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Case Report

Cutaneous Carcinosarcoma of the Scalp - A Case Report and Literature Review

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Abstract: Carcinosarcomas are a group of biphasic tumours. They are composed of co-existing malignant epithelial and mesenchymal components. They are extremely rare in the skin. We present a case of a 43 year old female patient who came as a referral from a local district hospital with a chronic painless mass for 7 years. It had ulcerated 6 months prior to presentation with bleeding. The scalp mass was resected with a 10 mm margin. The histological diagnosis was carcinosarcoma as both carcinomatous and sarcomatous. Diagnosis of carcinosarcoma rests on demonstration of pleomorphic spindle cells with pressence of both epithelial and mesenchymal elements within the same tumour. The biphasic nature of the tumour may be seen on light microscopy showing atypical squamoid epithelial cells admixed with mucoid secreting variants as shown in our case. Immunopositivity to both cytokeratin and vimentin on a background of the described light microscopy findings confirms the diagnosis. Generally, surgery with resection of at least 10mm margin is the primary therapeutic modality. Local spread is rare. In conclusion, cutaneous carcinosarcoma of the scalp is rare and management is patient dependant with surgery being the mainstay treatment.

Keywords: Carcinosarcoma, Cutaneous Fibro-sarcoma, Surgical Resection

1. Introduction

Carcinosarcoma is a biphasic tumour which is composed of 2 different malignant tumour components occurring in a variety of organs [1]. The oronasopharynx, larynx, the gastrointestinal systems, kidneys, uterus, lungs and skin have been involved in some cases [2–4]. Primary carcinosarcoma of skin is very rare and literature has less than 60 cases reported to date. However identification of both the elements is critical since these tumours are frequently associated with a poor prognosis especially the visceral ones. In reported cases of carcinosarcoma, the epithelial component was basal cell carcinoma, squamous cell carcinoma or adnexal carcinoma of the skin. The mesenchymal component included osteosarcoma, chondrosarcoma, fibrosarcoma or malignant fibrous histiocytoma. We report a rare case of primary cutaneous carcinosarcoma presenting as an ulcerated scalp mass in a 43 year old female patient.

2. Case Presentation

A 43 year old female patient presented to us with a slowly enlarging scalp mass for 7 years. It had ulcerated with bleeding since 6 months prior to presentation. She had developed shortness of breath and general body weakness. She was hemodynamically unstable with a low grade fever. Had a negative HIV status with no history of smoking and also negative family history of skin cancers.

Her examination had significant conjunctival and palmar pallor and was ill looking. She had an ulcerated mass on the

right frontoparietal scalp measuring about 12x8x6cm, firm and mobile with clearly defined circumferential edges and non-pulsatile as seen in Figure 1. The mass was noted to have areas of sepsis. Patient was managed with antibiotics and transfused with packed cells then after stabilising was taken to theatre for excision of the mass which was uneventful. A 10 mm resection margin was achieved. The mass was sent to the laboratory for histological diagnosis. The scalp defect was closed with primary split skin graft which took well as shown in Figure 2.

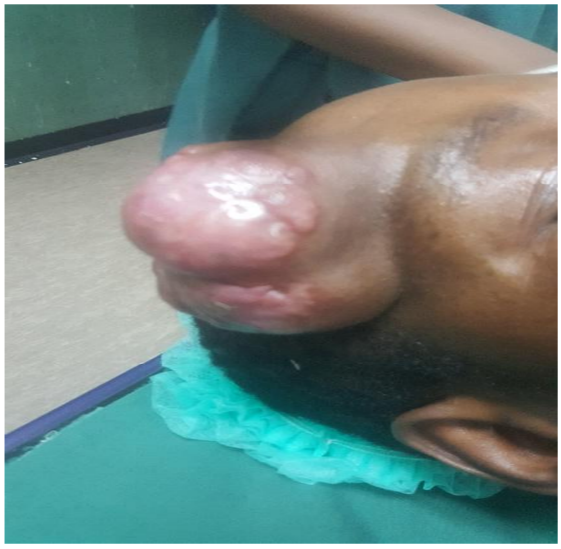

Figure 1. *The macroscopic picture of the carcinosarcoma on the frontoparietal scalp.*

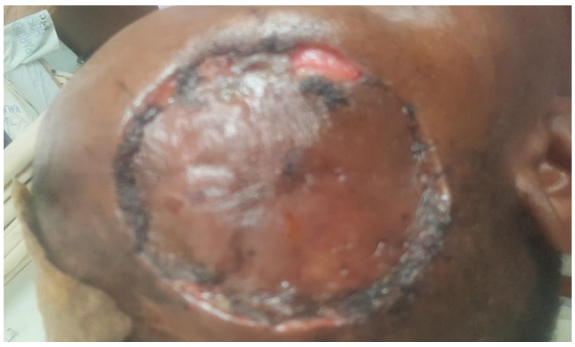

Figure 2. *Day 4 post operation showing a well taken split skin graft.*

Macroscopic examination confirmed an extensive fungating and ulcerated rubbery tumour that had 2mm clear off the tumour from the deep margin, and an average of 12mm cut margins in the anterior, posterior, medial and lateral sides.

Microscopic findings confirmed a scalp mass showing ulcerating tumour comprised of anastomosing cords, and nests of polyhedral atypical cells with eosinophilic to pale cytoplasm (Figure 5 and 6). There were areas with pleomorphic spindle cells, squamoid atypical cells and mucin secreting atypical cells were identified. The tumour was noted to be biphasic with both carcinomatous and sarcomatous elements. It was positively immunoreactive to cytokeratin and vimentin which was consistent with features of a carcinosarcoma (figures 3 and 4).

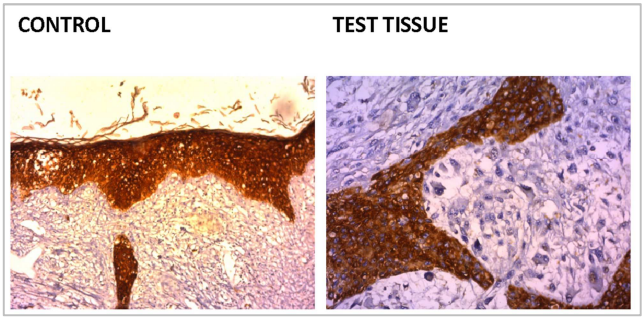

Figure 3. *Cytokeratin immunohistochemistry positivity.*

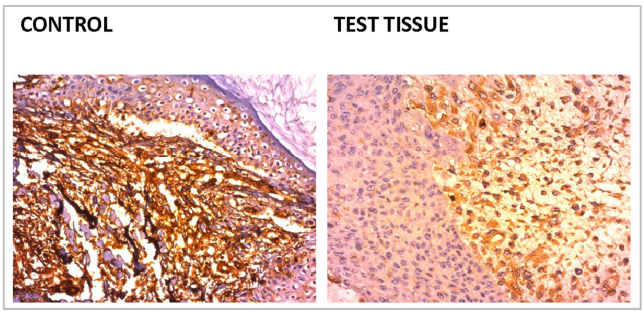

Figure 4. *Vimentin immunopositivity.*

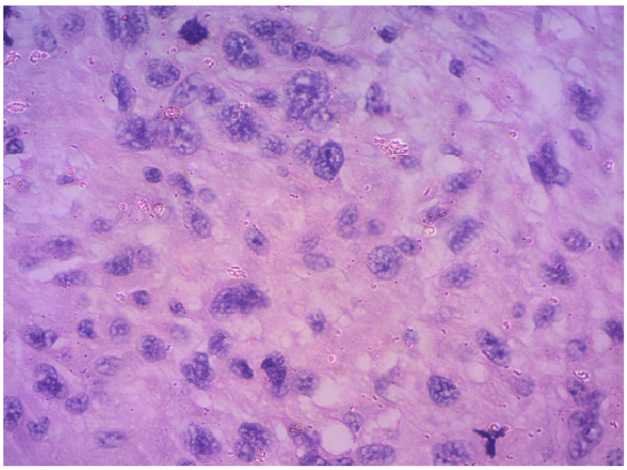

Figure 5. *Sarcomatous component with bizarre mitotic figures.*

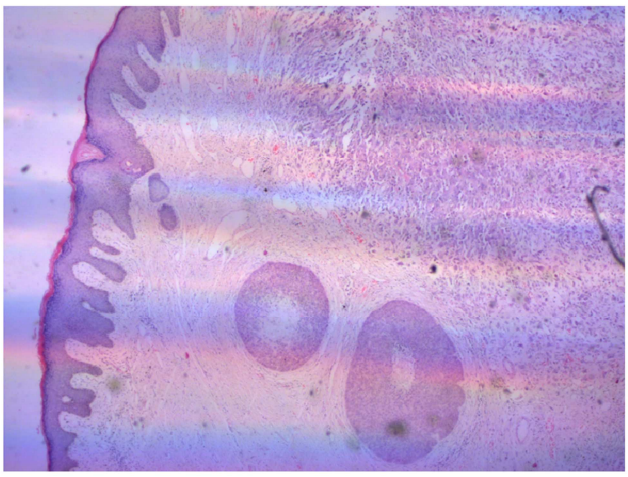

Figure 6. *A mixture of sarcomatous and carcinomatous components.*

3. Discussion

The first case of primary carcinosarcoma was published in 1972 by Dawson etal [12]. It is very rare [13]

Four main hypotheses have been postulated for the histogenesis of carcinosarcoma [5]. These have been based on the pathology of the disease as seen in other organs such as the female genital tract and lungs [5, 6] The collision tumour hypothesis proposes a smash of two independent tumours resulting in a single neoplasm. This is based on the observations that skin cancers and superficial malignant fibrous histiocytomas (sarcomas) are commonly seen in patients with sun damaged skin. In this hypothesis, the epithelial and mesenchymal components are believed to come from different undifferentiated progenitor cells. This is supported by the fact that the 2 different tumour components have no common features as observed through immunohistochemical and ultrastructural differentiation [7, 8]. Secondly, the monoclonal hypothesis suggests that the undifferentiated, totipotential neoplastic cells undergo multiple pathways of terminal differentiation into histologically recognisable mesenchymal and epithelial elements. This is based on the fact that the different tumours have common features which demonstrate the clonal origin [4, 5].

The combination hypothesis suggests that the mesenchymal components represents a pseudosarcomatous reaction to the epithelial malignancy. The final hypothesis is called the diversion/conversion hypothesis which regards the sarcomatous components of the tumour as a product of metaplasia from the epithelial component. However despite the various explanations as to the origin of these tumours, the medical advances in immunohistochemistry and molecular genetics suggest and favour the hypothesis of monoclonality in carcinosarcoma [5, 6].

Loss of chromosome 9p and to some extent abnormality has been found in both the mesenchymal and epithelial components with great consistency. The chromosome 9p has the tumour suppressor gene p16 which suggest its possible role in the pathogenesis of these tumours [4, 10]. Dedifferentiation can lead to a phenotypic change [14]. Insufficient data has been reported to draw conclusion on predisposing factors, but the history of previous skin tumours and anatomical distribution of lesions suggest that the sun exposure may be a factor [11].

Our case had both epithelial and mesenchymal components. This was evidenced by vimentin and cytokeratin immunopositivity. This demonstrated the existence of 2 different tumours, one of epithelial origin (squamous cell carcinoma) and the other of mesenchymal origin (fibrous sarcoma). These tumours have different histogenic mechanism. The carcinomatous component had a dominant contribution of 60% in comparison to the sarcomatous part. The collision tumour theory could fit into the explanation of our case as it occurred in a sun exposed area of the frontoparietal scalp and particularly in typical hard working African woman who exposed to the scotching heat of the sun.

Locoregional spread is rare [13]. Surgical resection with removal of deep fascia affording a 10 mm margin is advised [7]. The sarcomatous type has poor response to chemotherapy and radiotherapy [7]. Long term follow up is advised for optimal results on this very rare entity [7].

4. Conclusion

Primary carcinosarcoma are rare tumours of unknown aetiology and it is important to recognise the 2 malignant components of the tumour based on both the morphogenic and immunohistochemical features. The theories about histogenesis may be applicable to some tumours in particular locations but not all cases. More studies are needed in order to understand the exact natural history of these tumours.

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