Primary Melanoma of Infra Temporal Fossa: A Case Report

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Abstract This is the second case report in English literature of a primary non-cutaneous non mucosal melanoma (NCNM) originating from Infratemporal fossa (ITF). A 21 year woman who was previously treated for craniopharyngioma 14 years back with surgery and radiation, presented with left ITF swelling which after excision was histologically confirmed to be NCNM. She has been disease free till date. Therapeutic difficulties and diagnostic dilemmas in such a case are discussed. We propose the term of non cutaneous non mucosal melanoma (NCNM) for this rare entity.

Keywords Primary melanoma · Infratemporal fossa · Non cutaneous non mucosal

Introduction

Malignant melanoma is the most aggressive form of skin cancer. Although it accounts for less than 5 % of all cutaneous

malignancies, it is responsible for more than 75 % of skin cancer-related deaths [1]. The age-adjusted incidence has increased more than 200 % from 1975 to 2008 [2]. Melanomas are clinically classified as cutaneous, primary meningeal, uveal, and mucosal; cutaneous being the most common. Here we present a case of primary non cutaneous non mucosal melanoma (NCNM) of infratemporal region. Review of the English literature showed only one case report till now at this site.

Case Report

A 21 year old woman who was diagnosed as a case of craniopharyngioma at age of 6 years and treated primarily by right pterional craniotomy with decompression of a cystic mass in sellar and suprasellar region presented at the Head and Neck Clinic of Regional Cancer Centre (RCC) in October, 2012 with complaint of swelling at the left temporal region. Further enquiry revealed that in April, 1998, she had received post operative radiation (RT) of 45 Gy. She was on regular follow up and was asymptomatic for the past 14 years. CT scan showed 3.4×2.5 cm heterogeneously enhancing soft tissue density in left temporal fossa (Fig. 1).

FNAC from this lesion was suggestive of round cell tumor. Wide excision of the lesion was done under General Anaesthesia; it was a fleshy mass infiltrating temporalis muscle, however, bone and skin were free. The gross specimen measured 4.5×3.5×1.5 cm. Cut section of the growth was grey-white, myxoid. Microscopy showed a cellular neoplasm composed of nests and sheets of cells separated by fibrovascular septae (Fig. 2a). Individual cells were oval /spindly with moderate eosinophilic cytoplasm, vesicular nucleus, many showing prominent nucleoli. Some of the cells

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Fig. 1 CT image showing heterogeneously enhancing soft tissue density in left ITF

showed intracytoplasmic brownish pigment (Fig. 2b). With a provisional diagnosis of malignant melanoma, immunohistochemical panel of markers including S100, HMB45 and cytokeratin (CK) was done. The tumor cells showed diffuse strong positivity for S100 (Fig. 3a), focal positivity for HMB45 (Fig. 3b) and negative staining with CK. Mean while a section from the tumor was also subjected to bleach. The brownish pigment was removed with bleach, confirming melanin. Correlating histopathology and immunohistochemical findings a diagnosis of malignant melanoma was made. The closest margin had clearance of 2 mm.

In view of previous radiotherapy for craniopharyngioma, further RT was not given. Patient and relatives opted for close follow up to avoid morbidity of re do surgery.

After 9 months, she presented with mild pain at local site and low backache. CT scan showed asymmetric soft tissue thickening suggestive of post operative changes or recurrent disease. Ultrasonography (USG) neck showed left level 2 node with preserved fatty hilum, FNAC was negative for malignancy. Serum LDH was 638 (313–618 U/L). X-ray dorsolumbar spine was

normal and so was bone scan. Chest x-ray was normal. USG abdomen showed a celiac artery node measuring 10×15 mm. After appropriate symptom control, she was advised to continue close follow up.

Discussion

Conley et al. [3] in 1964 classified tumors of ITF. Majority of the lesions are contiguous from adjacent anatomical sites where as few are purely primary. Metastasis from elsewhere are rare. This classification system is practical and universally accepted. Among primary tumors of infratemporal region, adenoid cystic carcinoma is most common followed by squamous cell carcinoma, fibrosarcoma, hemangiopericytoma, histiocytosis-X, Hodgkins disease, osteosarcoma and rhabdomyosarcoma.

Although malignant melanoma of nasopharynx or maxillary sinus may encroach into the ITF, primary melanoma of ITF is an absolute rarity. An extensive search

Fig. 2 **a** Section from the neoplasm showing sheets of atypical cells. (H&Ex400). **b** Atypical cells with prominent nucleoli and intracytoplasmic brownish pigment. (H&Ex400)

Fig. 3 **a** Atypical cells showing diffuse strong positivity for S100. (IHCx400). **b** Atypical cells showing focal positivity for HMB45. (IHCx400)

of literature showed only one case of primary NCNM melanoma of ITF which was treated with local excision and post op RT and the patient was reported as disease free till 1 year of follow up [4].

In this present report, the patient was a case of craniopharyngioma managed with surgery and post op RT who after 14 years of asymptomatic follow up presented with the ITF swelling leading to a suspicion of sarcoma. The diagnosis of melanoma on final pathology report was a surprise. History of previous radiation therapy doses of >15 Gy in childhood has been implicated in the onset of future melanomas [5]. As this patient had received 45 Gy radiation, it could perhaps be the risk factor here.

Although more than 90 % of melanomas have a cutaneous origin, occasionally it is discovered first as a secondary deposit in a lymph node or visceral site, without evidence of a primary. This entity is known as melanoma of unknown primary (MUP) which was first described by Das Gupta in 1963 [6]. In his study he excluded patients fulfilling any of the following criteria: (1) patients with metastatic melanoma with evidence of previous orbital exenteration or enucleation; (2) patients with metastatic melanoma with history of excision, electrodesiccation or cauterisation of a mole, birthmark, freckle, chronic paronychia or skin blemish; (3) patients with metastatic melanoma in one of the node-bearing areas who presented a scar of previous local treatment in the skin area drained by this lymphatic basin; and finally (4) patients with metastatic melanoma who did not have a thorough physical examination including ophthalmoscopy and adequate examination of the anus and the genitalia.

The incidence of MUP varies considerably between the different studies, ranging from 1.2 % [7] to 18 % [8]. Various theories have been postulated to explain MUP such as the spontaneous regression of the primary site due to involvement of immunological mechanisms. The first one proposed by Smith and Stehlin in 1965 support the disappearance of a primary lesion as a result of spontaneous regression after metastasis has occurred [9]. Partial or complete spontaneous resolution of melanoma cells from the primary site is fairly common and it has been actually described in various studies [10].

Second theory assumes that melanoma originates in lymph nodes or other subcutaneous tissues or in viscera supported by the observation that ectopic benign nevus cells have sometimes been found in lymph nodes and other tissues [11]. Third, a synchronous unrecognised melanoma and fourth possibility, a previously excised or traumatically removed melanoma have been proposed as explanations. The most likely explanation for MUP remains to be immune induced regression of the primary tumour [12].

Surgery is the principle treatment for the management of malignant melanoma of the head and neck, with local excision of the primary tumor and operative resection of the involved lymphatic basins. Radiation therapy is offered to selected patients, and systemic therapies are indicated for selected stage II patients, most stage III patients, and for the majority of stage IV patients. As to date no systemic therapies have been proven to significantly improve overall survival (OS) in randomized trials in patients with stages I to III melanoma. For cutaneous melanoma, the 5 year survival drops from 97 to 40 % when disease progresses from early stage to regional nodes [13]. Melanoma of mucosal surfaces of the head and neck represents an aggressive form of the disease with a high propensity for regional spread and distant metastasis. Margin control in these patients is difficult due to the proximity of the tumors to critical neurovascular and bony structures and is associated with adverse outcomes. The addition of adjuvant irradiation improves local-regional control, but not OS. For all mucosal sites in the head and neck, 5-year survival is approximately 25 % [14,15].

Conclusion

Although non cutaneous melanomas of head and neck have been reported from upper aerodigestive tracts, this is the second case report of primary melanoma originating from infratemporal region with a disease free interval of 9 months and no imaging or histologic proof of recurrence. Further

follow up is needed to assess the natural history of disease due to paucity of literature about this rare entity.

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- Non cutaneous non mucosal origin of melanoma is absolutely rare
 - Immunohistochemistry confirms diagnosis
 - Management is on the lines of those of cutaneous/mucosal melanoma
 - It is supposed to have aggressive course
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Authors' Contributions RKS: Did the literature search and prepared the manuscript.

NL: analysed and edited manuscript.

BTV: overall supervision, concept and design, preparation of final manuscript.

KRA: provided pathology slides and review.

All authors read and approved the final manuscript.

Competing Interests The authors declare that they have no competing interests.

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