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Case Report

Admixed foamy cell and signet ring cell change in a cutaneous angiosarcoma – A diagnostic pitfall

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ABSTRACT

Cutaneous epithelioid angiosarcomas are known to exhibit various morphological variations. The present case is first of its own kind describing admixture of foamy cell and signet ring cell morphological patterns in a case of cutaneous angiosarcoma occurring in a 64-year-old female. The initial incisional biopsy revealed predominantly cytoplasmic vacuolations in the tumor cells with only focal presence of typical angiosarcomatous component, while in subsequent resection specimen, these variations were present only focally with predominant classical angiosarcomatous areas. This case highlights the importance of awareness of such morphological variations in angiosarcomas as these can be easily missed or misdiagnosed in small biopsies due to sample selection.

1. Introduction

Cutaneous angiosarcoma is the malignant neoplasm of endothelial cells primarily affecting elderly males typically on the head and neck. These tumors grow rapidly and have extremely infiltrative margins making them difficult to excise completely, and hence carry a poor prognosis. Several cytoplasmic alterations are described in epithelioid cell variant of cutaneous angiosarcoma and include foamy cell, signet ring cell and granular cell angiosarcoma.[1–3] These variants can be diagnostically challenging and can mimic a variety of benign and malignant lesions. We recently came across a case of cutaneous angiosarcoma with admixture of foamy and signet ring cells, a rare entity.

2. Case report

A 64-year-old female, known case of hypothyroidism and type II diabetes mellitus, noticed a small swelling over left side of chin 6 months back. It was gradually increasing in size and was not associated with pain or any discharge. On local examination, the swelling was 6 cm × 3 cm in size and firm in consistency. The overlying skin was erythematous and showed patchy bluish black discoloration. The lesion was not adhered to the underlying bone. The oral mucosa corresponding to it was also normal. An incision biopsy followed by wide local excision of the lesion was performed. No past history of any malignancy was present.

The skin biopsy measured 1 cm × 0.8 cm × 0.6 cm. Sections from the biopsy showed thinned out epidermis with an

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infiltrating tumor involving whole of the dermis and subcutaneous fat (Fig. 1A). The tumor was composed of sheets and cords of bland looking, largely polygonal cells dissecting through the dermal collagen and entrapping the skin appendages. The cells were variably sized, closely packed and fused with each other at places. They showed abundant multivacuolated (bubbly) to univacuolated clear cytoplasm. Mucin stain was negative in the cytoplasmic vacuolations. Some of the cells showed central while other showed peripherally placed indented and compressed nuclei with mild pleomorphism and indistinct nucleoli (Fig. 1B). Few inconspicuous foci showed classical angiosarcomatous pattern (constituting <5% of total biopsy area) composed of intercommunicating irregular vascular channels lined by cells having eosinophilic cytoplasm and plump nuclei (Fig. 1C). The tumor also showed significant lymphocytic infiltrate with occasional lymphoid aggregate formation (Fig. 1D). No significant nuclear pleomorphism, mitotic activity or tumor necrosis was identified. On immunohistochemistry (IHC), the tumor cells were positive for CD31 and CD34 (Fig. 2A and B). In focal areas, tumor cells were also positive for CD68 (Fig. 2C). Ki67 proliferation index was about 10% (Fig. 2D). The stains for CK and EMA were performed to rule out metastasis from a carcinoma. Both were found to be negative. An initial diagnosis of angiosarcoma, foamy cell variant was given on the biopsy.

Subsequent wide local excision specimen was received measuring 9 cm× 6 cm × 2.5 cm, which showed a bosselated tumorous area with congested and focally excoriated overlying skin (Fig. 3A). Serial slicing showed a poorly circumscribed gray white tumor in the dermis measuring about 6 cm × 5 cm which was focally extending into the subcutaneous tissue. The tumor was firm in consistency (Fig. 3B). Sections from the tumor confirmed the diagnosis of angiosarcoma. The predominant tumor component showed features of classical angiosarcomatous pattern comprising vascular arrangement with plump atypical endothelial cells. The morphological change described above (vacuolated cells) was noted only focally (comprising about <20% area of total tumor including the initial biopsy and excision specimen). So a final diagnosis of angiosarcoma with focal foamy cell change was given on excision specimen. There were no postoperative complications. Subsequent PET CT scan of the patient did not reveal any metastasis.

3. Discussion

Foamy cell and signet ring cell variants of angiosarcoma are rare and only handful of cases with such morphological patterns are described in the literature. On reviewing the exact cellular morphology and IHC findings described in the

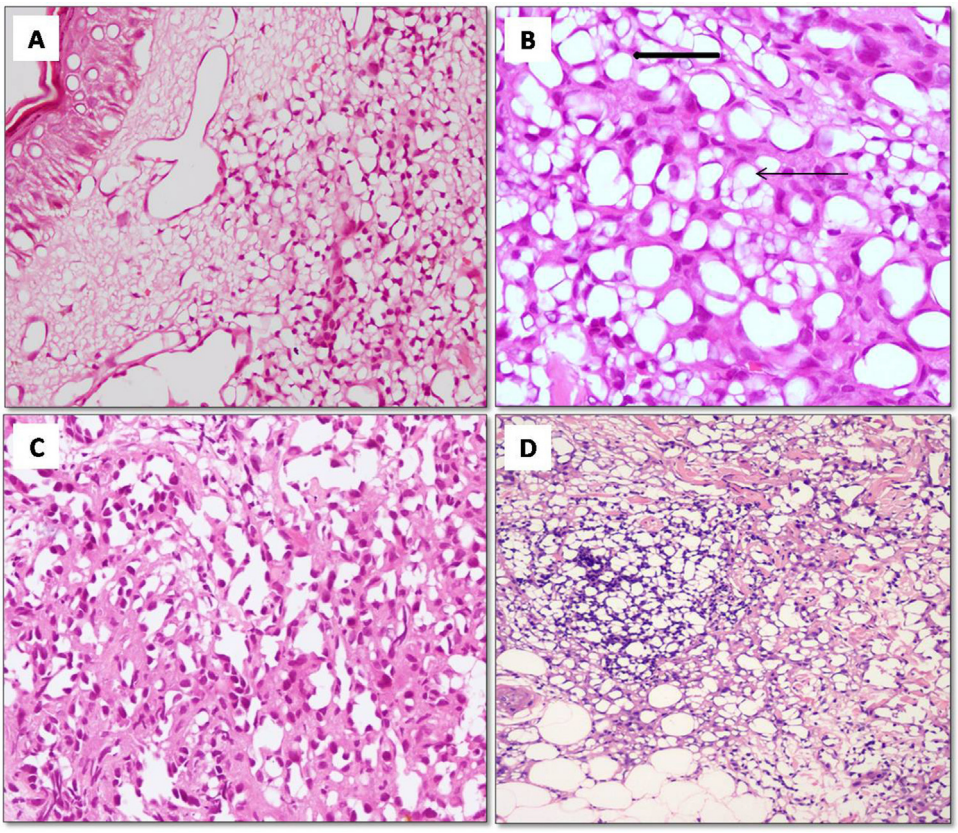

Fig. 1 – Microphotographs showing tumor in the dermis (A – H&E 20×); bland looking tumor cells with abundant clear cytoplasm. Some of the cells showing central (thin arrow) while some showing peripherally pushed nucleus (broad arrow) (B – H&E 40×); typical angiosarcomatous areas with intercommunicating channels lined by atypical endothelial cells (C – H&E 40×) and tumor with lymphoid aggregates (D – H&E 20×).