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Case report

Cutaneous angiosarcoma masquerading as photodermatitis: a case report

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Abstract

Angiosarcoma (AS) is a rare and aggressive soft tissue sarcoma originating from endothelial cells, with cutaneous manifestations often seen in the head and neck region. Despite its rarity, AS poses significant diagnostic challenges due to its variable presentation and ability to mimic other dermatological conditions. We report the case of an 87-year-old female that presented with a 4-month history of an asymptomatic nodule on her neck, which rapidly progressed into an indurated plaque spreading to her face, chest, and scalp. Initially misdiagnosed as cellulitis and dermatitis, the lesion was unresponsive to antibiotics and steroids. Imaging showed extensive infiltration in the neck, precluding surgical resection. This case underscores the diagnostic difficulty of AS, which can be mistaken for benign skin conditions. Despite a multidisciplinary approach, the prognosis for AS remains poor, with a 5-year survival rate of approximately 35%. Treatment options include surgery, radiation, chemotherapy, and immunotherapy tailored to the patient's condition and tumor characteristics.

Keywords: angiosarcoma, dermatology, dermatopathology, skin cancer, vascular

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Introduction

Angiosarcomas (AS) are rare soft tissue sarcomas originating from endothelial cells, known for their poor prognosis (1). The majority of cutaneous AS lesions arise in the head and neck region (2). Due to the diagnostic complexity and aggressive nature of the tumor, optimal management requires a multidisciplinary approach. Imaging techniques such as computed tomography (CT) and magnetic resonance imaging (MRI) can help assess the extent of disease, although they are not reliable for differentiating soft tissue sarcomas from other benign or malignant tumors (3). A biopsy with immunohistochemical staining is recommended to confirm the diagnosis. Early detection of cutaneous AS is challenging due to its ability to mimic various skin conditions. This case report highlights the diverse clinical features of cutaneous AS and provides a list of reported presentations it can imitate. To date, we believe this is the first reported case of angiosarcoma mimicking photodermatitis.

Case report

An 87-year-old female, with a known history of diabetes and hypertension, presented to our dermatology clinic. She had no personal or family history of cancer, radiation therapy, or lymphedema. She reported a 4-

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month history of an asymptomatic flesh-colored nodule on her neck, which progressed over 2 months into a coalescing plaque. The lesion spread centrifugally, extending inferiorly to her chest and superiorly to the right side of her face, forehead, and scalp, eventually reaching the left side of her face. Prior to presenting at our clinic, she was diagnosed with cellulitis, dermatitis, and photosensitive dermatoses, and she was treated with a 2-month course of oral antibiotics, topical, and oral steroids, but she showed no improvement.

On clinical examination, there were well-defined erythematous subcutaneous nodules coalescing into an indurated plaque on a background of erythematous, woody, hard, non-pitting edema extending from the jawline to the chest (Fig. 1a). In addition, there was a well-defined erythematous violaceous bruise-like patch over the face, forehead, scalp, cheeks, and nasal bridge, sparing the nasal tip and perioral area (Fig. 1b). A few violaceous and black nodules were observed just below her right earlobe and scalp. There was also marked edema of the lips and lower right eyelid, leading to eyelid closure and restricted eye movement (Fig. 1b). No lesions were found in other photodistributed areas, and no mucous membrane involvement was noted. The lymph nodes were not palpable due to skin rigidity, and the lesions bled easily. Two punch biopsies were performed, and the patient was sent for laboratory and radiological tests.

Complete blood cell counts, basic biochemistry results, and inflammatory markers were within normal limits, except for an elevated lactate dehydrogenase (LDH) level of 325 U/l (normal range 0–247 U/l). Histology from two punch biopsies taken from the postauricular nodule and chest revealed anastomosing blood vessels with dense hypercellular proliferation of spindle-shaped and epithelioid malignant cells, featuring enlarged, pleomorphic, and hyperchromatic nuclei, some with prominent nucleoli (Fig. 2). The tumor was vascularized with irregularly shaped blood vessels and hemorrhagic areas, along with foci of tumor necrosis. Atypical mitotic figures were seen. Immunohistochemical staining showed that the target cells were positive for cluster of differentiation (CD) 31 and CD34, and negative for CD45, pan-cytokeratin (PanCK), desmin, human melanoma black 45 (HMB45), and S100. Ki67 immunostaining showed high cell proliferative activity throughout the tumor, supporting a diagnosis of AS.

Radiologically, a CT scan of the neck with contrast revealed diffuse soft tissue swelling and edema on the right side of the neck, involving the right periorbital region, parotid space, masticator space, buccal space, carotid space, prevertebral space, and superior mediastinum. There was diffuse enlargement and hyperenhancement of the right parotid gland and submandibular gland, along with surrounding fluid, edema, and fat stranding, extending inferiorly to the right side of the neck. Multiple lymph nodes were identified, with the largest noted in the left submandibular region, measuring approximately 11 mm. Mild mucosal thickening of the right sphenoid sinus was also noted (Fig. 3). A targeted non-enhanced MRI of the neck showed right-sided superficial skin thickening with subcutaneous fat stranding.

Due to the extensive infiltration, complete surgical resection was not feasible. The patient was referred to a tertiary care center for a comprehensive evaluation to determine the most appropriate treatment strategy.

Discussion

AS is a rare malignant neoplasm of endothelial cell origin, accounting for less than 1% of all sarcomas. It can develop in any part of the body, with the skin and superficial soft tissue being the most common sites (4). Although the exact cause of cutaneous angiosarcoma is unknown, several factors are associated with its development, including chronic lymphedema, prior radiation therapy, and exposure to exogenous toxins. In our patient's case, none of these established risk factors were present.

The clinical presentation of cutaneous angiosarcoma is highly variable. As reported in the literature, these lesions can present in different forms, ranging from single or multifocal bluish or violaceous nodules, plaques, or flat infiltrative areas. This variability poses a significant challenge, particularly in the early stages, as initial signs can be subtle, appearing as mild erythema or bruise-like lesions that often mimic other skin conditions. In addition, various atypical presentations have been documented, further complicating diagnosis.

Delayed diagnosis of AS is common due to frequent misdiagnosis. A thorough review of the literature highlights numerous atypical features of cutaneous angiosarcoma (Table 1). These cases have been reported to mimic a range of benign and malignant dermatological conditions, including hematoma (5),

keratoacanthoma (6), scarring alopecia (7) and others. Our patient's case provides yet another example of diagnostic difficulty because she exhibited an unusual photodistributed pattern.

Accurate diagnosis of AS is often challenging. A definitive diagnosis typically requires a biopsy, histopathological confirmation, and immunohistochemical analysis. MRI is also valuable in assessing the extent of the primary lesion before surgery. Despite optimal care, the prognosis remains poor. Whereas primary soft-tissue sarcomas have a 5-year survival rate of 50% to 60%, angiosarcomas show an overall 5-year survival rate of approximately 35%, even in cases with localized disease (1). Clinically, our patient presented several poor prognostic indicators, including advanced age, large tumor size, and necrosis, as revealed by histopathology.

There is no specific treatment guideline for cutaneous AS. Treatment depends on various factors, such as the extent, site, and size of the disease, as well as the patient's overall condition. Therapeutic options include surgery, radiation therapy, chemotherapy, targeted drug therapy, and immunotherapy. A multidisciplinary approach is recommended to optimize outcomes.

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Table 1. Published atypical clinical presentations of cutaneous angiosarcoma.

Ref. no.	Author	Masquerading condition
5	Ambujam et al. (2013)	Hematoma
6	Kong et al. (2013)	Keratoacanthoma
7	Knight et al. (1980)	Scarring alopecia
8	Kwak et al. (2023)	Bilateral facial cellulitis
9	Chow et al. (2021)	Periorbital oedema
10	Kast et al. (2012)	Herpes zoster
11	Aguila et al. (2003)	Rhinophyma
12	Moon et al. (2017)	Erysipelas
13	Dhanasekar et al. (2012)	Squamous cell carcinoma
14	Gonzalez et al. (2016)	Atypical fibroxanthoma
15	Bray et al. (1995)	Xanthelasma and ptosis

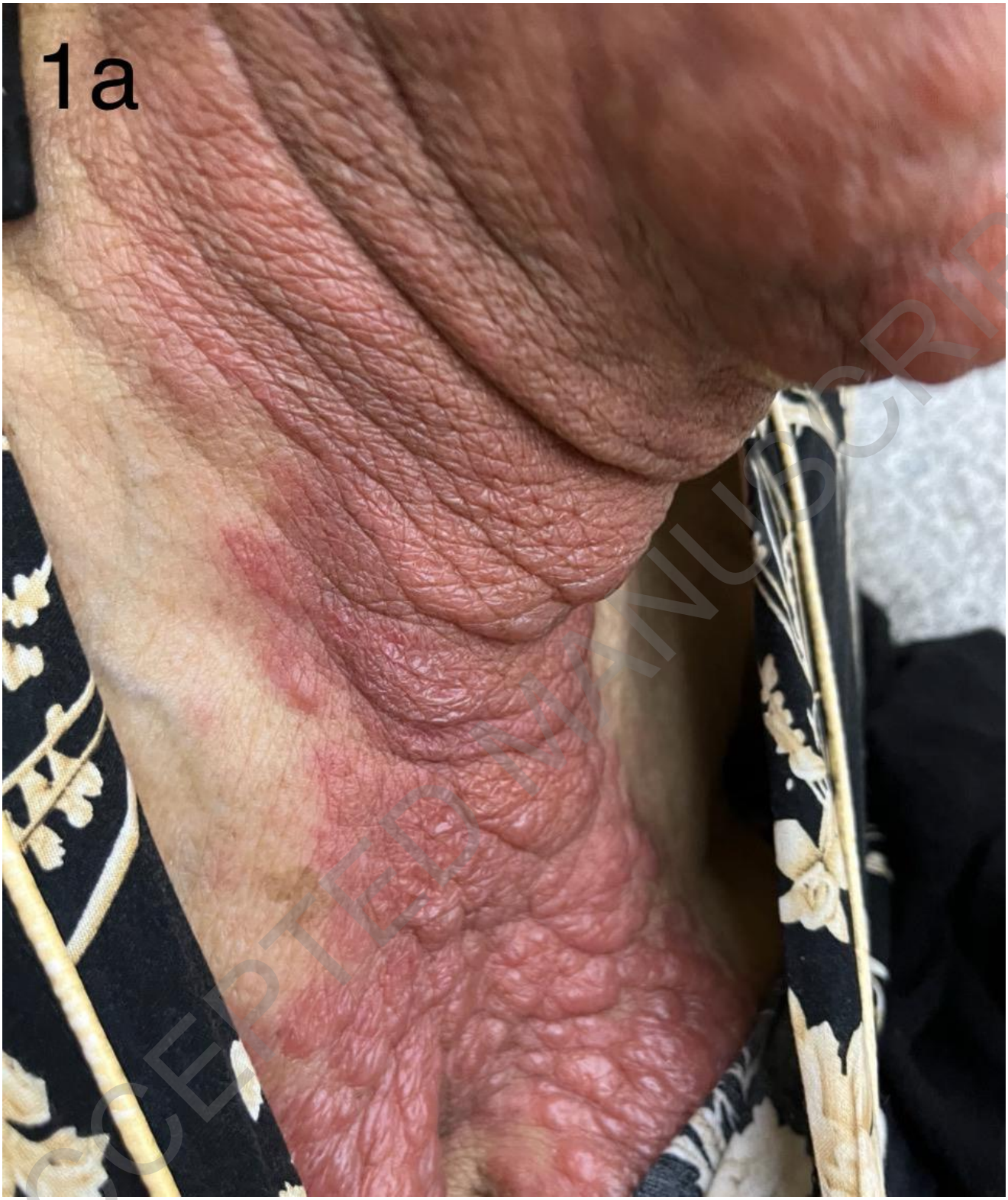

Figure 1a. Well-defined erythematous subcutaneous nodules coalescing into an indurated plaque on a background of erythematous, non-pitting edema extending from the jawline to the chest.

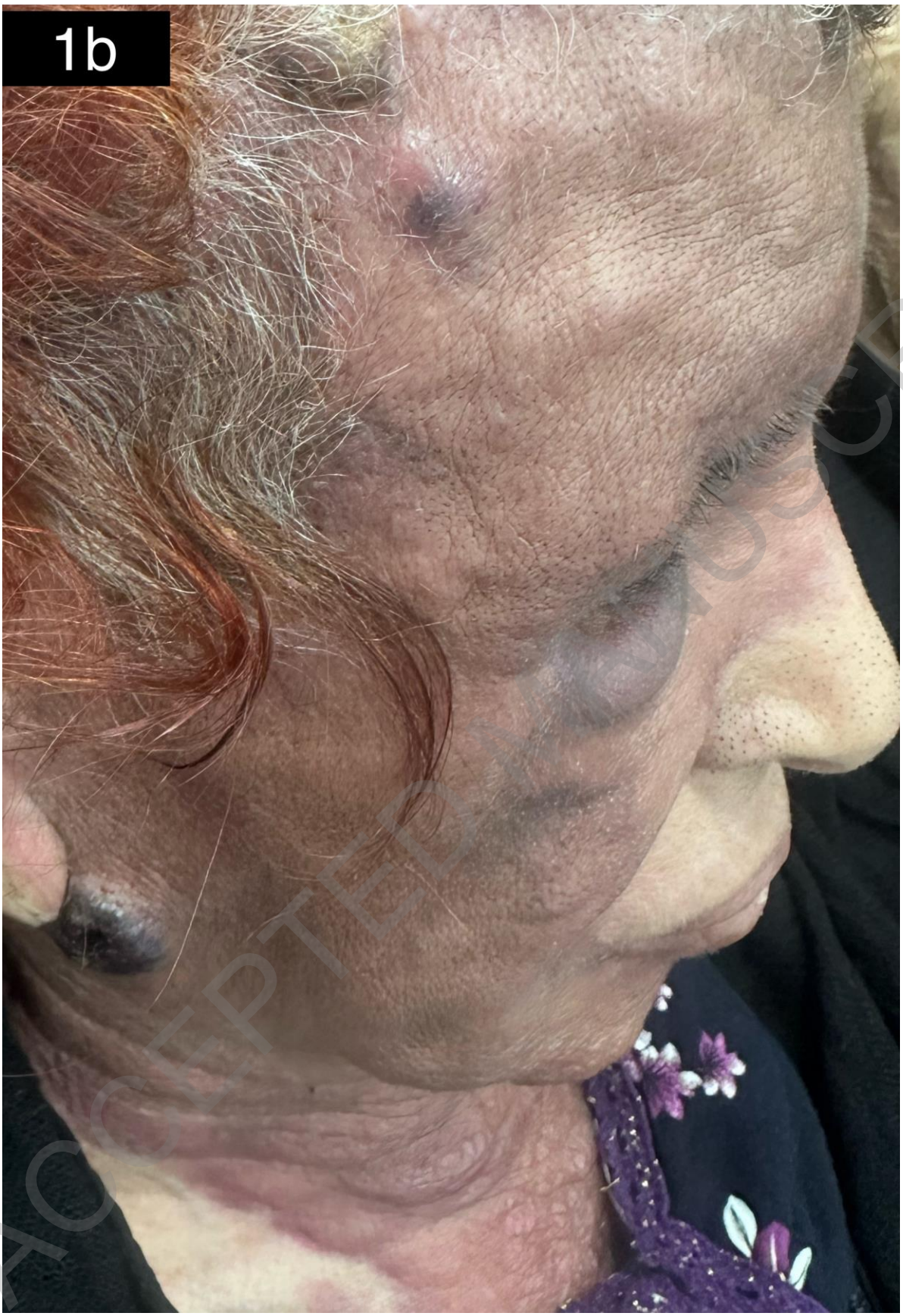

Figure 1b. A few violaceous and black nodules are visible just below her right earlobe and on the scalp, along with edema of the lower right eyelid. Sparing of the nasal tip and perioral area can be noted.

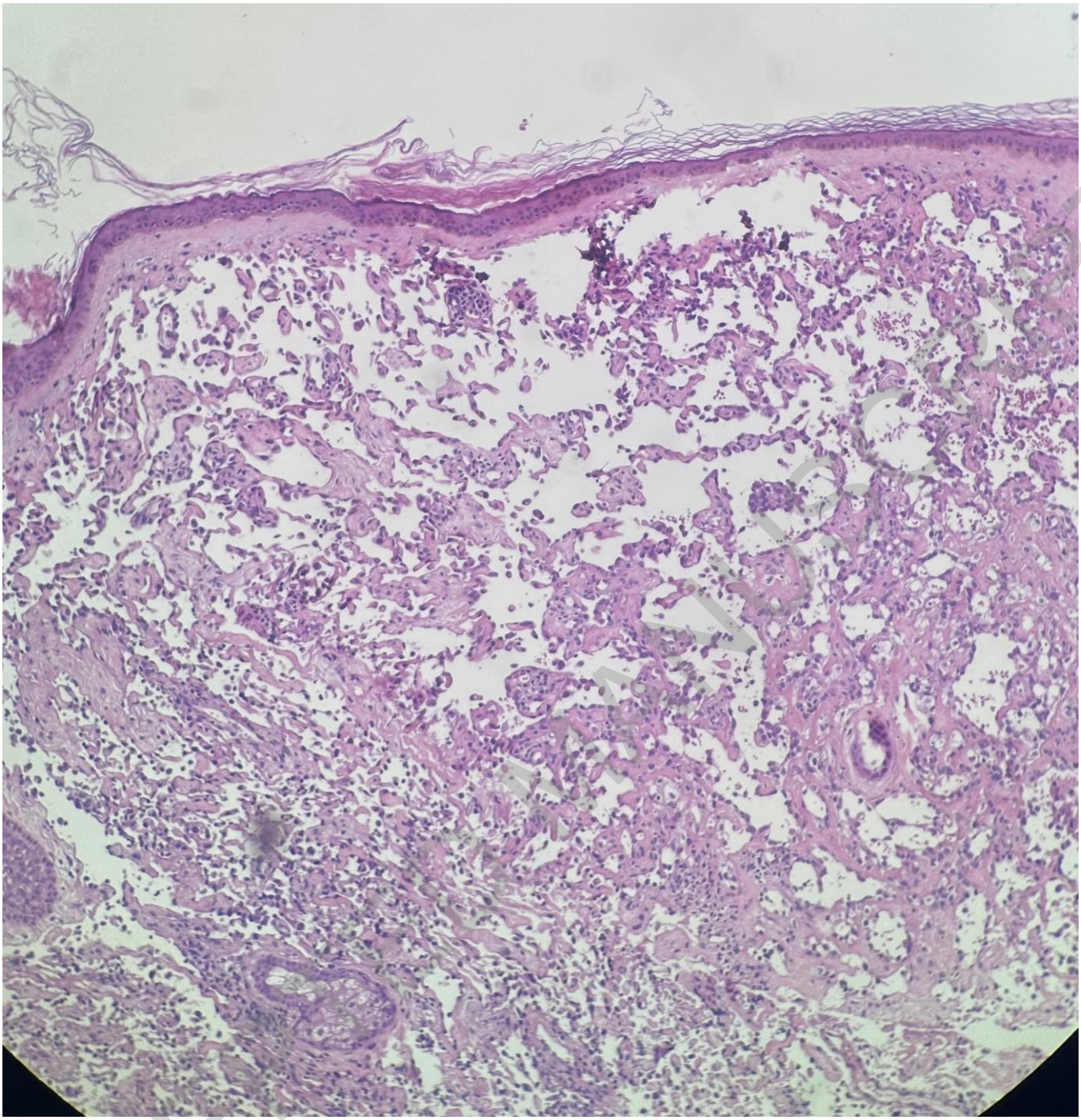

Figure 2. Anastomosing blood vessels with dense hypercellular proliferation of spindle-shaped and epithelioid malignant cells with enlarged pleomorphic and hyperchromatic nuclei, some containing prominent nucleoli. The tumor is vascularized with irregularly shaped blood vessels and hemorrhagic areas. Foci of tumor necrosis are present. Atypical mitotic figures are observed. Immunohistochemical staining results show that target cells are positive for cluster of differentiation (CD) 31 and CD34. Ki67 immunostaining reveals high cell proliferative activity throughout the tumor.

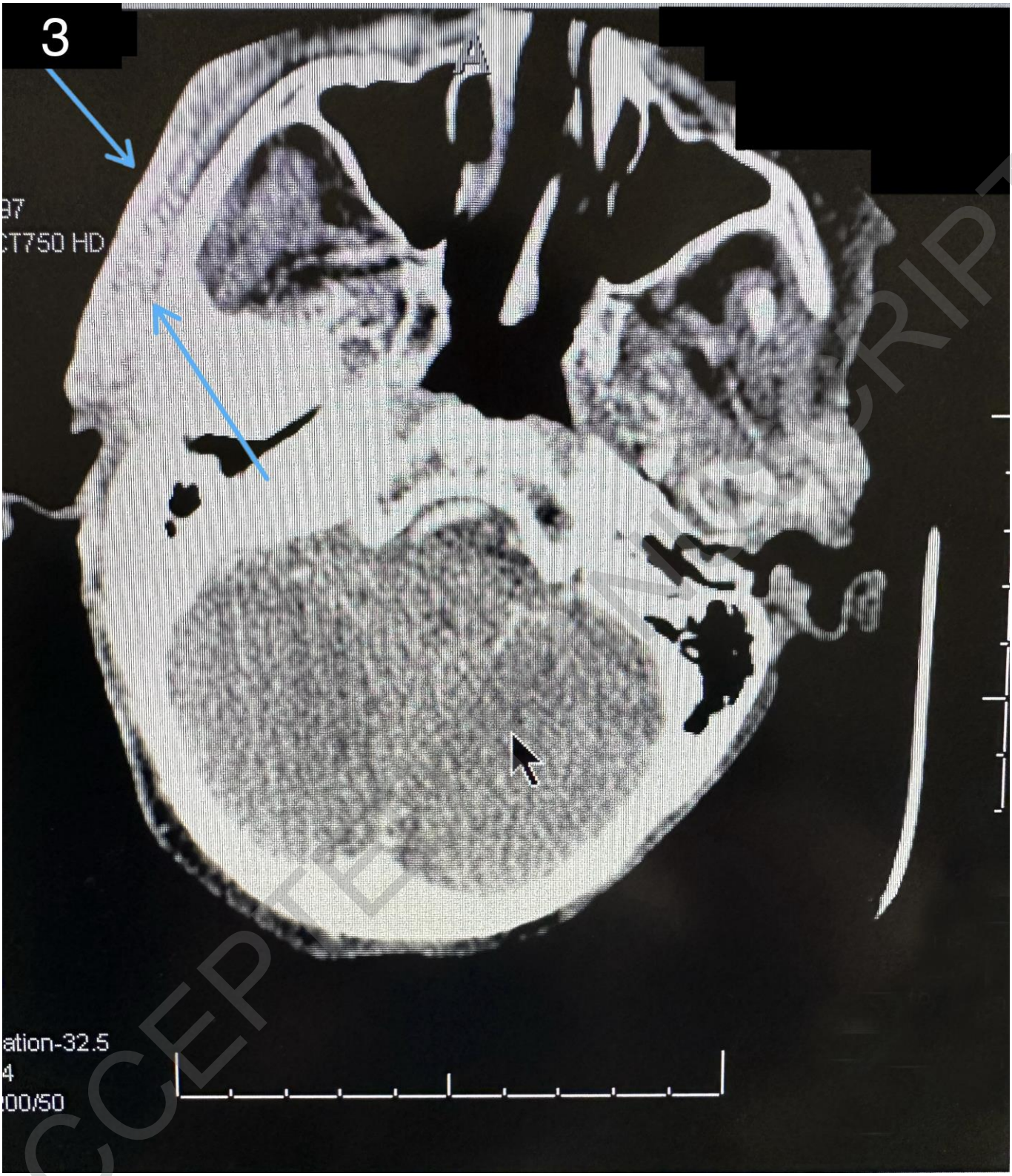

Figure 3. Computed tomography scan showing diffuse soft tissue swelling/edema on the right side of the neck, involving the right periorbital region, right parotid space, masticator space, and buccal space.