A rare instance of intraoral malignant melanoma in a young female

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Abstract:
Intraoral malignant melanoma is an extremely rare aggressive neoplasm constituting 0.8 to 1.8% of all oral malignancies and 0.2-8% of all melanomas. Mucosal melanomas tend to present at a higher stage, are more aggressive, and in a vertical growth phase of disease.

Key words: Melanoma; Oral Malignancies; Mamata General Hospital; Khammam

Introduction
Intraoral malignant melanoma is an extremely rare aggressive neoplasm constituting 0.8 to 1.8% of all oral malignancies and 0.2-8% of all melanomas [1,2]. Mucosal melanomas tend to present at a higher stage, are more aggressive, and in a vertical growth phase of disease [1,3]. The pre-existing melanoses seemed to be the most common precancerous lesion of the oral mucosa [4]. The diagnosis is often missed or delayed for long period because of its asymptomatic nature and inaccessible site [5]. The clinical details of a patient with an intraoral mucosal melanoma will be described and discussed for its rarity.

Case Report
A 28 years old female patient was referred to Mamata General Hospital OPD, with an intraoral pigmented patch present since her childhood. It started as a small (pea sized) brownish black pigmented, asymptomatic patch, initially over the proximal part of the hard palate and gradually progressed to the border of upper gums (Fig.1a,b,c). Mucosal biopsy carried out from the patch 10 years back showed no evidence of malignancy. Minimal discomfort with gradual increase in size of the lesion for the last one year. None of her family members had history of similar features.

Her intraoral examination revealed an asymmetric, bluish black pigmented patch of 8x6 cm
size with irregular borders over anterior one-thirds of the hard palate extending up to the upper gums with no evidence of bleeding or ulceration. Pigmentation is patchy interspersed with three small brown macules seen over lower gums. Bilateral, firm, discrete, nontender submandibular lymphadenopathy is present. Her upper jaw is conspicuous with overcrowding of teeth. Apart from acne vulgaris on her face, no other cutaneous and systemic abnormalities were present.

Routine haematological tests were normal. Chest X-ray, abdominal scan, CT brain and FNAC of submandibular lymph node showed no evidence of metastasis. Whereas, histological examination of lesional mucosal biopsy confirmed the diagnosis of malignant melanoma with abundant intracytoplasmic melanin pigment, nuclear atypia and mitoses infiltrating through the entire thickness of the mucosal and submucosal tissue. (Fig.2)

Patient was referred to MNJ cancer institute, Hyderabad for further management of melanoma, where resection of the entire lesion including palate was advised. However, the patient refused to undergo surgery and failed to report for further followup.

Discussion:

Intraoral malignant melanoma is an extremely rare condition, predominantly affecting the palate (32%) followed by maxillary gingival [1,5,6]. Malignant melanoma can also involve other mucosal sites like anal, vaginal, nasal, laryngeal, conjunctival, and sinus mucosas [7]. Blacks, Japanese and Asian patients are more affected than whites [1,8]. Unlike cutaneous melanomas, the oral lesions are more common in men than in women [8]. Oral melanoma presents as asymmetric pigmented macule with irregular borders which may bleed or ulcerate. Pre-existing melanosis occurs in about one third of all patients with oral melanoma [7]. Such macular hyperpigmentation probably represents a radial growth phase of the tumor, and it may persist for years before submucosal invasion occurs. Differential diagnosis includes oral melanotic macule, physiological pigmentation, nevus melanocanthoma and for amelanotic melanoma angiomata and other vascular lesions, giant cell granuloma, ill fitting denture, fibrosarcoma and carcinoma are to be considered [5,8]. Melanocyte markers like S-100, HMB-45, Melan-A helps to differentiate from other conditions [5,8]. Our patient had the pigmented lesion since her childhood. The lesion was ignored due to its intra oral (concealed) site and asymptomatic nature which progressed to extend up to the gums. Irregular margins, patchy pigmentation, gradual increase in size and associated swelling in the gums has raised the suspicion of malignancy and a mucosal lesional biopsy was performed, which confirmed the diagnosis of malignant melanoma. A radical local surgery has been advised.

Mucosal melanomas show far more aggressive behavior as compared to skin melanomas.6 They are more inclined to metastasize into regional and distant sites with a rate of 5– 48% of regional and 4–14% of distant dissemination, with 5-year survival rates reportedly ranging from 0% to 55%.9 Surgical excision with tumour free margin is the treatment of choice [1,3,5]. However, complex anatomy of the oral cavity makes complete surgical excision difficult. Histological staging of Clark’s levels (Clark et al. 1975) does not have any relevance to tumors of oral cavity due to different histological structure of mucosa and skin [10].

Poor prognostic factors include advanced clinical stage, thickness more than 5mm, vascular invasion and nodal and distant metastasis. Aggressive nature, tendency for metastasis, difficulty in surgery, grave prognosis, indicates the importance of high clinical suspicion and prompt biopsy for pigmented lesions in the oral cavity to avoid delay in the diagnosis.

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Fig.1a: Note bluish black, irregular, variegated pigmented patch present over the roof of hard palate and upper gums

Fig.1b: Note bluish black, irregular, variegated pigmented patch present over the roof of hard palate and upper gums

Fig.1c: Irregular pigmented lesions over upper gums

Fig.2. Histological examination of lesional mucosal biopsy showing intracytoplasmic melanin pigment, nuclear atypia and mitoses infiltrating through the entire thickness of biopsy confirmed the diagnosis of malignant melanoma.