A synchronous presentation of malignant melanoma and dermatofibrosarcoma protuberans in a Nigerian female: A case report

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A B S T R A C T

INTRODUCTION: The simultaneous occurrence of multiple primary malignancies in a patient has been widely reported. Oral mucosal malignant melanoma is a rare lesion of the oral cavity. The occurrence of this lesion with a very rare skin sarcoma at the same time has never to the best of our knowledge been reported previously.

PRESENTATION OF CASE: A 64-year-old Nigerian woman with a 27-year history of a dark lesion in the right maxillary buccal and palatal mucosa with an anterior abdominal swelling of 24 years which was discovered on systemic examination. Both lesions were later discovered to be malignant melanoma and dermatofibrosarcoma protuberans respectively. She had a right hemimaxillectomy for the maxillary lesion and wide excision of the abdominal wall lesion, followed by six courses of chemotherapy. She has been undergoing regular reviews for the past 5 years and remains free of all malignancy; however, she died shortly after noticing the reappearance of both lesions.

DISCUSSION: While evaluating patients with a tumor, it will be rewarding to rule out other similar swellings in the body. This is important to avoid metastasis or multiple primary malignancies, which can help prolong patient life.

CONCLUSION: The synchronous occurrence of two different neoplastic lesions is rare. Our literature search revealed no previous report of such occurrence of oral malignant melanoma and DFSP at the same time in one patient. The importance of thorough examination of patients cannot be overemphasized as concurrent lesions may be missed.

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1. Introduction

Malignant melanoma (MM) is a neoplasm of melanocytic origin that arises from the proliferation of neural crest cells in the basal layer of the oral mucous membrane hence, they are virtually found at all sites and organs where neural crest cells migrate [1]. The vast majority of head and neck melanomas are cutaneous melanoma followed by oculary and mucosa types including sinonasal cavity, oral cavity, pharynx, larynx, and upper esophagus in descending order of frequency [2].

The oral variant is quite rare and represents 0.2–8% of all melanomas, 1–2% of all oral malignancies, and also accounts for 1.6% of head and neck malignancies [1]. Oral malignant melanomas (OMM) occurs in all races but it is found to occur more in countries like Japan, Uganda, and India [3]. It preferentially affects the hard palate, maxillary alveolar mucosa, and gingiva with a slight male predilection [1, 4, 5]. Published age of occurrence of OMM is between 30–90 years of age with a higher incidence in the 6th decade [4, 5]. In Nigeria OMM account for about 0.6% of all melanomas [6, 7]. A WHO report put the global annual incidence at 160,000 and death rate of over 45,000 [8].

Dermatofibrosarcoma protuberans (DFSP) is also an uncommon skin malignancy [8, 9, 10]. It is considered an intermediate neoplasm with a locally aggressive and slow-growing pattern and rarely metastasizes [9, 11]. DFSP has a yearly incidence of only 0.8 cases per million and presents commonly at mid-adult life with a slight male predominance [11]. Clinically, it appears as a slow growing nodular mass of the skin on the trunk or proximal extremities [9, 11].

The simultaneous occurrence of two different tumor entities in the same patient is a rare occurrence.

The authors report a case of a 64-year-old female managed in the oral and maxillofacial surgery department of the University of Nigeria Teaching Hospital Ituku/Ozalla, Enugu, Nigeria.

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1.1. Aim

This case report is aimed at highlighting the possibility of multiple tumors occurring at the same time in an individual; emphasis should be laid on proper examination while reviewing the patient. Several treatment options are available and they depend on the presentation.

2. Presentation of case

A 64-year-old female from the southern part of Nigeria presented with a one-year history of a dark painless ulcerated mass on the palate. She had had an incisional biopsy done on a 27-year-old dark leathery palatal patch one year before presentation to our clinic which was reported as a pigmented epitheloid melanocytoma. There was no associated medical condition, nil history of tobacco use in any form, and did not take alcohol and nil family history of similar conditions.

Clinical examination reveals an ulcerated, melanotic nodular mass on the right side of the hard palate with irregular surfaces. The swelling extended from the upper right lateral incisor to the upper last molar (buccally) and from the gingival margin of the upper right lateral incisor to the soft palate (palatally). It measured about 5 cm by 3 cm and was firm in consistency. No palpable cervical lymph nodes were found.

Abdominal examination revealed a 24-year-old firm, round, painless, non-tender mass that was slowly enlarging located on the anterior abdominal wall (supraumbilical wall) which measured about 4 cm by 4 cm. The swelling was attached to the skin but not to the underlying structures, there were no palpable lymph nodes and, internal organs were normal on palpation.

Magnetic Resonance Imaging (MRI) of the skull showed a localized lesion in the right hemi palate extending vertically into the antrum with features suggestive of malignancy. Differential diagnosis of oral mucosa malignant melanoma was made.

She had a right hemi maxillectomy and excisional biopsy of the abdominal wall mass performed simultaneously under general anesthesia by the lead maxillofacial surgeon who had both local and foreign exposure and both specimens sent to a pathology department in the United Kingdom on the patient’s request. A diagnosis of pT4aNxMx stage IVa OMM and DFSP pT2, N0, M0 respectively were made (Figs. 1–3).

She was subsequently referred for 6 courses of chemoradiotherapy sterilization of the margins and any occult tumor while awaiting the result of the biopsy. The chemotherapy regimen used was Cisplatin 50 mg, Vinblastine 5 mg, Decarbazine 1200 mg, and Vincristine 2 mg all via intravenous route and were well tolerated. Five years after treatment patient was clinically free of any tumor. However, during the sixth year post-surgery, her condition suddenly deteriorated when both lesions were noticed to have recurred. She passed on before we could offer further treatment. This work has been reported in line with the SCARE 2018 criteria [12].

3. Discussion

The occurrence of Multiple Primary Malignancies (MPM) has been known for centuries; it can involve a single organ or many organs at the same time [13]. The North American Association of Central Cancer Registries (NAACCR) classified MPM into Synchronous and Metachronous. Synchronous malignant lesions are two or more primary carcinomas co-existing at the same time of diagnosis in a patient [13]. Warren and Gate [14] based its diagnosis on three criteria; each of the tumors must have a definite feature of malignancy, each must be histologically distinct and should not be a metastasis of the other.

OMM and DFSP are both rare malignancies; their simultaneous occurrence is very uncommon. To our knowledge, this is the first reported case involving OMM and DFSP. The closest of similar cases seen by the authors we saw in the literature was that of DFSP and
Merkel Cell Carcinoma occurring simultaneously in a 74-year-old woman [9].

Synchronous neoplasms may occur at any age; however, literature suggests that these patients appear to be older than those with only one tumor [15,16]. Although the association between the two lesions may most likely be coincidental, long exposure to environmental causative agents, hereditary factors, and reduced immunity from long-standing malignancy may increase the risk of other primary neoplasms occurring [9,16]. Whereas the etiology of oral mucous melanoma largely remains unknown, denture irritation, use of tobacco, alcohol, race, exposure to sunlight, and many other etiologic factors have been suggested as possible causes of palatal melanoma, however, evidence to support this remains scarce [8,15,17]. Epitheloid melanocytoma is a rare low-grade variant of melanoma with indolent behavior and a relatively good prognosis but is known to have the potential to transform to melanoma which appears to agree with our case [18,19]. DFSP may affect patients of all ages but it is uncommon during childhood and mainly occurs between the ages of 20–40 years of age [9]. It is known to involve the dermis and subcutaneous tissue and has asymmetric growth and pseudopod-like extensions which make it difficult to completely resect which accounts for its high rate of recurrence which ranges from 10 to 80% [9,11]. This tumor is considered to be of low-grade malignancy [9]. Microscopically, it appears as monomorphic proliferation of spindle cells which present as a whorled pattern, evidence of mitotic activities may be present occasionally [11]. It can transform into Fibrosarcoma DFSP which has a high malignant and invasive potential even though death from this tumor is rare [9].

Late presentation of this patient with advanced (stage IVa) oral mucosa melanoma may have been due to the asymptomatic nature of the tumor, coupled with its rapid growth rate and the thinking that the initial diagnosis of epitheloid melanocytoma did not require aggressive treatment [18].

The cause of the rapid growth of the earlier lesion was not clear however, trauma from the biopsy and its longstanding duration may have contributed.

For both malignancies, surgical resection with free margins was the main treatment option along with adjunct chemoradiation to achieve further local tumor control [5,9,10]. The use of regional chemotherapy as a potent therapy for tumor shrinkage has also been reported as a treatment modality [20].

4. Conclusion

OMM and DFSP are uncommon tumors of the skin found commonly in the elderly. The synchronous occurrence of these two tumors has never been described. Wide resection and combination chemo radiation are the best treatment options.

Declaration of Competing Interest

The authors report no declarations of interest.

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Ethical approval

Ethical approval was sort and obtained from the University of Nigeria Teaching Hospital Health Research Ethic Committee with number NHREC/OS/01/2008B-FWA00002458-IRBO00002323.

Consent

“Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.”

Author contribution

All of us participated in the management of the patient and while writing this case Drs Okechi and Uguru did the write up. Drs Onwuka and Umeanuka contributed to the literature reviews and we all read through the work and make corrections.

Registration of research studies

N/A.

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