiple myeloma, solitary plasmacytoma of bone, and extramedullary plasmacytoma. The most common radiographic finding was a radiolucency that many times was superimposed on the roots or apices of nearby teeth. The most common symptom was localized pain, and the most frequent clinical sign was a raised red lesion on the alveolar ridge. The disease affects older persons. Radiotherapy is the primary treatment [16].

Materials and methods

This is a retrospective study which was conducted in the Department of Surgical Oncology, Gujarat Cancer and Research Institute (GCRI), Ahmedabad and which covered a period of 7 years from January 2001 to December 2007. All patients having malignancies of sinonasal track other than SCC were

included in this study. SCC, non-neoplastic and inflammatory conditions were excluded. All information regarding history, clinical examination, investigation measures, treatment plan and follow up were extracted from hospital records and analyzed.

Result

Total 472 cases of sinonasal malignancy were reported in this institute during 2001–2007. Out of them 351 cases were of SCC of maxilla. Tables 1–3 shows incidence, clinical presentation, TNM staging, treatment done at GCRI and prognosis of rare tumor of sinonasal track.

Discussion

As a subset of maxillary sinus malignancies, non-squamous cell cancers of the sinonasal track are rare entities. Therefore, limited data exist in the literature regarding survival for patients with these lesions. In addition, the bone and soft tissues of the sinonasal track may give rise to several different histologic types of malignant tumors. These include sarcomas, melanomas, carcinomas, and minor salivary gland malignancies. Although staging and survival for patients with SCC of the sinonasal track have been well established, development of survival data for patients with non-squamous cell malignancies of the maxillary sinus is hampered by the rarity and variety of these malignancies. In many instances, small numbers of patients with non-squamous cell malignancies of the maxillary sinus have been aggregated with those with squamous cell cancers, rather than reported separately [17–21]. In this study we found some very rare type of tumor of maxilla like angiosarcoma, chondrosarcoma, chordoma, plasmacytoma, etc. Lymph node metastasis is uncommon in non-SCC. Distal metastasis is more common in non-SCC. It is not possible to separate the maxillary tumor from ethmoid because at the time of presentation, all most all tumors had already involved more the one subsite of sinonasal track.

The primary treatment is surgery except for advanced disease, lymphoma, and plasmacytoma rhabdomyosarcoma. Most of the patients are required postoperative radiotherapy as most of them have stage IVa disease at presentation. Three years survival rate of adenocarcinoma, adenocystic carcinoma, MEC are 77%, 72% and 50%, respectively. For undifferentiated carcinoma 1 year survival rate is only 25% and none of the patient is alive at the follow up of 3 years. Ewing sarcoma has 3 years survival rate of 100%. Esthesioneuroblastoma has over all good prognosis but in this study both the patients died before 1 year and the cause may be the advanced disease at presentation. Malignant melanoma had 1 year survival rate of 33% and none of the

patient is alive at the follow up of 3 years. Osteogenic tumor had 3 years survival of 50%.

Patient of angiosarcoma, chordoma and plasmacytoma were still alive at the follow up of 3 years. The prognosis of the non-SCC is very poor as 11 out of 89 died due to disease and 31 out of 89 patients had persistent disease at last follow up. The worst prognosis is of undifferentiated carcinoma, malignant melanoma and rhabdomyosarcoma. Salivary gland has good prognosis. In this study 31 patients were in stage IVb and stage IVc and had taken palliative treatment and 11 patients had not taken treatment or had taken incomplete treatment. Most of the patients had undifferentiated carcinoma, malignant melanoma and rhabdomyosarcoma. Many factors should be taken into account in the prognosis of non-SCC of maxilla, including the histological and clinical stages of the disease.

Conclusion

Because of the varied types of non-squamous malignancies rare malignancies of maxillary sinus, the prognosis of this group of malignancies varies with the subtype of tumor. Each of these tumors has their own unique presentation, growth pattern, presentation, prognosis which will determine the aggressiveness and type of treatment.

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