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Case Report:

Malignant Melanoma of Nose and Paranasal Sinuses: 2 Case Reports

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Abstract:
Malignant melanoma is one of the rare and highly aggressive diseases of the sinonasal cavity. High index of suspicion is required for diagnosis as the patient usually presents with non specific signs and symptoms. In the natural course of the disease, higher rate of loco regional recurrences and distant metastasis are seen making the overall prognosis of disease very poor. In reviewing the various treatment modalities used in the past, surgical resection of the tumour with postoperative radiotherapy is preferred one. Advances in surgery, radiotherapy and chemotherapy don't have any impact on improved survival, which remains poor in this disease. We report two cases of malignant melanoma, which were treated at our institute.
Key Words: Malignant Melanoma, Nose, Paranasal sinuses

Introduction:
Malignant melanoma is rare and aggressive tumour of nose and paranasal sinuses with high incidence of local recurrence and distant metastasis. They account for less than 1% of all melanomas and less than 4% of sinonasal tumours.[1] At intial presentation, the tumours are usually confined to the nasal cavity.[2] Diagnosis is often delayed because of onset of symptoms is insidious and non specific. Early diagnosis is important in management. Because of the rarity of disease, retrospective reviews and case reports are important source of gathering information about the course of disease, prognostic factors and response to treatment. Out of various treatment modalities, which have been used in the past, most of the surgeons prefer, surgical resection followed by postoperative radiotherapy.[3] Survival rates are generally poor in spite of aggressive treatment.

Case Reports:
Case I: A 50 year-old-female was referred to ENT OPD with history of nasal obstruction and occasional epistaxis for the last 3 months. Earlier patient presented to EYE OPD with the complaints of watering of left eye for 3 months, swelling medial to the left medial canthus for 3 months. On examination, there was epiphora of left eye, and a firm mass 1.0 X 1.5 cm medial to medial canthus causing lateral displacement of the canthus. Best-corrected visual acuity of the left eye was 6/9. Slit lamp examination showed a nuclear sclerosis of grade II and fundus was normal. The patient was treated as a case of sub-acute dacryocystitis for 1 month. On follow-up there was no improvement, hence, the patient was referred to us for evaluation. On examination of face irregular, elevated, tender swelling extending over the left side dorsum of nose and extending to medial canthus area was seen. Anterior rhinoscopy revealed fragile soft tissue mass in the middle meatus area. Left side of neck had a palpable lymph node 2 x 2 cm Ib (submandibular region). Diagnostic nasal endoscopy revealed fragile pinkish vascular mass, in the middle meatus area and septum, which bleeds on touch . Biopsy from mass confirmed the diagnosis of malignant melanoma, while FNAC from cervical lymph node was suggestive of amelanotic melanoma. Contrast Enhanced Computed Tomography (CECT) of the sinuses (Fig.1) revealed soft tissue mass involving left nasal cavity, left ethmoid cell region and causing erosion and destruction of lateral wall and protruding into the left orbit. Patient underwent excision of naso-orbital tumor mass by lateral rhinotomy approach with glabellar extension, and left side selective neck dissection. Patient was advised postoperative radiotherapy, which patient did not take. After 4 months patient presented with tonic-clonic seizures. Visual acuity was reduced to 1/60. There was a firm, tender, pigmented mass near the medial canthus 2 x 3 cm in size, proptosis, inferolateral globe displacement and fundus examination showed disc edema. CECT showed soft tissue mass in the ethmoid sinus extending into the left orbit, invading the extraocular muscles, optic nerve and maxillary sinus with erosion of cribriform plate and going intracranially, thereby suggesting recurrence of tumour. A craniofacial resection with left orbital exenteration was done Patient was given postoperative adjuvent chemotherapy (injection Dacarbamazine). Patient could tolerate only 2 cycles, before she expired.

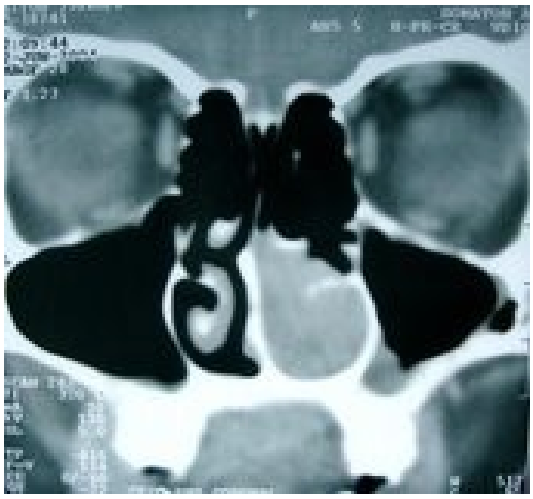

Fig.1 CECT of nose and paranasal sinuses showing soft tissue mass involving left nasal cavity, left ethmoid cell region and causing erosion and destruction of lateral wall and protruding into the left orbit.

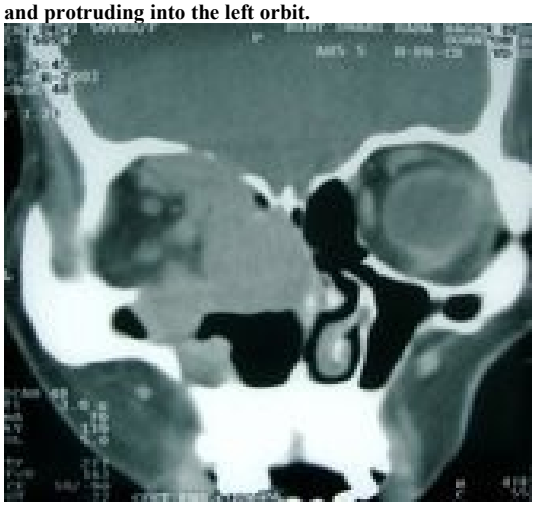

Fig.2 CECT Scan Nose and PNS showing mass confined to the left nasal cavity, septum and inferior turbinate

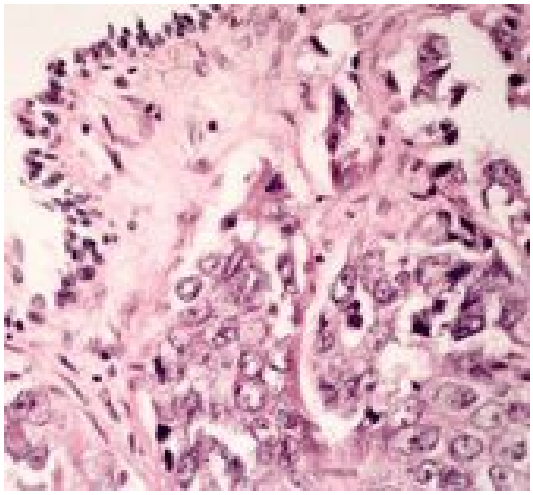

Fig.3 Microphotograph (H/E - 40x) showing features consistent with Malignant Melanoma

Case II: A 35-year-old female presented to ENT OPD with a 3-month history of recurrent epistaxis from right naris and gradually progressive nasal obstruction. Anterior rhinoscopy revealed pinkish, friable, polypoidal mass involving the right nasal cavity, septum, and inferior turbinate. Diagnostic nasal endoscopy was done, findings were confirmed, and biopsy was taken. Histopathology confirmed the diagnosis of malignant melanoma. Metastatic work up was within normal limits. CECT scan of nose and paranasal sinuses (Fig. 2) revealed enhancing soft tissue density mass lesion in right nasal cavity, inferior turbinate and septum. The patient underwent excision of the mass by lateral rhinotomy approach. Histopathology (Fig. 3) of the excised mass was consistent with malignant melanoma. Patient was given postoperative radiotherapy. Patient is under regular follow-up for more than one year without any local recurrence and distant metastasis.

Discussion:

In the head and neck, the incidence of primary melanomas is 25-30%.[4] However the incidence of mucosal melanomas arising from aerodigestive tract varies from 0.4 to 4%, out of which nose and paranasal sinus is the most common site.[5,6] Nasal cavity is being the commonest site of tumour presentation. Bridges et al[2] reported 70% of melanomas start in nasal cavity and 30% from paranasal sinuses. The incidence of the disease is roughly equal in both sexes, and predominantly affecting the age group over 60 years.[7] This is not in conformity in one of our case as the disease was seen in early thirties. Clinical presentation depends upon size and site of the lesion. Presenting symptoms include nasal obstruction, epistaxis, swelling of the nose, mass at the vestibule, epiphora, diplopia and proptosis.[7] Nasal obstruction with epistaxis are the two common symptoms with which majority of patients present.[3,8] Despite its lower incidence these tumours invade the orbit frequently because of easy access to the orbit from the ethmoidal sinus, given the thinness of lamina papaaracea.[9] One of our case presented with chief complaints of epiphora and swelling medial to medial canthus and non specific complaints of nasal obstruction and occasional bleeding. Patient was treated as a case of sub-acute dacryocystitis, and after persistence of symptoms after one month, the patient was referred to us for nasal endoscopy.

The presence of positive lymph node and advanced clinical stage at presentation has been implicated as poor prognostic indicators.[10] This is in conformity with one of our case where the patient presented with early local and nodal metastasis. The primary treatment modality is complete surgical excision with or without postoperative radiotherapy.[2] Although radiotherapy alone has been shown to result in complete or partial cure initially, neither radiotherapy nor chemotherapy, singly or combined, has been shown to affect overall survival. Chemothearpy should be reserved for patient with systemic disease. Temam et al[11] reported that postoperative radiotherapy improved local control, independent of T stage, but distant metastasis free survival and overall survival were worse when compared with surgery only group. Conversely, Kingdom and Kaplan[12] reported that postoperative radiation therapy was associated with better overall survival and better disease free intervals. They recommended postoperative radiotherapy in all patients and even those with negative surgical margin. Disease- related mortality is associated with local or regional recurrence. The lung, liver, bone, and brain are the most common site for metastasis.[7] In reviewing the previous series on sinonasal melanomas high rates of local recurrence (31%-85%) and distant metastasis (25%-50%) and poor 5-year survival rates (13%-45%) have been reported[8]. Close follow-up is warranted as local recurrence is common and resection of recurrent tumour may result in longer survival.

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