

Angiosarcoma of the scalp : A zosteriform presentation

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Abstract

Cutaneous angiosarcoma is a rare and aggressive tumor with a poor outcome, and early diagnosis is paramount, but it's often delayed because of the long delay in the appearance of lesions and the variable clinical presentation of this neoplasia. This article reports the case of an 84-year-old patient who presented with a persistent ulcerated plaque on the scalp and face evolving for one year that was initially treated as herpes zoster ophthalmicus. Incisional biopsy revealed a morphological aspect of high-grade angiosarcoma with CD31+/CD34-. An extension workup, consisting of thoraco-abdominal and pelvic CT scans, showed hepatic, adrenal, and bone metastasis. Given his advanced age, general condition, and visceral metastasis at the time of diagnosis, the decision of the multidisciplinary consultation meeting was to provide palliative care. He passed away 8 months after the diagnosis of angiosarcoma was made, 18 months after the occurrence of the first symptoms. Cutaneous angiosarcoma (CA) is a rare aggressive malignant tumor that occurs in elderly white men. Due to the variability of its presentation and its insidious evolution, the appearance of lesions may not concern patients or practitioners for some time. Therefore, correct diagnosis may be delayed, often dramatically limiting the available treatment options and impacting the outcome of the patients

Keywords: Ulcerated placard, Cutaneous angiosarcoma, Neoplasm, Scalp, Herpes zoster ophthalmicus.

Introduction

Cutaneous Angiosarcoma is an aggressive malignant tumor of the vascular endothelium that accounts for approximately 1,6% of cutaneous soft tissue sarcomas [1]. It mostly occurs in elderly white men, and can appear anywhere on the skin with a predilection for head and neck in 60% of cases and can be unifocal or multifocal at presentation [2]. Timely diagnosis is often delayed, because of the very variable clinical presentation of this tumor and its slow onset and thus a dark prognosis with a 5-year overall survival less than 26% [1]. We report the case of a patient with Angiosarcoma of the scalp mimicking an herpes zoster ophthalmicus.

Observation

84 year man, farmer with history of diabetic and arterial hypertension for 20 years, was admitted to our institution for indurated and ulcerated purple plaques on the scalp evolving one year prior to consultation and treated as an herpes

zoster phthalmicus on several occasions. General clinical examination revealed a conscious patient, with a ECOG Scale of Performance Status : 2, dermatological examination found an ulcerated and indurated purplish scalp plaques, which seemed to occupy the territory of the left ophthalmic nerve with a circumferential extension to the right hemiscalp [figure 1]. This plaques extended to the forehead and left cheek, with multiple ulcerated nodules, we also found edema of both eyelids more pronounced on the left eye, obscuring the vision [figure 2]. Further examination revealed multiple tender lymph nodes occupying the left cervical region. An incisional biopsy was performed and revealed a morphological aspect of high grade Angiosarcoma with CD 31 + / CD 34 -. Thora-coabdominal and pelvic CT scans showed hepatic, adrenal and bone metastasis. The patient was discussed in multidisciplinary concertation meeting and the decision was to put him under palliative care, given his advanced age, general condition and visceral metastasis. He passed away eight month later, about 18 months after the occurrence of the first symptoms of the tumor.

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Figure 1: Ulcerated and indurated purplish scalp plaques.

Discussion

The originality of our observation lies in the atypical aspect of the lesions, simulating an herpes zoster ophthalmicus. Angiosarcoma is a rare and very aggressive tumor that develops in the vascular endothelial cells of the skin and superficial soft tissues, it mostly occurs in elderly people, men are more affected than women, the main risk factors are ionizing radiation and the longstanding solar exposure [3], as was the case of our patient. Reported cases in the literature have described the lesions as infiltrated purplish-red papules, plaques, and nodules that may be misdiagnosed as rosacea, eczema, and hematoma. These lesions may also mimic cutaneous lymphoma, metastatic carcinoma or Kaposi's disease[3]. To our knowledge, this is the first case of Angiosarcoma taking on a metameric arrangement and thus the diagnosis of herpes zoster ophthalmicus complicated by cellulitis and treated with antivirals and antibiotics was given before the correct diagnosis of Angiosarcoma provided.

As in our patient's case, 50% of Cutaneous Angiosarcomas occur in the head and neck. The scalp is the most affected area, which also holds the worst prognosis, The 5-year survival rate for patients with Cutaneous Angiosarcoma of the head and neck ranges from 10% to 54% [2].

Prompt diagnosis and treatment is an important factors in improving survival, but may be delayed due to insidious tumor progression as lesions can be hidden by hair for a long time and the initial benign presentation, as was the case for our patient. Jonathan M. et Al showed that the average delay between the appearance of the lesions and the positive diagnosis was 4 months, in our case the delay was one year [2].

Despite its infiltrative nature, visceral metastases of Angiosarcoma are uncommon. In a study of 52 patients, only 2% had

distant metastases [2]. Their presence at the time of diagnosis is considered as a poor prognostic factor, as was the case of our patient.

Several treatments are available for Angiosarcoma. Traditionally, the combination of large surgical excision of the tumor and preoperative or postoperative radiotherapy provides the basic treatment [4]. However, complete surgical resection can be challenging because of the very infiltrative and multifocal nature of the tumor like was the case of our patient, which often leads to local recurrence and metastatic lesions.

The median survival time of patients with Cutaneous Angiosarcoma of the scalp ranges from 12 to 42 months [5]. the median survival time of our patient since the first appearance of the lesions to the time of diagnosis was 18 months. Age over 70 years, tumor size larger than 5 cm, location in the scalp, multifocality ,ulceration and metastases in the moment of diagnosis are predisposing factors for poor prognosis of Angiosarcoma, our patient had them all.

Conclusion

Cutaneous Angiosarcoma is an aggressive malignant tumor, the variable presentation and insidious onset of the lesions may not trigger concern among patients or practitioners for sometime and thus correct diagnosis can be delayed, often drastically altering outcome and treatment options. Our case serves as a reminder to physicians that an abnormal skin finding in elderly adults should prompt them to suspect Angiosarcoma and perform an early biopsy.

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