Case report

Cutaneous angiosarcoma: A case report

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ABSTRACT

Introduction: Angiosarcoma is a very rare malignancy, which varies based on the location and organ affected. A clinicopathological form of cutaneous angiosarcoma (CAS) involves the head: scalp, face, and neck. We report a 59-year-old female patient with CAS on the temporoparietal region of the scalp.

Case presentation: The patient presented with lesions in the head area, which appeared suddenly in the last 6 months, (before her surgery). Excision was performed under general anesthesia for bleeding as indicated on the right temporal region. The excision procedure was carried out three times at various stages, and then closed with skin grafts. Pathological examination of the three excised tissues showed progression, leading to CAS. Based on the clinical picture and anatomical assessment, a consultation with the surgical oncology department was necessary for further treatment.

Discussion: Angiosarcoma has a high rate of progression. The onset of lesions, which are difficult to detect, does not often lead to progression. Other macroscopic features appear as bluish macules, which can be perceived as bruising. Wide surgical resection is the optimal treatment and is usually combined with radiotherapy and/or chemotherapy. This patient underwent gradual excision, based on bleeding in the right temporal lesion, along with progressive pain.

Conclusion: Surgical is the preferred management for our patients. Determination of multimodality therapy as treatment for CAS requires assessment of all factors related to age and patient condition. Follow-up evaluation is carried out after palliative therapy – to observe the general condition of the patient, tumor progression, tendency for metastases, and excision of any remaining lesions.

1. Introduction

Angiosarcoma is a very rare malignancy, about 2% of other skin malignancies: given their similar nature compared to other tumors, angiosarcoma can be seen anywhere on the body. Variations are distinguished by location and the organ attacked. A clinicopathological form involves the head, particularly the scalp, face, and neck. It begins on the skin but can extend to the subcutis [1]. The ratio between men and women is 2:1 [2]. In elderly people, sun exposure can be a factor if it occurs on the scalp 4,5. This malignancy is similar to other types of tumors, in terms of lesion appearance: they may appear as bluish bruising or as a reddish nodular rash [1].

Angiosarcoma tumors are sheets of endothelial cells, with several irregular anastomotic channels. The canals may be filled with blood, or empty and lined with one or more layers of atypical endothelial cells. Another examination must distinguish angiosarcoma from other tumors, especially if benign [1,3]. Its morphological features fall in a spectrum, from well-differentiated angiosarcomas of complex anastomotic
vascular spaces, lined by one or several layers of atypical endothelial cells, to poorly-differentiated atypical endothelial cells. Tumor cells are spindle-shaped or epithelioid [4]. This case was reported using the Surgical Case Report Guidelines (SCARE Guidelines) 2020 [5].

2. Case presentation

A 59-year-old female presented with ulcerative lesions on the head area, which appeared suddenly in the 6 months before surgery (Fig. 1). Initially appearing on the left side of the head, they slowly expanded to the back and right side. The right lesion seemed to be accompanied by bleeding.

We performed a head CT scan, the results were mass impressions on the cutan-subcutan-temporal fascia on the right temporal-parietal-frontal, left parietal, and vertex (Fig. 2).

Excision was performed under general anesthesia for bleeding in the right temporal region. The excision defect was closed with the split-thickness skin graft, with skin from the patient’s leg. Tissue was removed at 8 cm × 6 cm × 1.5 cm and assessed in the pathology laboratory. On histopathology examination, tissues coated with a hyperkeratotic squamous epithelium form a horn-cyst structure, papillomatosis, or erosive epithelium. Stroma look like lymphocytic inflammatory cells with blackish-brown pigment of the hair follicles, with papillomatous seborrheic keratosis and irritation (Fig. 3). Our patient was stable in two weeks, as the wound healed without bleeding.

The second excision was performed after the first postoperative control excision, after following the patient’s condition. In this procedure, most tissue was removed in the parieto-occipital area, with a size of 13 cm × 10 cm × 7 cm and a tumor mass of 7 cm × 5.5 cm × 1 cm. The histopathology evaluation obtained pieces of the tissue, lined with inlaid flat epithelial cells, and subepithelium of connective tissue stroma; this shows pleomorphic epithelial cell groups with coarse hyperchromatic nuclei, from non-keratinizing squamous cell carcinoma. Subsequent controls were done every 2 weeks to observe postop wound healing, success of the skin graft attached to the excision, and the tendency for new lesions. The graft in the parieto-occipital area was well-fused, so the wound was well-maintained. However, in each control, the patient complained of pain in the right temporal lesion, along with wounds and bleeding found.

The third excision was carried out the following month, as the lesion was removed in the right temporal area, measuring 7 cm × 6 cm × 2 cm, with the left measuring 10 cm × 6 cm × 3 cm (Fig. 4). The histopathology results confirmed the diagnosis of cutaneous angiosarcoma (CAS) (Fig. 5).

Postop control was performed 2 weeks after the third excision, when the patient was found to have enlarged lymph nodes and a bluish bruise-like lesion in the right temporal region near her right eye and cheek (Fig. 6). Based on the clinical picture and pathology examination, a consultation with the surgical oncology department was arranged for further therapy.
3. Discussion

Primary CAS, unrelated to radiation or lymphedema, is the most common form of angiosarcoma, and is relatively the same in men and women. Angiosarcoma is a malignancy with a high rate of progression. The onset of lesions, which are difficult to detect, does not necessarily lead to progression [6]. Scalp, head, or neck are common sites of diffuse ecchymosis, involving large surfaces of the scalp, temporal area, or forehead. A previous study found that the most aggressive presentation was angiosarcoma of the face [7]. Lesions are usually well-demarcated and often show traversed areas with a subcutaneous hematoma [8]. The lesions spread from the scalp to the inferior temporal region, and eventually to the skin of the forehead, periorbital region, cheeks, chin, and neck. Other macroscopic features are shown as bluish macules, often seen as bruising. Distinguishing signs are a peripheral erythematous ring, satellite nodules, the presence of intratumoral hemorrhage, and a tendency for the lesion to bleed spontaneously (or with minimal trauma). The tumor progressively enlarges asymmetrically, often becoming multicentric, developing into indurated bluish nodules and plaques [9]. In this case, the patient presented with ulcerative lesions in the head area, appearing suddenly in the six months before surgery. Initially appearing on the left side of the head, they slowly expanded to the back and right side. Lesions that appear bluish-red or the color as the scalp multiply and spread to the temporoparietal area, with dense consistency and overgrown hairs on top of the lesion. We did not find the palpable lymph node enlargement at the bilateral neck in the initial and secondary examination until the third post-op examination. A soft tissue mass was seen in the cutaneous-subcutan-temporal fascia area on other investigations, such as a CT scan. Nevertheless, examining soft tissue tumors has many causes, so it is not specific to a particular type of tumor. Therefore, the pathological examination is carried out in establishing the diagnosis [10].

Initially, the patient underwent excision of the tumor in the temporal area: the lesion was suspected to be a turban tumor, considering its macroscopic appearance: multi-clusters that resemble a turban-like shape over the scalp. The appearance of lesions that are nodular, skin-colored, and varying in size also strengthens the initial suspicion [11]. Due to their variable appearance, lesions were seen as benign. There was difficulty assessing them, due to the patient’s predisposition for benign tumors: the pathology exam before the first excision showed papillomatous seborrheic keratosis. Similar lesions between tumors, squamous
cell or basal cell carcinoma, complicated the diagnosis. However, the clinical examination showing progressive enlargement, increased tenderness, discoloration of the old ulcerative lesion or bleeding, and the appearance of enlarged lymph nodes made us suspect malignancy. Therefore, the diagnosis has not been made until the results of the third pathological examination are available.

Morphologically, CAS is similar to other tumors: that is, infiltrative dermal growths of irregular vascular space that anastomose with characteristic dissection between collagen bundles [12]. CAS lesions are divided into two grades: low-grade lesions of irregular vascular channels, lined with atypical endothelial cells in one or several layers, whereas high-grade lesions are sheets of undifferentiated pleomorphic cells, and difficult to distinguish from carcinoma. The definitive diagnosis can be done by biopsy and immunohistochemistry, using antibodies for the endothelial markers CD31 and Factor VIII-associated antigens [1]. The third excision showed epidermis and dermis, with proliferative blood vessels, some anastomosing, and lined with atypical, hyperchromatic endothelial cells; there were also solid areas of epithelioid cells and those with spindle nuclei, which are features of low-grade CAS. There are many limitations regarding supporting examination where we cannot perform further pathological examinations such as immunohistochemistry due to the covid-19 pandemic.

Treatment of CAS, especially lesions on the scalp, is a challenge for researchers. In recent years, treatment of patients with angiosarcoma has undergone many changes, including focused surgery followed by multimodal therapy with radiation and chemotherapy [6]. Wide surgical resection is the treatment of choice, and as stated, usually combined with radiotherapy and/or chemotherapy with taxanes, ifosfamides, or anthracyclines [13]. However, surgical treatment is often difficult, as tumors tend to have indistinct borders with skipped lesions. Surgical procedures, such as reconstruction combined with free muscle flaps and skin grafts may be performed, considering scalp contour and skin tone matching [14]. A previous study found complications, in which many patients experienced local recurrence outside the resection limits. Other problems were also found while determining the accurate extent of the lesion as well as identifying incorrect boundaries [14,15]. Surgery may be performed for problems of local control (bleeding and pus), or for those who have only one site of disease via clinical and imaging studies for systemic therapy [14]. Other studies show that radical surgery with adjuvant radiation therapy is optimal [15]. This patient underwent gradual excision, based on bleeding in the right temporal lesion, and progressive pain. The excision was accompanied by a skin graft, taken from the skin on the leg. The patient had enlarged lymph nodes, and a bluish bruise-like lesion in the right temporal region near her right eye and cheek. At the latest follow-up after the third excision, a new bluish lesion radiated from the right temporal area to the right eye and cheek, with enlarged lymph nodes in the neck. Based on these findings, the patient consulted with the surgical oncology department for chemotherapy or radiation. These measures are based on patient outcomes, considering age and disease progression. Further excision will be reconsidered after the patient completes chemotherapy. As stated, the 5-year survival rate of CAS in the head and neck region varies widely in terms of severity, but tends to be low [16].

4. Conclusion

Surgical management is the best treatment for our patients. Multimodal therapy for CAS requires consideration of age and patient condition. Follow-up evaluation is done after palliative therapy for tumor progression, with metastases and excision of any remaining lesions.

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Ethical approval

The study is exempt from ethical approval in our institution.

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.

Registration of research studies

Not applicable – single case report.

Guarantor

Maximillian Christian Oley and Mendy Hatibie Oley.

Credit authorship contribution statement

Mendy Hatibie Oley: study concept and surgical therapy for this patient. Maximillian Christian Oley, Regina Elizabeth Meriam Kepel, and Melany Feronika Durry: Data collection and Writing-Original draft preparation. Mendy Hatibie Oley: senior author and the manuscript reviewer. Muhammad Faruk: Editing and Writing. All authors read and approved the final manuscript.

Declaration of competing interest

Nothing to declare.

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