Primary Malignant Melanoma of the Tongue

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The oral cavity is a rare location for the development of primary malignant melanoma. The most common primary lesion sites are the palate and gingiva. Melanoma of the tongue is specifically uncommon. A 66-year-old woman was referred to our clinic with a complaint of a huge, painless, black, discolored mass on the right side of the oral tongue for 7 years. There were no cutaneous lesions suggestive of malignant melanoma over the rest of her body. The biopsy of the tongue lesion revealed a histopathology consistent with primary malignant melanoma. Computed tomography of the neck showed no significant cervical lymphadenopathy. Chest radiograph, whole body bone scanning, and abdominal sonography revealed no definite distal metastatic lesions. She received composite resection of the tumor on the right side of the tongue and right functional neck dissection. The patient had an uneventful recovery and received regular follow-up examinations. She was free of disease for more than 2 years. The treatment principle for primary tongue melanoma is wide surgical excision. Early diagnosis will be promoted by careful oral examination and early biopsy of pigmented and non-pigmented masses. We reviewed the published reports in the English literature since 1970 and fewer than 30 cases of primary tongue melanoma were presented. We present a case report and a review of the relevant literature. (Chang Gung Med J 2002;25:764-68)

Key words: malignant melanoma, tongue.

Primary malignant melanoma of the oral cavity is a rare neoplasm. The incidence of oral cavity melanoma is about 0.2% to 8% of all malignant melanoma cases. Primary lesions arising from oral mucosa occur most frequently on the maxillary gingiva and palate, with the lips following as the third most common in frequency. Melanoma of the tongue is specifically uncommon and represents less than 2% of all oro-nasal melanoma cases. A review of the literature revealed fewer than 30 cases of primary malignant melanoma of the tongue had been reported and a tumor size of more than 6 cm was even more unusual as discovered in our review.

CASE REPORT

A 66-year-old woman was referred to our clinic with the complaint of a painless mass on the right side of the oral tongue. She had been aware of the dark discolored mass for 7 years; however, it gradually increased in size within the past 6 months. On examination, a black, pigmented and ulcerated mass measuring approximately 6 cm in size was found on the right side of the tongue with floor extension (Fig. 1). There were no cutaneous lesions suggestive of malignant melanoma over the rest of her body. The biopsy of the tongue lesion revealed a histopathology consistent with primary malignant...
melanoma. Computed tomography of the neck showed a right anterior lateral tongue mass with high intensity after contrast enhancement (Fig. 2). There was no significant cervical lymphadenopathy. Chest radiograph, whole body bone scanning and abdominal sonography revealed no definite distant metastatic lesions. She received a composite resection of the tumor on the right side of her tongue and right functional neck dissection. The histopathological findings revealed a malignant melanoma characterized by neoplastic proliferation of epithelioid to spindle melanocytes with melanin deposits and underlying skeletal muscle invasion. Scattered tumor cell nests were also present in the overlying squamous epithelium, suggesting that the tumor was a primary rather than a metastatic lesion (Fig. 3). The resection margin and base of the tumor were clear and no evidence of metastasis was found in the tissue of the functional neck dissection. The patient had an uneventful recovery and received regular follow-up examinations. She has been free of disease for more than 2 years, with no clinical or biochemical evidence of metastasis.

DISCUSSION

The mucosal membranes are rare sites for primary malignant melanoma. The presence of melanocytes in the mucosal membrane of respiratory, alimentary and urogenital tracts explains the occurrence of malignant melanoma in these sites.\(^1\) Melanoma of the oral cavity mucosa is a distinctly

Fig. 1 A black, pigmented and ulcerated mass measuring approximately 6.5 cm was found on the right side of the tongue with floor extension.

Fig. 2 Computed tomography of the neck showed a right anterior lateral tongue mass with high intensity after contrast enhancement.

Fig. 3 Histopathological findings of malignant melanoma. The tumor presents as a protruding mass composed of sheets of epithelioid to spindle-shaped neoplastic melanocytes. The tumor cells arranged in whorling fascicles or nests with focal melanin deposition. There are also scattered tumor cell nests noted in the squamous epithelium (arrows) (H&E, 100x).
rare occurrence with an incidence of 0.012/105 for combined primary and metastatic lesions to oral cavity. The tumors are commonly found in patients older than 40 years and there are no significant differences between genders. The oral cavity may be a site of predilection for melanomas in Japanese, although it is very rare in the white populations. We reviewed the reports in the English literature and fewer than 30 cases of primary malignant melanoma of the tongue were found. Men were more commonly affected than women in primary malignant melanoma of the tongue which was in contrast to skin melanoma where the incidence between genders was roughly equal.

Oral pigmentation preceded the development of malignant melanoma in about a third of the patients. Takagi et al. reported that mucosal melanosis was associated in 66% of oral melanoma, pre-existing in 36.2% and concurrent in 29.8%. There are many situations to be considered in the clinical differential diagnosis: Tattoos, melanotic macules, Laugier's disease, melanocytic nevus, drug intake, some vascular lesions, and oral pigmented lesions associated with endocrine disorders or different syndromes. Our patient had had the oral dark pigmentation for 7 years, but she did not pay much attention to it. We suggested that a deep biopsy should be performed on any intra-oral pigmented lesions with the tendency of malignant transformation.

Oral melanomas may present as flat, painless, dark brown or black discoloration macules or nodules, sometimes with erythema or ulceration. As the disease progresses, bony erosion is common. A very important point in the management of malignant melanoma of the oral cavity is to exclude the possibility of it being a metastasis from a cutaneous melanoma. This is because metastasis plays a large role in determining the goals and method of treatment. In the histopathologic distinction, Billings et al. found that all metastatic lesions lacked evidence of junctional activity in the overlying mucosa and showed no epidermal migration. This is in contrast to primary lesions, in which 44% and 38% had junctional activity and epidermal migration, respectively. A unique feature seen in the primary lesions (25%) was the presence of extensions of the melanotic pigment into the minor salivary glands. However, these findings may be inconsistent, and the diagnosis of a primary oral mucosal melanoma requires the careful search for and exclusion of any suggested cutaneous or mucosal lesions. For this patient, there was no history of melanoma-like lesion excision. We did not find any cutaneous lesions suggestive of malignant melanoma over her body, extremities, head or neck; there were not any pigmented lesions in the nasal cavity, pharynx and larynx. The histopathological findings revealed scattered tumor cell nests that were also present in the overlying squamous epithelium, suggesting that the tumor was a primary rather than a metastatic lesion. Physical examination and histopathologic findings suggested the diagnosis of primary melanoma.

The immunohistochemical profile of oral malignant melanoma was similar to that of cutaneous melanoma, with the exception that no oral malignant melanoma was positive for cytokeratin. HMB-45 are regarded as showing greater specificity for melanoma than S-100 protein. The immunoperoxidase stains of our patient showed positive finding in S-100 protein and HMB-45 stains.

Surgery is believed to be the most effective treatment for melanoma. Wide resection with a surgical margin 2 to 5 cm is necessary for cutaneous melanoma, but is difficult to achieve for oral melanoma because of anatomical reasons. Our patient received a composite resection of the tumor on the right side of the tongue and right functional neck dissection. The histopathological findings revealed no evidence of metastasis and the resection margin was clear. A series of studies showed no evidence of distal metastasis. The role of radiotherapy is controversial because many authors believe melanoma to be a radioresistant neoplasm, and it is frequently used for palliation. Radiotherapy and chemotherapy play an important role in the primary management of unresectable diseases. Because the resection margin was clear and no loco-regional recurrence or distant metastasis have been found till now, our patient do not received any radiation therapy or chemotherapy. In recent years, immunological therapies have been used. The most widely used cytokines are interferons and interleukin-2. However, immunotherapy has not improved survival or local regional control rates in patients with mucosal melanoma.

In general, the prognosis for patients with oral malignant melanoma is poorer than that for patients.
with cutaneous lesions. The 5-year survival rates were 6.6% to 20%. Several factors may contribute to this poor prognosis including lack of symptoms early in the disease, difficulty in achieving wide radical excision because of anatomic limitations, and rich blood supply that may facilitate hematogenous spread.

Early diagnosis will be promoted by careful oral examination and early biopsy of pigmented and non-pigmented masses. Early diagnosis and treatment will improve the prognosis of patients with oral malignant melanoma.

REFERENCES

舌部原发性恶性黑色素瘤

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原发於口腔的恶性黑色素瘤在临床上是非常少见的。口腔黑色素瘤最常见的是颈部和肩颈部，颈部其次；舌部的黑色素瘤则极为罕见。本科经历一名66岁的女性患者，主诉舌部右上一巨大、无痛的黑色斑块已有七年，因此到本院求诊。除上和其它身体部位并不存在类似黑色素瘤且病理组织切片证实为恶性黑色素瘤。临床检查发现并无法明確颈部淋巴结转移。胸部X光、腹部超音波和全身骨骼扫描显示没有转移的迹象。患者经医师广泛的术前评估，行手术切除，术后至今逾2年，无复发迹象。原发性舌部恶性黑色素瘤的治疗上，尚宜以广泛性手术切除为主。为了早期诊断，对于任何口腔中的色素性和非色素性肿瘤均需切片检查。回顾自1970年以来的文献文献报告，舌部原发性恶性黑色素瘤的个案数少于30个。本文提出一个新的案例并做文献之回溯。(長庚醫誌 2002;25:764-68)

關鍵字：恶性黑色素瘤，舌頭。