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Case Report
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Primary Angiosarcoma of the Scalp - A Rare Entity

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Abstract

Angiosarcoma is a rare soft tissue tumor of vascular endothelial origin with the predilection for head and neck region. It frequently affects elderly people. Its presentation is variable, causing difficulties in diagnosing it timely. Most of the patients are treated by wide excision with reconstruction of defect. The role of radiotherapy and chemotherapy are well documented for advanced disease. Post-operative radiotherapy also decreases likelihood of local recurrence. We are reporting a case of 70 yr female with bleeding scalp lesion. After thorough work up, wide excision with cover of defect with local transposition flap was done. Post-operative radiotherapy was given.

Keywords: Angiosarcoma; Scalp; Radiotherapy

Introduction

Angiosarcoma is a rare soft tissue sarcoma of vascular endothelial origin accounting for less than 1% of head and neck malignancies [1]. It frequently involves the scalp and face. Wilson-Jones initially described a separate subgroup of angiosarcoma involving scalp of elderly patients and confined to skin and soft tissues only [2]. Various etiologic factors have been suggested, however many patients are not found to be associated with any pre-existing conditions [3]. The presentation of patients is variable [4]. As it possesses a relatively benign nature and rarity of incident it is diagnosed late and in advanced stages. We are reporting a case of 70 yr female presented with angiosarcoma of the scalp.

Case Presentation

A 70 yr female patient presented to the outpatient department with the scalp swelling with occasional episodes of bleeding. The patient noticed this swelling 6 months back during combing of her hair when some bleeding took place. The patient went to some quack for treatment where the patient received periodic dressing after which it subsided. But later it again reappeared and then increased in size with more frequent episodes of bleeding, even with minor trauma.

On examination, a 6 cm × 5 cm × 1 cm lesion was seen over left parieto-occipital region of the scalp. It was brownish black in color, firm to feel with central part of lesion showing ulceration with irregular margins and bleeding was noticed on palpation (Figure 1). Rest of the scalp was normal. There were no cervical lymph nodes palpable. Contrast enhanced CT head was done. It showed a heterogeneously ill-defined soft tissue lesion over left parietal area with areas of necrosis within it. No evidence of underlying bony erosion was noticed. No other evidence of metastasis was noticed.

Patient underwent wide local excision of the lesion along with underlying pericranium with cover of the raw area by transposition flap (Figure 2). Histopathology of the specimen showed atypical vascular spaces lined by spindle to plump epithelioid shaped atypical endothelial cells with multi-layering and intra cytoplasmic lumina having RBG formation. Immunohistochemistry (IHC) showed diffuse strong positivity for CD31. Ki-67 proliferation index was ~15% to 20%. Overall morphology and IHC findings were consistent with cutaneous angiosarcoma. All margins and base were free from tumour cells.

Sutures were removed on 10th post-operative day and wound healed well (Figure 3). Post-operative radiotherapy was given to local area. Total dose of 70 Gy in 2 Gy fractions was given. The Patient tolerated radiotherapy well. No evidence of local or distant recurrence noticed in 6 month follow up.

Discussion

Angiosarcomas of the skin are aggressive tumors of vascular endothelial origin with high rates of recurrence and metastasis. Various factors have been found to be associated with increased risk of angiosarcomas. Previous exposure to radiation, exposure to various chemicals (polyvinyl chloride, arsenic), chronic lymphedema, increasing age remains some factors among them. Most of

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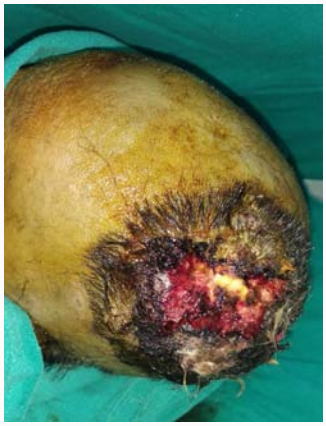

Figure 1: Pre-Operative Photo showing lesion over left parieto-occipital area of scalp.

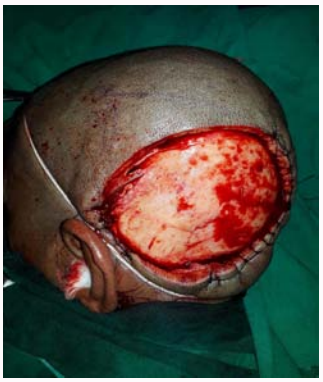

Figure 2: Intra operative photo after excising lesion along with pericranium.

the patients don't have any underlying factor. In our patient also, no underlying predisposing factor was noted. Angiosarcoma of the scalp occurs more frequently in elderly patients with male to female ratio being 2:1 [4]. Our patient, however, was a female.

The diagnosis of early angiosarcoma is difficult as it presents as a bruise like lesion or purplish macule causing confusion with hemangioma [5]. Ulcerated, fungating masses or hemorrhagic lesions represent signs of advanced diseases. The 5-year survival rate for patients with cutaneous angiosarcoma of the head and neck region ranges from 10% to 54% [6-8].

Surgical wide excision of the lesion to achieve tumor free margins is considered to be first line of treatment. If the underlying pericranium remains intact, raw area created after excision can be grafted. In our case as the pericranium was also excised, it was covered with the local flap (transposition flap). The free flap is also an option in cases of extensive lesions involving whole scalp.

Post-operative radiotherapy is indicated in patients to decrease chance of local recurrences and improve 5-year overall survival [9]. Recently researchers at Memorial Sloan-Kettering Cancer Centre, New York had reported paclitaxel to be active against angiosarcoma of the scalp due to its antiangiogenic properties [10].

In our case we have managed the patient with wide local excision and post-operative radiotherapy.

Conclusion

Angiosarcoma being a rare and aggressive tumor needs prompt diagnosis and aggressive treatment. In elderly patients having scalp lesions, suspicion of angiosarcoma should be kept in mind. It will help in managing them in the early stage of disease. Timely treatment increases chances of better outcomes and prognosis. Due to aggressive nature of disease life-long, to follow up is required for detecting local recurrence or distant metastasis.

Figure 3: Post-operative follows up after 2 months.

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