PRIMARY MUCOSAL MALIGNANT MELANOMA OF MIDDLE EAR – A CASE REPORT

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Abstract: Primary mucosal malignant melanoma reported in this case is a rare clinical entity. Very few cases of primary mucosal malignant melanoma of middle ear have been reported in the literature till date. The etiopathology, clinical features and management of this disease has been described here.

Keywords : Primary mucosal malignant melanoma, Middle ear.

INTRODUCTION

Malignant melanoma is a rare malignancy primarily affecting the skin. It could be either cutaneous or mucosal. Mucosal malignant melanoma, especially of head and neck is an extremely rare entity. Very few cases of primary middle ear malignant melanomas have been reported in the world literature till date. Here is another example of this clinical rarity

CASE REPORT

A 63 years old female presented in the E.N.T. Department with complaints of Tinnitus and blockage of the left ear of three months duration. On routine clinical examination of the ear, patient had a reddish brown mass in the middle ear, seen through the intact and bulged tympanic membrane; rest of the E.N.T. examination was normal. Nasal endoscopy revealed fullness around the Eustachian tube of left side. Otomicroscopy confirmed the clinical findings and depicted an intact bulging drum with reddish brown mass having irregular margins and the mass was non pulsatile. (Fig. 1)

There was no history of ear ache or ear trauma. Pure tone audiometry was suggestive of 45 dB conductive hearing loss. Impedance audiometry was suggestive of middle ear mass lesion. CT scan of temporal bone showed mass in the middle ear cavity extending up to the Eustachian tube eroding inferior and anterior

bony boundaries of middle ear cavity. CT scan showed lesion extending upto nasopharynx, though nasal endoscopy did not reveal the extension.

Exploratory tympanotomy was done under general anaesthesia and biopsy material was sent for histopathological examination. Histopathological examination showed extensive infiltration of large elongated polygonal or irregular cells, with pleomorphic deep staining nuclei having increased mitotic activity. The cells were predominantly of the epitheloid type with prominent nucleoli, and containing abundant intra and extra cytoplasmic melanin there by suggestive of malignant melanoma, which was further confirmed by Masson Fontana and Melanin bleach histochemical test.

Careful dermatological examination revealed no other lesions. The fact that symptomatology and physical findings were only confined to the ear, which was supported by relevant investigations and that there was no involvement of skin elsewhere, diagnosis of primary mucosal malignant melanoma of the middle ear was established.

Seven cycles of Radiotherapy comprising each of radiation dose of 150 Gy/day was given for 5 days in a week. The Chemotherapy consisted of Dacarbazine (220mg/m²) and Cisplatin (25mg/m²) iv daily for 3 days every 3 weeks, Carmustine (150mg/m²) iv every 6

Fig.1: Otomicroscopy showing intact bulged drum with reddish brown mass inside the middle ear.

Fig.2: CT - Scan showing mass lesion in middle ear cavity, extending to nasopharynx.

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weeks, for a total period of 6 months. Patient didn't respond to the complete and adequate course of Radiotherapy and chemotherapy. Therapy was monitored by otomicroscopy, nasal endoscopy, pure tone and impedance audiometry.

Gradually there was worsening of symptoms with addition of new symptoms like epistaxis and headache. Later on patient developed progressive facial palsy, trismus and features of intracranial extension such as severe headache, vomiting, convulsions and papilloedema. Patient died after 18 months of completion of radiation because of wide metastasis and intracranial extension.

DISCUSSION

Melanoma originates from the neural crest – comprising of melanocytes, which are normal pigment cells present in the epidermis and sometimes in the dermis. 15-20% of all melanomas occur in the head and neck (I.W. Sherman)³. Only 0.5 to 0.3 percent of all cases of malignant melanomas originates from the mucous membrane of the head and neck (Hesham)². Within upper respiratory tract the commonest site of occurrence is the nose and paranasal sinuses with decreasing incidence in the lower parts of the respiratory tract. (I.W. Sherman)³.

There are significant racial difference, the incidence being highest in Japanese and Africans but cutaneous melanoma is uncommon in them. Mucosal melanoma of the head and neck occurs more frequently between the ages of 40-70 years. with a slight male preponderance (Snow and Vander Waal 1986)⁴.

Melanocytes are distributed through out the mucous membrane of head and neck. The gross appearance of the lesion depends on the site of its origin. The nasal lesions are usually polypoidal while palatal lesions are flat. Usual histological findings are same as described in the case report and can be confirmed histochemically by Masson Fontana and Melanin bleach tests. The epithelium from the affected part shows a marked increase in dysplastic melanocytes, in situ. Though Malignant changes and areas of invasion of the underlying tissue, is suggestive of its primary origin, yet it is difficult to differentiate primary and secondary lesions.

Because of the extreme rarity of the disease, different biological behaviour of the disease is not very well known. So conclude malignant melanomas are difficult to treat and prognosis of primary mucosal malignant melanoma is very poor because owing to its aggressive course and unpredictable response to various

modalities of treatment like radio therapy and chemotherapy, wide excision or combination of both and wide spread metastasis. Some authors suggest wide surgical resection as the treatment of choice for both cutaneous and mucosal melanoma (snow)⁴. However in many cases, this is not feasible and not advisable due to its extension.

For many years Melanoma was considered as a radio resistant tumor. However now there are evidence that radiotherapy with high dose fraction is an effective modality and can be an adjuvant to chemotherapy (Habertmalz)¹. Reports of Snow and Vander Wall (1986) and Batsakis (1982) suggests better five years survival rates of nasal lesion. Total five year survival rate is approximately 20-30 percent.³

CONCLUSION

Malignant melanoma it self is a rare condition and mucosal malignant melanoma of middle ear is still a rarer entity. Diagnosis is confirmed by histopathological findings. Primary mucosal tumours are more difficult to treat as compared to cutaneous one because of its aggressive clinical course, wide spread metastasis and unpredictable response to treatment. however, wide surgical resection with or without adjuvant radiotherapy remains the treatment of choice.

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