International Journal of Oral and Facial Surgery

# Sarcomas of the scalp: a case series

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## ABSTRACT

Sarcomas of the scalp are a diverse group of rare and aggressive neoplasms of the head and neck originating from bony or soft tissue elements. Clinical manifestations of sarcomas of the scalp vary from small plaque-like lesions to multifocal nodules with involvement of deeper layers, and it is the extent and grade of the tumor that determines the treatment plan. Majority of the cases are managed surgically with three-dimensional wide excision and robust reconstruction. The role of adjuvant radiotherapy and chemotherapy is limited to those cases with poor prognosis. Very few cases of sarcomas involving the scalp have been reported in the literature. We wish to share a series of three rare mesenchymal tumors of the scalp, two of which were locally advanced at the time of presentation. This case series highlights the surgical management, options for reconstruction and outcomes of these tumors.

**Keywords:** Epithelioid sarcoma; Angiosarcoma; Dermatofibrosarcoma; scalp; Mesenchymal tumors; Undifferentiated Pleomorphic sarcoma.

**ISSN:** Awaiting
Case Report

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**Article Info**

Received on: 26-06-2020
Revised on: 03-07-2020
Accepted on: 10-07-2020



## INTRODUCTION

Sarcomas are malignant neoplasms of mesenchymal origin [1]. Sarcomas of the head and neck (H&N) are relatively rare neoplasms and account for less than one percent of all the head and neck malignancies [2]. They arise from bony or soft tissue elements. On average, 80% of the sarcomas are of soft tissue origin and only 20% are of bony origin [3]. Soft tissue sarcomas are more common in the head and neck region and are associated with high recurrence and mortality rates. Soft tissue H&N sarcomas account for more than fifty different histological types [4]. The most common variants are malignant fibrous histiocytoma, angiosarcoma, malignant peripheral nerve sheath tumor and non-classified or non-differentiated sarcoma [5]. Soft tissue sarcomas arising in the H&N have less overall survival rate and poor prognosis than those arising in the other sites. This may be due to the increased occurrence of aggressive histological types like fibrosarcoma & angiosarcoma and/or due to inability to obtain wide surgical margins, resulting in high rates of local relapse (LR) [5]. The site of origin of these sarcomas play an important role in planning the treatment, as it influences the safe margin, aesthetics, post-operative function and quality of life. Due to their rarity, literature available on scalp sarcomas is limited and management is guided by multidisciplinary team inputs. We present a case series of advanced mesenchymal tumours of the scalp with an insight into its management and prognosis.

## CASE REPORTS

### Case Presentation 1

A 16 year old male patient from the Pacific island of Tonga reported to the head and neck out patient department with the complaint of a swelling on his forehead for the past four months. Patient gave a history of injury to the forehead from a wooden plank following which the swelling developed. He reported a progressive growth of the swelling which was associated with occasional pain, ulceration and foul smell. Clinical examination revealed a highly vascular mass of size 15 X 10 cm in the fronto-parietal scalp which bled on touch, with a high clinical suspicion of malignancy. Biopsy done at Tonga was suggestive of epithelioid sarcoma. Whole body PET CT scan did not show any uptake elsewhere in the body except in the scalp. Wide excision of the lesion on the frontal region of the scalp was done with 3 cm margin as shown in the Figure 1. Considering the size of the defect, an

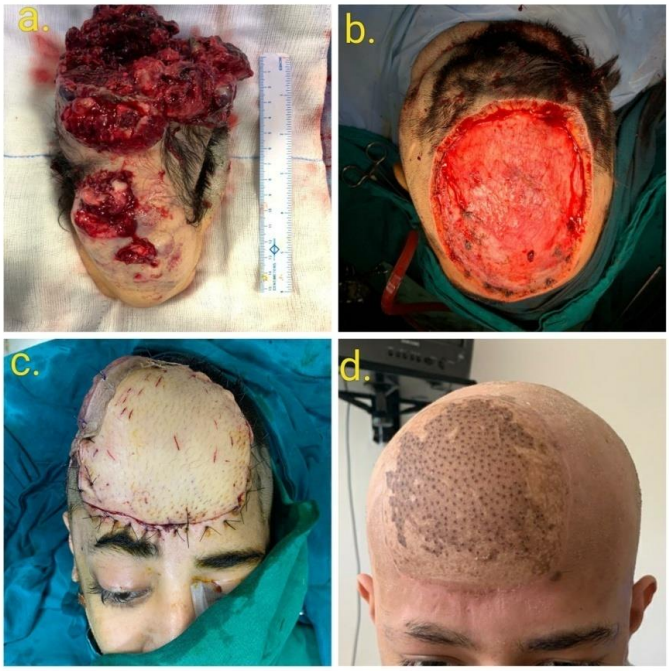


**Figure 1: Case Presentation 1: Showing a. Excised specimen b. Scalp defect post excision c. Reconstruction with anterolateral thigh flap d. Follow up**


**Figure 2: Case Presentation 1: Showing anastomosis to superficial temporal artery and vein using venous grafts (markings 1 and 2)**

anterolateral thigh flap was planned for reconstruction. Superficial temporal vessels were isolated and prepared through a pre-auricular approach. Reconstruction of the defect was done using an anterolateral thigh flap and split thickness skin graft. Vein grafts harvested from the thigh were used to bridge the gap between the donor and recipient vessels (Figure 2). The final histopathology of the excised specimen was consistent with an undifferentiated sarcoma with a possibility of an atypical fibroxanthoma. All margins were free of malignancy. Patient was advised adjuvant Chemotherapy and Radiotherapy considering the high grade histology of the tumor. Radiation of 60 Gy was given to the tumor bed in 30 fractions. Post radiation, he received 6 cycles of Adriamycin and Ifosfamide. Unfortunately his follow up whole body PET CT scan revealed multiple lung metastases. There was no loco-regional relapse.

### Case Presentation 2

A 60 year old male patient presented to the head and neck department with the complaint of multiple, large, foul smelling, and non healing ulcers over the scalp region since one year. He reported a progressive growth in the size and number of the ulcers which gradually involved the entire scalp region. Clinical examination revealed multiple ulcerated and fungating lesions over the scalp in various regions with the largest measuring 10 X 8 cm on the right occipital region. On neck examination, a 5 X 4 cm node was identified on the left level V region. The biopsy from one of the lesions along with IHC

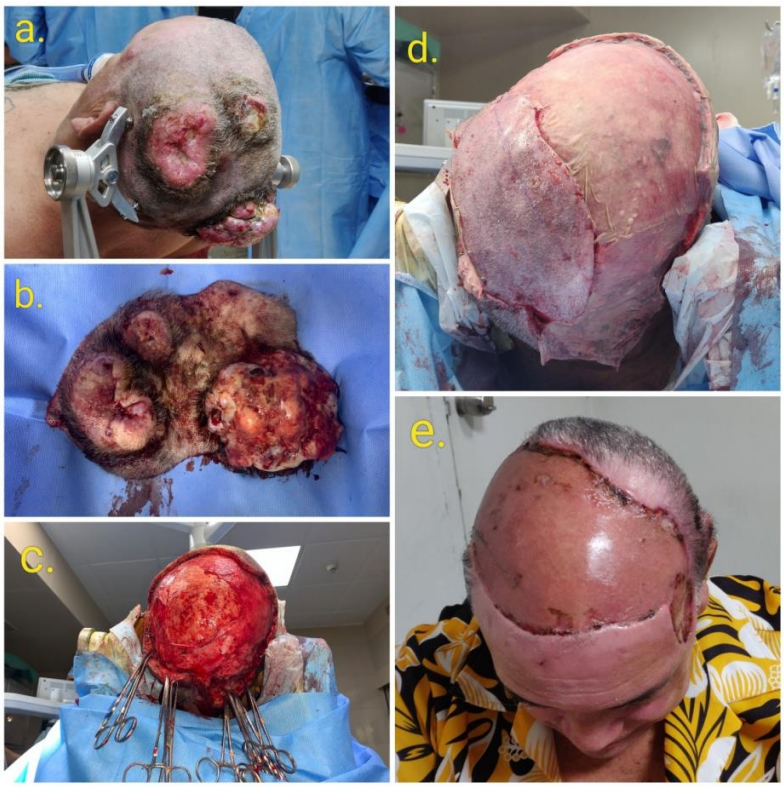


**Figure 3: Case Presentation 2: Showing a. multiple ulcerated lesions b. Excised specimen c. Scalp defect post excision d. Split thickness skin graft insitu e. Follow up**

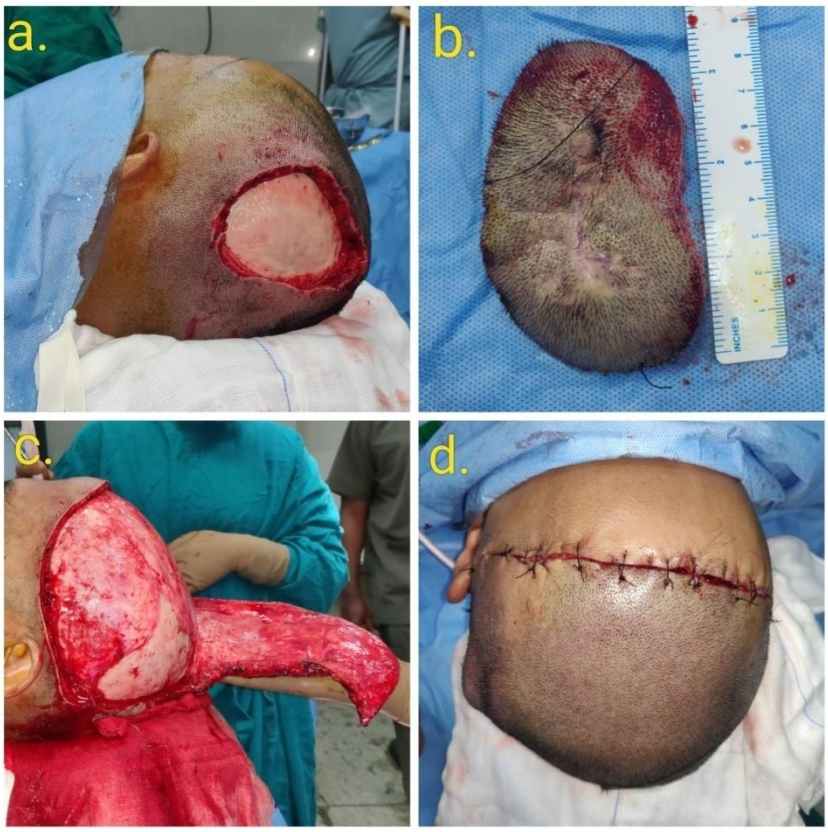


**Figure 4: Case Presentation 3: Showing a. Scalp defect post excision b. Excised specimen c. Raised anteriorly based scalp rotation flap d. Closure**

studies was reported as an Angiosarcoma. PET CT scan was suggestive of extensive multiple scalp lesions with no erosion of the bone and presence of left neck secondaries. Surgical treatment was planned after evaluating the patient's general state and stage of the disease. Wide excision of the

extensive lesions on the scalp along with neck dissection was done. Pericranium was also excised in the areas with pericranial infiltration. A single lesion over the left temporal region was excised separately. Multiple frozen sections were done in the suspicious areas and all were reported negative for invasive malignancy. Reconstruction was done using anteriorly based rotation of the remaining temporal scalp over the area where bone was exposed and the remaining defect was reconstructed using split thickness skin graft from right thigh (Figure 3). Left postero-lateral neck dissection was done clearing level II, III, IV, V with sacrifice of sternocleidomastoid muscle and preserving internal jugular vein and spinal accessory nerve. Radiation was planned from the 3rd week post surgery in view of the aggressive nature of the disease. A radiation planning PET scan was done & is suggestive of multiple lung nodules and hence radiation was deferred and palliative chemotherapy was considered. There was local disease progression during the course of the treatment and best supportive care was offered to the patient.

**Case Presentation 3**

A 30 year old male patient, a known case of dermatofibrosarcoma of scalp s/p excision with positive margins in the histopathology report presented to the head and neck out patient department for further management. Clinical examination revealed a 4 X 3 cm scar over the left parietal region with no other lesions or ulcers elsewhere on the scalp. PET scan did not show any uptake elsewhere in the body. Surgery was planned and wide excision was done with 3 cm margin around the scar up to the bone. Reconstruction of the defect was done with an anteriorly based scalp rotation flap as shown in the Figure 4. Patient is presently doing well and free of any relapse.

**DISCUSSION**

Sarcomas of H&N soft tissues are unusual malignancies. Risk factors for the soft tissue sarcomas include genetic abnormalities, irradiation and conditions like type I neurofibromatosis, Li-Fraumeni syndrome etc. [1]. Environmental factors also contribute to the development of sarcomas. Scalp sarcomas usually present as progressive, necrotic lesions of size more than 5 cm and with irregular borders and irregular septa [1]. They present as a painless mass [2] with diverse symptoms depending on the site of involvement. Scalp sarcomas are best imaged by a computed tomography or magnetic resonance imaging.

The rapid progression of sarcomas follows an extensive mechanism. Due to the local inflammatory response, the adjacent tissue forms a pseudo capsule which is filled with malignant and inflammatory cells [3]. Clinical examination of the tumor reveals the extension of tumor along the fascial planes, bone and muscle and this dictates the necessity of en bloc resection of the tumor with adequate safe margins [2]. Each and every case requires personalization of the available treatment options.

Surgical resection and reconstruction is the recommended treatment modality for the soft tissue sarcomas of the scalp [1]. Radiotherapy is advised for cases with poor prognosis due to a positive margin or advanced histologic grade [1]. Histological types like rhabdomyosarcoma and angiosarcoma are prone to higher recurrence than other variants [6], which implicates the histologic grade as an important predictive factor of prognosis. It is reported that 10 to 40% of the sarcomas develop distant metastasis, most commonly to the lung [4,5]. Local recurrence is usually predicted by the margin status and the grade.

**Undifferentiated Pleomorphic Sarcoma**

Pleomorphic sarcoma is a rare tumor originating from the mesenchymal tissue. It is clinically and histologically similar to the atypical fibroxanthoma. The common features between these tumors include pleomorphism, presence of atypical mitotic figures, spindle or round cell histology and CD10 marker positivity [7]. Being more aggressive, the pleomorphic sarcoma occasionally shows perineural invasion (PNI), necrosis, lymphovascular invasion (LVI) and/or deeper layer involvement [8].

In the case presented here, the initial biopsy of the lesion reported it as epithelioid sarcoma, but the final histopathology revealed an undifferentiated pleomorphic sarcoma without any LVI or PNI and also suggesting another possibility of atypical fibroxanthoma. In view of the deeper pericranial involvement, the diagnosis of an undifferentiated pleomorphic sarcoma was entertained. However, considering the size of the tumor and its highly vascular nature, a wide surgical excision with 3 cm safe margin was performed in the above case. Studies also favoured the fact that surgical excision should always be the primary modality in treating a pleomorphic sarcoma [9]. Tardio et al reported pleomorphic sarcoma as a more aggressive tumor than previously estimated, with 20% local recurrence when incompletely resected and 20% distant metastasis after the treatment [10]. Unfortunately, the above case reported back with multiple lung metastases though there was no loco-regional relapse. Within the limited literature available regarding the management of pleomorphic sarcoma, it is of the essence for the surgeons to treat this tumor aggressively surgically with adequate margins.

**Angiosarcoma**

Angiosarcoma is a rare soft tissue head and neck sarcoma mostly occurring in the face and scalp in elderly patients and it varies from well differentiated to poorly differentiated. Angiosarcoma doesn't usually associate with any pre-existing condition [11]. Differentiation of angiosarcoma of the scalp and face from the benign vascular lesions is often difficult.

Microscopic examination reveals the involvement of dermis, whereas deeper structures are found involved in the poorly differentiated cases. High grade angiosarcomas are usually poorly differentiated and the low grades ones are well differentiated. However, the survival rate has no association with the grading of angiosarcoma [12,13]. The treatment outcome of angiosarcoma depends on the tumor size and potentiality of excising the tumor completely with wide margins [14]. The primary objective in treating angiosarcoma is controlling the local spread of the tumor. Studies reported surgery as the primary treatment modality for Angiosarcoma and also to routinely provide adjuvant radiation therapy to improve the survival [13]. In our case, there were multiple scalp lesions involving pericranium and also with neck secondaries. We had treated the patient with wide resection and reconstruction. Considering the presence of multiple lung nodules, the planned adjuvant radiation therapy was deferred and palliative chemotherapy was given to the patient. However, there was local disease progression too despite extensive resection indicating its aggressive nature and propensity for early recurrence. Understanding this fact, the only way to ameliorate the patient is by early diagnosis and performing adequate surgical resection. Long term follow up is mandatory to detect any late distant metastasis.

### Dermatofibrosarcoma

It is a nodular cutaneous tumor with a notable storiform pattern. Initially, the tumor manifest as a firm, plaque-like skin lesion surrounded by a discoloration [15]. The paucity of deeper scalp tissue suggests fixation of the tumor to the underlying periosteum. Dermatofibrosarcoma is a highly aggressive tumor with almost 50% to 75% of patients suffering recurrence [16,17]. The best treatment for dermatofibrosarcoma is wide local resection with more than 5 cm margins [18]. It has been reported that, an excision margin of 2 cm or less will result in 41% recurrence whereas, a margin of 3 cm results in 20% recurrence [19]. There is no literature suggesting radiation or any other treatment modality benefitting the management of dermatofibrosarcoma [15]. The patient mentioned in the above case had undergone excision of the lesion elsewhere and reported to us with positive margins, we re-excised the lesion with 3 cm margins and post-operatively he is doing well without any relapse for the past one year.

## CONCLUSION

Soft tissue sarcomas of the scalp are a diverse group of unusual tumors. Effective use of various diagnostic and therapeutic measures will improve the treatment outcome. Clinical trials dictating the treatment options for the majority of scalp sarcomas are still limited. A multidisciplinary treatment plan is always needed in treating these kinds of tumors and it should be based on the available standard literature. Future studies and case series on these rare but aggressive tumors will definitely play a role in improving the survival rate of sarcomas.

## ACKNOWLEDGEMENTS:

We would like to acknowledge the Department of Radiation oncology, Department of Medical Oncology and the Department of Pathology for their support in managing these patients.

The authors would also like to acknowledge the patients mentioned in the case series.

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