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Case Report

# Gingival metastasis of epithelioid sarcoma: A case report

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ABSTRACT

Metastatic epithelioid sarcoma to gingiva is extremely rare. One case has only been reported in the literature. We describe our experience for a rare case of female gingival metastasis, which the primary lesion occurred on the volar surface of the forearm and has been treated with wide surgical resection, chemotherapy, and radiotherapy 24 months earlier. Histological and immunohistochemical features of tumor cells led to the diagnosis of epithelioid sarcoma.

## 1. Introduction

Epithelioid sarcoma is a rare of malignant soft tissue neoplasm commonly involves flexor surfaces of fingers, hands, and forearms. It generally presents as a painless firm solitary or multiple nodules. The tumor mainly afflicts in teenagers and young adults that are likely to be disoriented with a variety of benign and malignant conditions, especially a granulomatous process, ulcerating squamous cell carcinoma, and synovial sarcoma. Despite its indolence and slow growth, the prognosis is poor because of high recurrence and early metastasis via lymphatic and hematogenous [1,2]. The principle common sites of epithelioid sarcoma metastasis are lungs, lymph nodes, and scalp, respectively [2]. Likewise, oral metastatic tumors are uncommon and comprise approximately 1% of malignant oral neoplasm which the most primary sites are from the breast, lung, and kidney [3,4]. In the oral cavity, furthermore, the most common site of metastatic lesions is mandible with the molar area, followed by premolar area [5]. Therefore, distant metastasis to gingiva is extremely rare [6], we can misdiagnosis in early clinical finding and delay proper treatment. The purpose of this report is to raise awareness of this uncommon entity by describing the clinical, radiographic and pathologic findings. The patient was informed and consented that the case would be submitted for publication.

## 2. Case report

A 31-year-old woman with no medical history presented to the department of orthopedics with a three-month history of a painless mass in the mid-forearm and developed another mass in the distal forearm in a month after that. No lymph node enlargement when diagnosis and follow-up patient. She was completely investigated with magnetic resonance images (Fig. 1) and biopsy. Results of technetium-99 radionuclide whole body bone scan, computed tomography scan of the chest and abdomen were negative. The preliminary pathologic report was fibromatosis then she underwent wide surgical excision. The post-operative pathologic report was epithelioid sarcoma and not free margin. Histologically reports showed aggregation of epithelioid tumor cells arranged in a solid pattern, tumor cell showed polygonal shaped with hyperchromatic vesicular nucleoli without clear demarcation (Fig. 2). Adjuvant chemotherapy (adriamycin and ifosfamide) was offered and followed by radiotherapy.

Two years later, she developed the ulcer and painless mass in the right maxillary attached gingiva extended to alveolar mucosa around anterior teeth and buccal surfaces around the molar area which multiple, erythematous nodes, granular surface, and extension of the lesion from the labial to the palatal gingiva. The involved teeth were the second degree of mobility, vitality test was positive, and sensation of the upper lip is normal (Fig. 3).

Panoramic radiography and a magnetic resonance imaging (MRI) revealed the mass in pterygoid muscle involved inner cortex of maxilla and right central incisor to canine (Fig. 4).

The pathologic report from an incision biopsy at the oral lesion was the metastatic epithelioid sarcoma. Histologically reports showed squamous mucosa with infiltration of sheets of epithelioid cells in the underlying submucosa. The neoplastic cells contained hyperchromatic, pleomorphic nuclei and increased mitosis (Fig. 5). The histological difference from the primary site was a less epithelioid tumor cell, appeared monolayer and seen vascular proliferation. The

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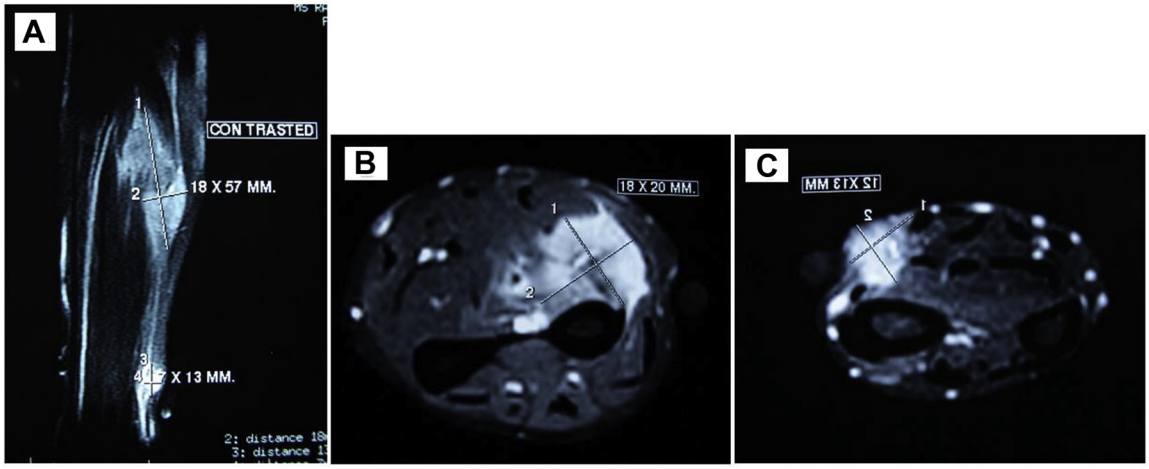


**Fig. 1.** Magnetic resonance images (MRI) of forearm taken at the initial visit. (A) Contrast-enhanced fat-suppressed coronal T1-weighted MRI of two forearm masses. (B) Contrast-enhanced fat-suppressed axial T1-weighted MRI of 18 × 20 mm. forearm mass. (C) Contrast-enhanced fat-suppressed axial T1-weighted MRI of 12 × 13 mm. forearm mass.

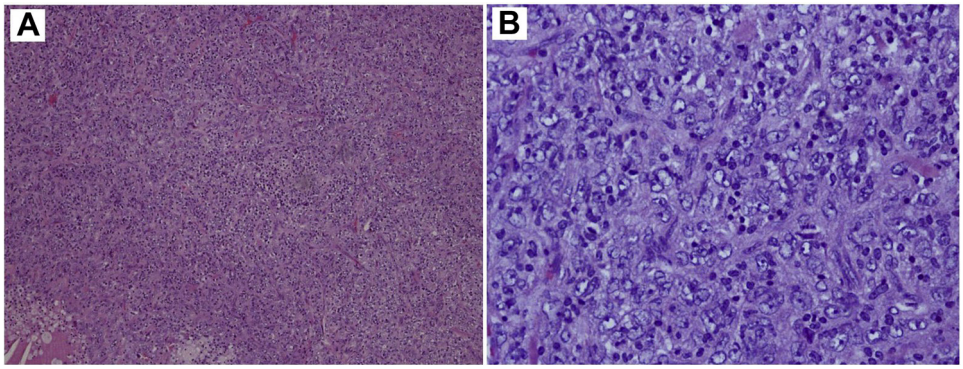


**Fig. 2.** Histopathological Image. (A) ×100 and (B) ×1000.

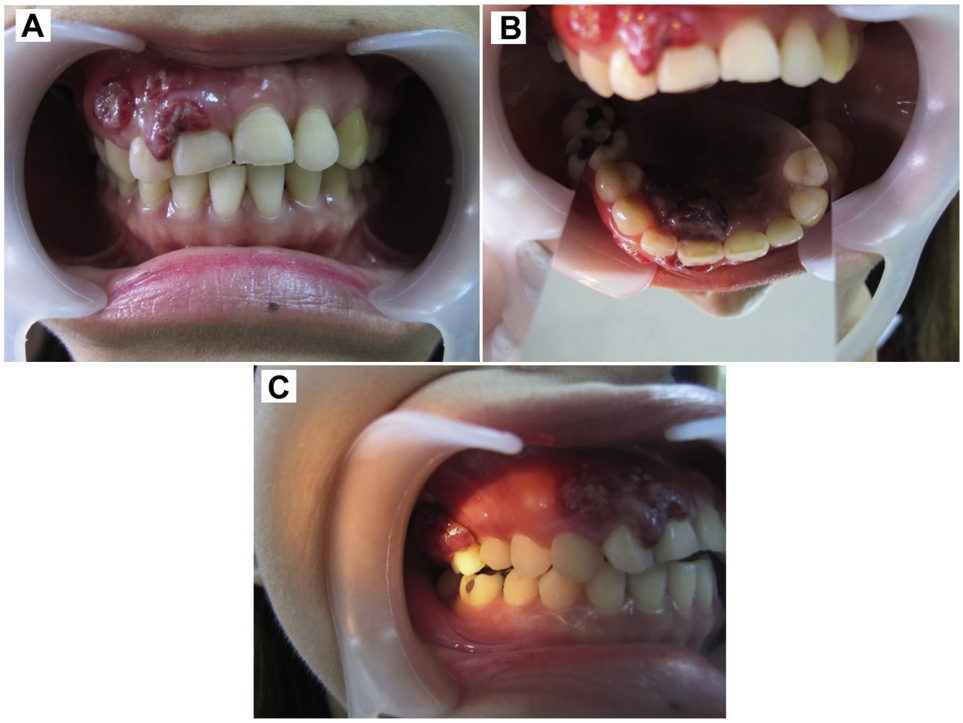


**Fig. 3.** Oral findings. (A) Frontal view of attached gingival masses on the right maxillary anterior teeth area. (B) Upside down view of attached gingival masses on the right maxillary anterior teeth area. (C) Lateral view of attached gingival masses on the right maxillary anterior and posterior teeth areas.

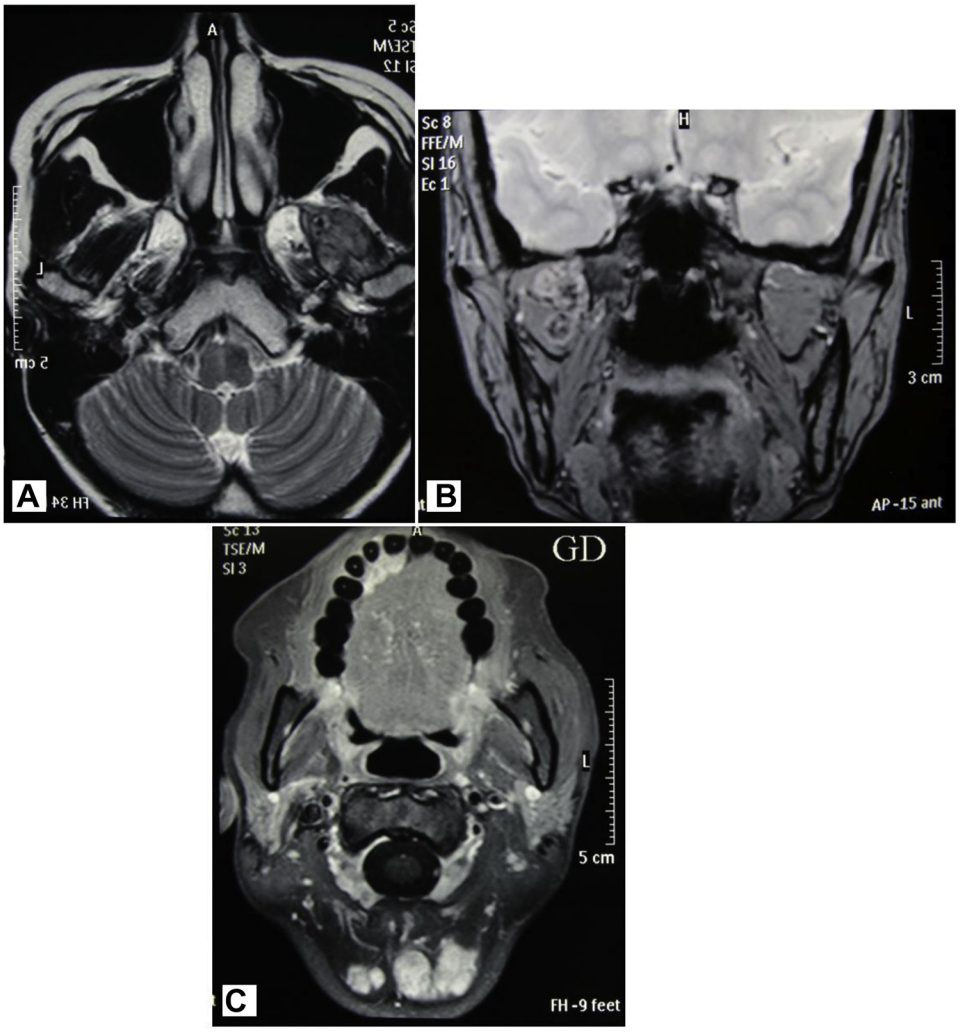


**Fig. 4.** Magnetic resonance images (MRI) of oral and maxilla. (A) Axial T2-weighted MRI of maxilla. (B) Coronal T2-weighted MRI of maxilla. (C) Contrast-enhanced fat-suppressed T1-weighted MRI of maxilla.

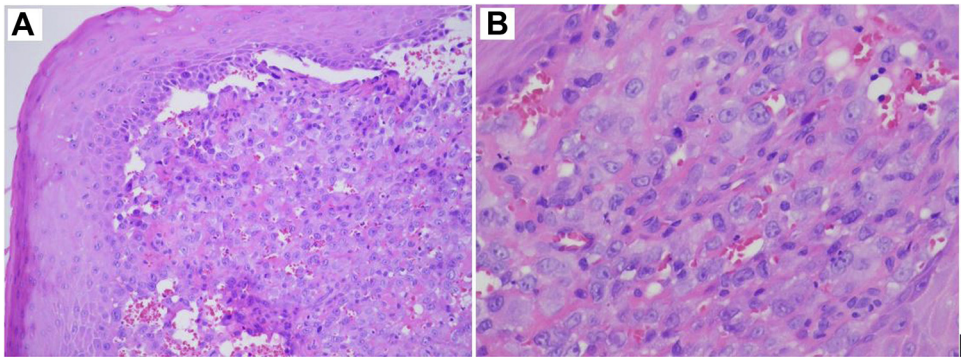


**Fig. 5.** Histopathological Image. (A) ×100 and (B) ×1000.

immunoperoxidase staining was positive for AE1/AE3, CD34, and epithelial membrane antigen (EMA) (Fig. 6) marker, also negative for smooth muscle actin (SMA) and S100 protein (Fig. 7) marker.

After the diagnosis of metastasis, the patient denied any further treatments except palliative care and she passed away after oral metastases for 6 months.

## 3. Discussion

The epithelioid sarcoma is a soft tissue malignant tumor of uncertain type. There are two principal types which are the classic form (distal type) mainly occurs in young adults involving the distal extremities as this case, a 31-year-old woman presented with a painless mass in the mid-forearm. Histologically, the classic type of epithelioid sarcoma has a distinct nodular arrangement of the tumor cells and epithelioid appearance with cytoplasmic eosinophilia. Most epithelioid

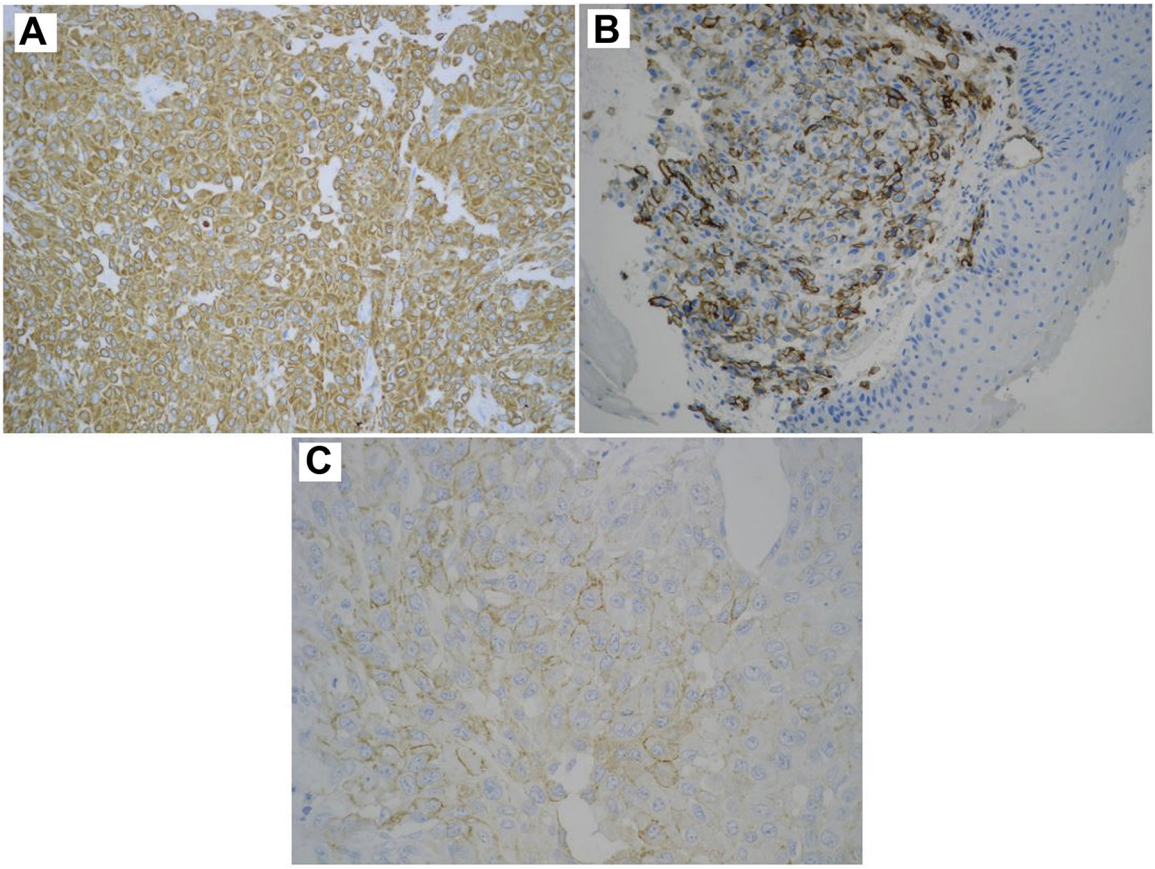


**Fig. 6.** Positive marker of immunoperoxidase staining in oral metastatic tumor. (A) AE1/AE3 (B) CD34 and (C) EMA.

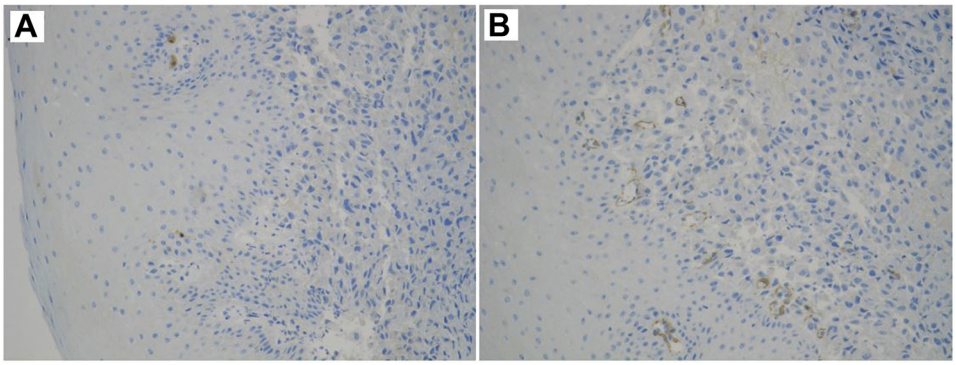


**Fig. 7.** Negative marker immunoperoxidase staining in oral metastatic tumor. (A) SMA and (B) S100.

sarcomas stain for EMA, low- and high- molecular weight cytokeratins and vimentin [8,9]. The rare form (proximal type) called large-cell tends to be more aggressive and usually affects in adults that arises in the deep parts of the pelvis, perineum, and proximal extremities. It consists of large epithelioid carcinoma-like rhabdoid cells that were intermediate between an epithelioid sarcoma and rhabdoid tumor and present with marked cytologic atypia, frequent mitosis, vascular invasion [7]. The overall survival and metastases-free survival are worse in proximal type.

Epithelioid sarcoma has a high likelihood of local recurrence (77%), pulmonary (51%), lymph node (34%) and scalp (22%) metastasis, respectively. The metastasis can be a direct extension along tendons, fasciae, nerves or periosteum and to lymph nodes or vascular invasion. It also related to location (axial vs appendicular) and limb location, size, depth, hemorrhage, mitotic figures, necrosis, vascular invasion, age, gender were identified as prognostic factors [2]. Epithelioid sarcoma tends to metastasis in early stage either via subdermal lymphatic vessels or hematogenous spreading, the tumor was associated with a higher incidence of lymph node metastasis compared with other soft tissue sarcomas [10]. From lymph node metastasis, the tumor cell may secrete vascular endothelial growth factor-C (VEGF-C) and VEGF-D that induce growth of lymphatic vessels (lymphangiogenesis) via activation of VEGF receptor-3 (VEGFR-3) localized on the surface of lymphatic endothelial cells [11,12]. From hematogenous spreading to gingival, tumor cells may be metastasis by the rich capillary network of the gingiva. The chronic inflammation of gingiva may be the good environment of the progression of the metastatic cells [13]. In molecular features, evidence of loss of tumor suppressor SMARCB1 protein expression and SMARCB1 gene deletion in the majority of epithelioid sarcoma cases [14].

In all cases, the multi-modality approach is the best which surgical treatment should combine with radiotherapy and multi-agent

chemotherapy. Metastatic epithelioid sarcoma to gingiva is extremely rare. Kao et al. reported the only one case of the metastatic epithelioid sarcoma to gingiva [15]. The 57-year-old man with primary lesion occurred on the wrist and was treated 18 months earlier by surgery and radiotherapy and then developed the painful granulomatous lesion on his right mandibular gingiva. They confirm the diagnosis by immunohistochemical and electron microscopic examination. The oral metastatic lesion was resected and controlled by chemotherapy. Others two reported of metastatic epithelioid sarcoma was tongue. The first case reported a 64-year-old man presented with epithelioid sarcoma of his right hand and developed metastasis to the tongue after the amputation 2 years later [16]. The second case reported a 45-year-old man with right axillary epithelioid sarcoma and developed metastasis to the tongue 4 months later [17]. Both of them developed pulmonary metastasis.

With respect to sarcomas other than epithelioid sarcoma metastatic in the oral-maxillofacial region. The Alveolar soft part of brachioradial muscle metastatic to tongue [18], angiosarcoma metastatic to mandible [19,20], Undifferentiated spindle-cell sarcoma of gluteal, elbow and lower extremity metastatic to mandibular gingiva and tongue [21,22], chondrosarcoma of femur metastatic to mandibular gingiva [21], Leiomyosarcoma of uterus metastatic to tongue [23] and myxofibrosarcoma of thigh metastatic to tongue [24]. From the literature, the tongue is the most common site of metastatic sarcoma.

To the best of our knowledge, we presented the second case of metastatic epithelioid sarcoma to gingiva which was the first case reported picture of metastasis and radiology. So, the purpose of this report is to increase awareness of this extremely rare entity. When the patient presented with the lesion at gingiva and history of epithelioid sarcoma, we should suspect metastatic lesion.

## Conflict of interest

The authors have no financial relationship to disclose.

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