Research Paper

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Mucosal malignant melanoma of nasal cavity – a rare case

Medical Science

KEYWORDS : mucosal melanoma; nasal cavity; surgery

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ABSTRACT

Mucosal malignant melanomas of the head and neck region are very rare. We report a rare case of mucosal melanoma of the nasal cavity of a 42 years old male who presented with recurrent episodes of epistaxis. Subsequently, the patient was treated with surgery with wide margins followed by radiotherapy.

Introduction

Mucosal malignant melanoma of the head and neck region is a rare disease entity.[1.] Nasal cavity, paranasal sinus and oral cavity are common sites of mucosal melanomas of head and neck. The treatment of choice is surgical resection with wide surgical margins. Some patients are benefited by post-operative radiotherapy.[2] The main cause of treatment failure is local recurrence, followed by the development of metastases.[3] The prognosis of the patients with mucosal melanomas of nose and paranasal sinuses is very poor with low disease specific five year survival rate.[2, 3] A rare case of mucosal malignant melanoma of a middle aged male is presented here.

Case report

A 42 years old male patient presented with a chief complaint of recurrent episodes of epistaxis from the right nostril of six months duration. The frequency of epistaxis was initially episodic and reported to have occurred every 20-30 days but had gradually increased since the past 2 months. The patient also complained of a right nostril blockage since 4 months which had progressed to the current stage. He also reported a blackish mass in his right nostril which he had noticed around 2 month before the time of initial reporting.

A thorough clinicopathological examination was carried out. The diagnostic nasal endoscopy revealed a blackish mass originating from the floor of nose involving anterior part of nasal septum and mucosa of the inferior turbinate. On histopathological evaluation, the diagnosis of malignant melanoma of nasal cavity was arrived at.

CECT of paranasal sinuses demonstrated a localized mass in the right nostril originating from floor of the nose involving anterior part of nasal septum, inferior turbinate, middle turbinate, anterior ethmoids and subsequently blocking the infundibulum. Posterior ethmoidal sinuses, frontal sinus, sphenoid sinus and the base of skull were free of tumor. No evidence of regional lymphadenopathy was noted. Metastatic work up was negative.

The patient was subjected to surgery with wide excision of nasal mass which included medial maxillectomy, anterior ethmoidectomy, entire nasal septal excision and part of the hard palate excision along with floor of nose excision by lateral rhinotomy approach. A custom made palatal prosthesis was designed for the post operative palatal defect to which the patient responded well. The lesional tissue was sent for histopathological examination. The reports confirmed the preoperative diagnosis of malignant melanoma of the nasal cavity. All margins were reported to be free of the tumor cells. Consequently, adjuvant radiotherapy was given. The post operative healing was uneventful and the patient is on regular follow up.

Discussion

Mucosal malignant melanomas of head and neck are rare. They are more aggressive and often show a different behavior from the cutaneous melanomas. Mucosal melanomas have a neuroectodermal origin, thus their impact on tissues with endodermal origin such as the oesophagus, larynx, and nasopharynx is scarce.[5] Chang et al (1998) reported a very small percentage of mucosal melanomas in the head and neck region (0.7%) in a study on 84,836 patients of cutaneous and non-cutaneous melanomas of the entire body.[1] Nasal cavity forms the most common site in the head and neck mucosal melanomas where the most common sub site reported is nasal septum (41%), followed by the middle turbine (29%), the inferior turbine (23%), the lateral nasal wall (7%), and the bottom of nasal fossa (1%).[4,5] In our case, the malignant melanoma was arising from the floor of nose which is a rare site. The most common symptom reported by the patients is epistaxis with or without nasal obstruction. Symptoms such as proptosis, diplopia, facial pain, and facial asymmetry are more frequent in the advanced cases, arising in the paranasal sinuses.[5]

Sino-nasal mucosal melanoma is primarily a disease of the adults and the elderly. Majority of cases in the English literature have reported the mucosal melanomas in the age range of 20 to 80 years with the most of cases are above 50 years of age.[6, 7]. There is an equal sex predilection with a slight preponderance of males in some studies. Nasal mucosal melanoma is however, more commonly seen in males. [6,8-11] In our case, a 42 years old male was diagnosed with mucosal melanoma.

Mucosal melanomas are characterized by being multiple, having satellite formations and an early angiolymphatic invasion. This results in advanced loco regional recurrences and early metastasis with a high mortality rate for the disease. Mucosa of the nasal cavity and Para nasal sinuses metastasizes less frequently to regional lymph nodes but distant metastasis to lung and brain are common.[5] Lymph node metastases are evident at the time of initial presentation in only 5.7% of the reported cases.[12] In our case, there was no evidence of regional lymphadenopathy or distant metastasis.

Due to rarity of this disease and absence of prospective studies, there are no clear cut guidelines for the management of sinonasal malignant melanomas. Surgery is the preferred treatment which depends on the location of the tumor and its extension. The most commonly used approaches are para lateronasal rhinotomy, craniofacial resections, and endoscopic surgery.[13] Prophylactic resection of lymph nodes is not recommended.[2,12] In our case, we operated the patient by paralateronasal rhinotomy approach with wide resection of mucosal melanoma followed by adjuvant radiotherapy. Post-operative radiation may be beneficial in the advanced tumor and cases of sin nasal melanomas because these are often resected with ''close'' margins owing to the anatomy of the region.[14]. Its use following surgery does seem to achieve local tumor control and patient survival. Immunohistochemical studies have shown the mucosal melanomas to be positive for S-100, HMB-45 and negative for cytokeratin expres-

sion. The immunotherapy and chemotherapy in the diagnosed melanoma cases have been used only with partial success.

The high rate of local recurrence and risk of developing metastases are the key reasons elucidating the low survival rate in melanoma subjects.[3] Improved survival depends on the invention of better systemic therapies aimed at a targeted treatment.

Conclusion

Sino nasal mucosal malignant melanoma is a rare but an aggressive disease. Despite advances in radiological, surgical and adjuvant therapeutics, the mean survival of patients with sinonasal melanomas has not improved since the past three decades. Surgery followed by adjuvant radiotherapy still remains the treatment of choice. The paucity of reported cases of mucosal melanomas and poor survival rate due to an early loco regional recurrence and distant metastases necessitates an increased awareness in the clinicians worldwide to report additional cases in literature. This would augment a better understanding of the lesion and aid in discovery of newer treatment strategies which would facilitate the improvement of outcome of the disease.

Figure 1. Intra operative photograph of mucosal melanoma of right nasal cavity

Figure 2. Enbloc resected tumor

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