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POSTIRRADIATION MULTIFOCAL CUTANEOUS ANGIOSARCOMA OF THE SCALP: REPORT OF A CASE AND LITERATURE REVIEW

SKALPTE RADYASYONA BAĞLI ÇOK ODAKLI KUTANÖZ ANJİYOSARKOM: OLGU SUNUMU VE LİTERATÜRÜRN GÖZDEN GEÇİRİLMESİ

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ABSTRACT

Purpose: Angiosarcoma is a malignant tumor of vascular endothelial cells that arises in the head and neck. The purpose of this case report is to present a very rare difficult to treat, lethal multifocal type of angiosarcoma.

Patient: A 65 year-old man was diagnosed with basal cell carcinoma on the superior occipital region 10 years ago. After 9 years of postirradiation controls, a new lesion occurred and was treated with cryotherapy . Five months after the onset of the second lesion the patient applied to our clinic with a rapidly growing plaque-like lesion on the vertex region that bled occasionally. Physical examination revealed 8x9 cm plaque, composed of a central necrotic and bleeding surface, surrounded by small purple-red satellite nodules. The incisional biopsy revealed the diagnosis of cutaneous angiosarcoma and concomitant basal cell carcinoma was made and the patient was admitted to the clinic for surgical therapy.

Method: Patient was evaluated with chest x-rays, computerized tomography and magnetic resonance imaging techniques. The incisional and excisional biopsies were stained with significant immunohistochemical stains such as, cytokeratin, CD34 and Vimentin.

Result: Multifocal angiosarcoma of the vertex and occipital region with concomitant basal cell carcinoma after radiation therapy is a lethal, aggressive tumor of the head and neck region.

Conclusion: A combination of good clinical prognostic factors (young age at presentation, fewer lesions on presentation, small tumor size, and perhaps the ability of clear margins) as well as definitive treatment with surgery and radiation offers the best hope of cure for patients with cutaneous angiosarcomas.

Keywords: postirradiation, multifocal, cutaneous angiosarcoma

ÖZET

Amaç: Anjiyosarkom, baş boyun bölgesinde vasküler endotelyal hücrelerden kaynaklanan malign bir tümördür. Bu olgu sunumunda, tedavisi oldukça zor olan multifokal anjiyosarkom vakası tartışılmaktadır.

Olgu: Altmış beş yaşında erkek hastaya verteks ve oksipital bölgede bazal hücreli karsinom tanısıyla 10 yıl önce eksizyon ve radyoterapi uygulanmış. Radyoterapiden 9 yıl sonra yeni bir tümöral lezyon oluşmuş ve kriyoterapi uygulanmış. İkinci lezyonun çıkmasından 5 ay sonra vertekse yakın bölgede hızla büyüyen kanamalı lezyonla hasta kliniğimize başvurdu. Fizik muayenede 6x8 cm çapında santrali nekrotik ve kanamalı alanlar içeren etrafında satellit nodüller bulunan bu kitlede bazal hücreli karsinom ve anjiyosarkomun eş zamanlı olarak bulunduğu gözlendi.

Sonuç: Multifokal anjiyosarkom ve radyoterapi sonrası oluşan bazal hücreli karsinom birlikte oldukça agresif bir seyir göstermektedir. Genç yaş ve lezyonların çapının küçük olması cerrahi tedavi ve radyoterapiyle iyi sonuçlar elde edilmesinde belirleyici faktörlerdendir.

GİRİŞ

Angiosarcoma is a malignant tumor of vascular endothelial cells that can occur in any region of the body which usually affects the face and scalp region, most often in the elderly patients. Overall, Meis-Kindblom showed that sarcomas occur uncommonly in the head and neck, constituting less than 1% of all head and neck malignancies.[1] According to Aust et al, fewer than 5% of soft tissue sarcomas occur in the head and neck, with only approximately 10% classified as angiosarcomas.[2] Angiosarcomas of the face and scalp are incidental, and their clinical presentation varies widely. In their early stages, they frequently appear clinically innocent and even may show benign capillary hemangioma-like structures histologically. This pattern, however, is deceiving, because angiosarcomas usually have an aggressive course. Tumor cells are located mainly in the dermis and may extend into the subcutaneous tissue. Angiosarcoma has a tendency for metastasis via lymphatic

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or hematogenous routes, and late local recurrence and metastasis after years of apparent remission and successful local control are well documented. According to Requena et al the overall prognosis for patients with angiosarcoma of the head and neck remains unsatisfying, with a reported 5-year survival rate of approximately 10%.[3]

Given the rarity of the tumor, relatively little is known concerning the features, natural history, or optimal treatment of face and scalp angiosarcomas.

Reports concerning the treatment of head and neck angiosarcomas are infrequent in the medical literature. There is no evidence, however, to suggest that sarcomas of the general head and neck region behave in the same manner as scalp angiosarcomas. Furthermore, given the small numbers, previous studies as stated by Verleysen et al, have been unable to analyze how various clinical and therapeutic modalities may affect the time to recurrence and overall survival in patients with scalp angiosarcoma.[4]

This case report describes a rarely seen multifocal localization of angiosarcoma of the scalp after radiation therapy and recurrent basal cell carcinoma. This case not only demonstrates the possible relationship between radiation therapy and cryotherapy later formed angiosarcomas, but it also stresses the possibility of multifocal sarcoma erupting eventually.

CASE REPORT

A 65 year-old man was diagnosed with basal cell carcinoma on the vertex region 10 years ago. Past history of the patient was significant for the presence of pulmonary hypertension. At that time the patient had rejected surgical resection of the lesion and was subsequently treated with radiation therapy. After nine years of postirradiation controls a new lesion occurred and was treated with cryotherapy in another hospital. Five months after the onset of the second lesion the patient applied to our clinic with a rapidly growing plaque-like lesion on the vertex region that bled occasionally. Physical examination revealed 8x9 cm plaque, composed of a central necrotic and bleeding surface, surrounded by small purple-red satellite nodules. An incisional biopsy from the erythematous area showed an ill-defined infiltrative intradermal mass with a pattern of hypercellular sheets of large cells alternating with areas of dilated , irregular, blood-filled channels, dissecting the collagen bundles (Figure1, 2). The diagnosis of cutaneous angiosarcoma and concomitant basal cell carcinoma was made and the patient was admitted to the clinic for surgical therapy (Figure 3).

Chest X-rays and computerized tomography of the chest and head and neck showed an 8 x 9 cm infiltrative tumor of the right frontotemporal region with invasion of subcutaneous tissue, temporalis muscle and the deep fascia planes. The lesion reached the zygomatic arch but showed no signs of further expansion.

Thorax and abdominal computerized tomography

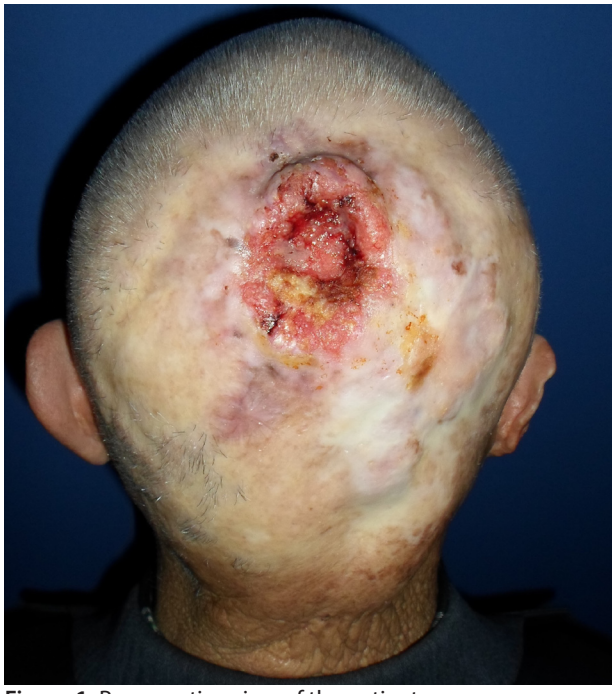

Figure 1. Preoperative view of the patient

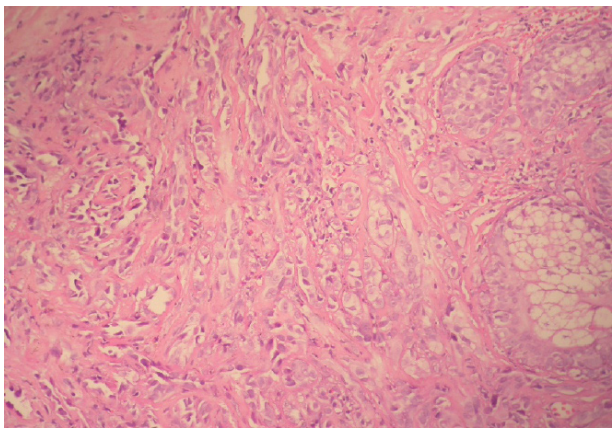

Figure 2. Incisional biopsy of high-grade angiosarcoma composed of ill-defined intradermal mass, well-formed vascular channels with enlarged endothelial cells infiltrating sebaseous glands, sweat glands and dermal collagen fibers. (H&E, x40)

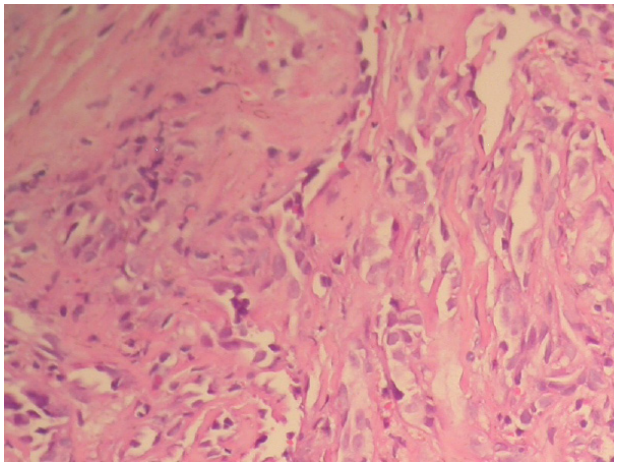

Figure 3. On higher magnification, the immunohistochemical stain is positive for Vimentin, which is a connective tissue marker that helps stain endothelial cells and collagen tissue, and is negative for adipose tissue. (Vimentin, x200)

didn't reveal distant metastasis. During the operation the tumor seemed to reach the periosteum of temporal bone, but no signs of dural invasion were noticed. Therefore, the reconstruction was done with split-thickness skin grafting and subsequent modified radical neck dissection with superficial parotidectomy. Postoperative pathology revealed that the tumor was continuing in inferior margins although the frozen section had revealed no malignancy. Neck and parotid dissection materials were reactive with lymphoid hyperplasia. Patient's general health condition was not suitable for a wider reexcision at the time so he was referred to radiation oncology for wide-field radiation therapy. Two months after the first operation the lesion on the temporal region rapidly grew towards the parietal region posteriorly, and to the orbit anteriorly causing significant angioedema over the orbita as concluded by Tay.[5] However a biopsy from the face was not done to confirm this . Meanwhile a second focus was noted to be forming on the left occipital region of the scalp. Its' margins were separated clearly from the original lesion. The tumor at this point reached approximately 18x12 cm in the temporoparietal region and 5x6 cm in the left occipital region .

A computerized tomography and magnetic resonance imaging was performed. Computerized tomography revealed invasion of orbital preseptal region and the MRI showed the extension of the tumor to the auricle, premaxillary region, preorbital region, requiring exenteration and total amputation of the auricle. No dural invasion or intracranial metastasis was reported. A second operation was planned where a wide-excision of the tumor was performed. Local flaps seemed to be a better choice of treatment so the large defect was reconstructed with cervicohumeral fasciocutaneous flap. The second focus on the occipital region was excised and grafted with a split-thickness skin graft. During the early postoperative period the patient's pulmonary hypertension was aggravated and unfortunately the patient died of pulmonary embolism.

DISCUSSION

Angiosarcomas are highly aggressive mesenchymal tumors with elements of vascular differentiation. As Fedok et al stated scalp angiosarcomas are recognized as a subgroup, usually occurring in the elderly white men, with an estimated male-to-female ratio of 3:1 and an average presentation age of 63 years(5). It may occur in any region of the body, but most frequently on the head and neck. Angiosarcomas are usually found on the scalp in the elderly population and they behave rather aggressively. Livingston and Klemperer, were the first to report the clinical features and histopathological characteristics of angiosarcoma in a case with presentation of a man with scalp lesion.[6] According to Ward et al location of angiosarcoma was the only statistically significant variable predicting survival.[7] All patients with angiosarcoma of the scalp ultimately died of disease. Few cases have been reported of postirradiation angiosarcoma as concluded by Caldwell et al(2). In most of the cases the primary lesion was a benign disease and the latency period ranged between 20-50 years. According to Livingston and Klemperer , the global incidence of irradiation-associated sarcoma is estimated as 0.03 and 0.08%.[8] Our patient had basal cell carcinoma as a primary lesion and 9 years of latency period.

This tumor expands with gradual infiltration, grows rapidly and has the highest rate of lymph node metastases of all soft tissue sarcomas of the head and neck. It may metastasize to cervical nodes and lungs. Extensive local spread is the rule as stated by Rosai et al, and the prognosis is dismal with a few patients surviving longer than 3 years.[9] Five year survival rates in the literature range from 12% to 41%. The scalp is a site where adequate surgical resection is rarely possible. The diagnosis is often delayed as stated by Pawlik et al, because this tumor usully presents as " a vicious tumor masquerading as a bruise with indistinct border".[10,11] Radiation therapy is frequently necessary as an adjuvant to surgery. This is of course a dilemma, radiation therapy being an etiological factor and an adjuvant therapy to surgery as Mark et al stated.[12]

Three known conditions are associated with development of angiosarcoma. They are long-standing lymphedema, prior radiation and a history of trauma to the lesion. Our patient had a history of trauma to the lesion, being cryotherapy, in addition to prior radiotherapy.

Higher grade lesions as our case are usually multiple and ulcerated with extensive spread by local infiltration. Multiple lesions are rare in literature and to the best f our knowledge only Rosai et al reported a rate of 40% of patients with multiple angiosarcomas. Satellite lesions were also seen by Requena and Sangueza in some undifferentiated cases.[13] Our patient presented with a second focus of angiosarcoma and from the history of the patient it seemed to have erupted like a separate lesion from the original tumor in the temporoparietal region. Also the margins of the two tumors were clearly defined and about 4 cm apart from each other predicting a separate lesion.

In this case report we emphasize the importance of multifocal scalp angiosarcomas, and the decreased disease-free survival in these patients. Disease stage and tumor size are also directly correlated with survival rate.

Achieving a negative surgical margin is exceptionally difficult. Pawlik et al, in scalp angiosarcomas and frozen section is not a definite marker for negative tumor margins as in our case.[14]

Potential treatment options for the patients include, surgery, radiation, and chemotherapy, possibly gene therapy more recently. Hodkinson et al, from the Mayo clinic reported that the only two survivors in their series of angiosarcomas received radiation therapy in addition to extensive surgery.[15] Patients who received radiation therapy had a median survival almost 4 times longer than patients who did not receive radiation the-

rapy (36.1 months versus 9.2 months respectively) ,according to a study by Pawlik et al. Compared with radiotherapy, chemotherapy is used much more sparingly. Paclitaxel is one of the agents that have activity against angiosarcoma. In one study by Fata et al, a response rate of 89% was seen in patients with angiosarcoma of the scalp or face, even in these patients who were treated previously with chemotherapy or radiation therapy. According to Pawlik et al and Gil-Benso et al's studies, few patients responded to gene therapy, which involved direct intralesional injection of the cDNA for Interferon α-2b.[16]

CONCLUSION

It appears that a combination of good clinical prognostic factors (young age at presentation, fewer lesions on presentation, small tumor size, and perhaps the ability of clear margins) as well as definitive treatment with surgery and radiation offers the best hope of cure for patients with cutaneous angiosarcomas.

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