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MYOEPITHELIAL CARCINOMA OF SOFT TISSUE- A CASE REPORT

General Surgery

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ABSTRACT

Soft tissue has been recently recognized as a primary site for myoepithelial tumour and only a small number has been reported to occur in the head and neck. Although the majority of myoepithelial tumours of the soft tissue are benign, some can be aggressive and develop distant metastases.1 We describe a patient with malignant myoepithelial tumour occurring in the soft tissue of the upper back. In contrast to previous publications warning for overtreatment of patients with myoepithelial tumours, our report suggests that early and correct diagnosis followed by a radical therapeutic approach is required when there is evidence of malignancy.

KEYWORDS

Soft tissue; Myoepithelial carcinoma; Head and neck cancer; Unilateral upper back mass

INTRODUCTION

Myoepithelial carcinoma of soft tissue is extremely rare although its counterpart of the salivary gland is relatively common and well-known with similar morphology.[2] In addition myoepitheliomas have been found to occur also in extrasalivary locations, such as soft tissue,[3,4] skin,[5-7] breast[8] and lung[9]. The separate entity of myoepithelial tumour arising primarily in the soft tissue was recognized for the first time about 23 years ago.[3-10] Only 15% of all these soft tissue myoepitheliomas occur in the head and neck region.[4] The great majority is found in the limbs and limb girdles. Although myoepitheliomas of the soft tissue are usually located in the subcutaneous tissue, less than 30% occur in the deep soft tissue.[4] The broad morphological spectrum of myoepithelial tumours probably is the mean reason why they were not easily recognized before.

Here we are reporting a case of myoepithelial carcinoma of soft tissue in upper back.

CASE REPORT

A 60-year-old female presented with swelling over right upper back for last six years. Initially was operated 10 years back (for which no documents were available). The swelling was insidious in onset and gradually progressive over last 6 yrs. On examination a 8x5 cm in size lump with well-defined margins, firm in consistency and reduced mobility noted over right suprascapular region. Overlying skin showed a healed old surgical scar.

USG UPPER BACK:

Multiple heteroechoic lesions in intra/inter muscular planes on right upper back Largest measuring 2.4x4.7 cm Minimal cystic lesions and internal vascularity

FNAC:

FNAC from the lesion revealed a cellular smear comprising of monomorphic spindle cells along with stromal fragments

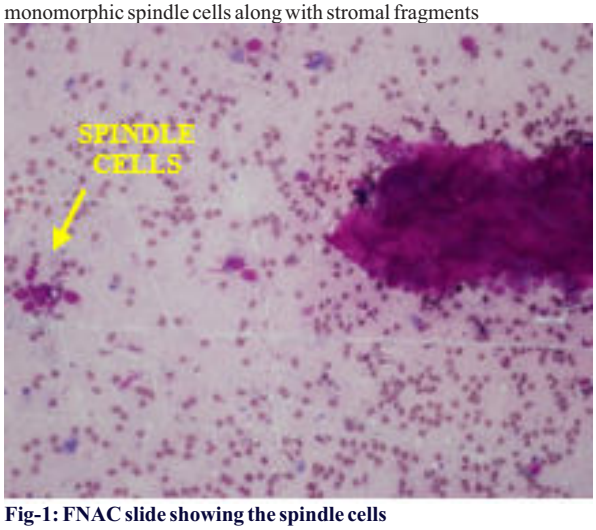

Fig-1: FNAC slide showing the spindle cells

MRI NECK & CHEST:

Multiple discrete/conglomerated, lobulated heterogeneous signal intensity mass lesion in right suprascapular region from C5 to D1 vertebral level Lesion seen involving trapezius muscle with 8 x 5 x 4.2 cm in maximum dimensions Predominantly hyperintense on T2W, hypointense on T1W images.

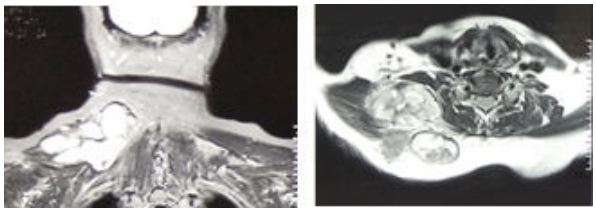

Fig-2: Coronal and sagittal section showing the tumour in relation to the trapezius

CORE CUT BIOPSY FROM THE LESION:

Showed fibro collagenous tissue only. Repeat Biopsy also was inconclusive.

TREATMENT:

In view of inconclusive tissue biopsy reports patient was taken up for surgery on the basis of FNAC report with WLE under GA Intra-op: Multi-lobulated, dumbbell shaped (superficial and deep to trapezius) soft tissue tumour with areas of myxoid degeneration. Areas of adhesions to old scar present. No significant adhesions in depth. Significant feeding vessels noted.

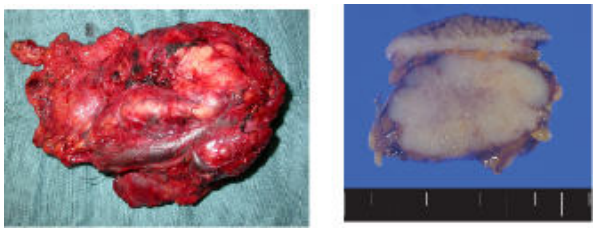

Fig-3: Macroscopic view of excised tumour with yellowish nodular surface and greyish tan cut surface

HPE:

Myoepithelial Carcinoma of Soft Tissue with deep surgical margins involved by the tumour

On IHC tumour cells were positive for S100, Cytokeratin, EMA, GFAP and p63.

FOLLOW UP:

Patient was reviewed in view of positive margin in HPE and was re-evaluated.

MRI NECK & THORAX:

Scar tissue is seen extending from skin till trapezius muscle at operated site. It appears hypointense on T1W1, T2W1 and hyperintense on STIR. Two well-defined lesions measuring 5.5 x 12 x 8.3 mm & 5.8 x 13.2 x 7.7 mm are noted in the lateral most and the medial most aspect of right trapezius muscle. They are hypointense onT1W1,

hyperintense on T2W1/STIR and show avid post contrast enhancement

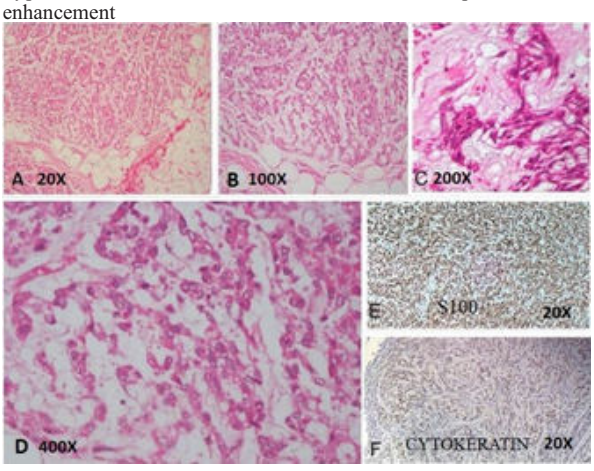

Fig-4: A to C: H & E staining showing the tumour arranged in lobules in a background of the abundant chondromyxoid stroma. D: This tumour showed focally high-grade morphology with sheets of cells with moderate amount of cytoplasm, vesicular chromatin, and prominent nucleoli. E & F: IHC

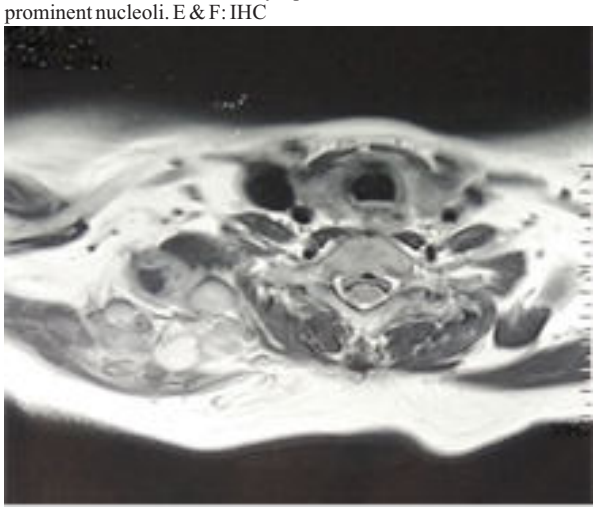

Fig-5: Axial section showing the residual tumour in respect to trapezius muscle

Patient underwent Exploration with revision of margins. Intra-op: 1 x 1 cm residual nodule medially over trapezius. Trapezius resected laterally proximal to acromion and medially proximal to C7 process

HPE:

Myoepithelial Carcinoma with margins free of tumour

IHC: Tumour cells were positive for S100, p63, SMA, GFAP and EMA

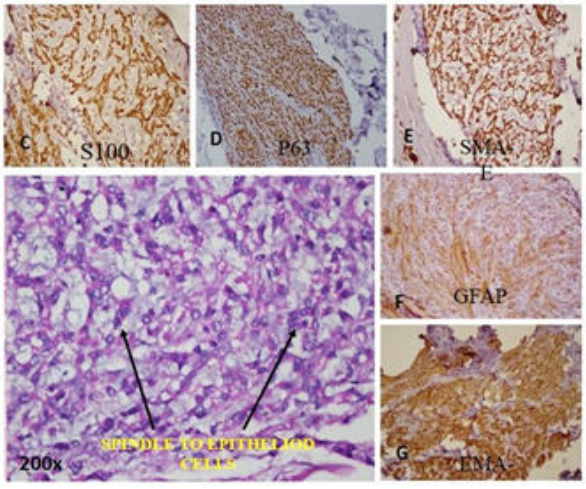

Fig-6: HPE of second resection showing the similar histological findings of previous HPE

DISCUSSION:

Primary myoepithelial tumours (myoepithelioma, myoepithelial carcinoma) are rare soft tissue tumours, which primarily occur in subcutaneous tissue. They affect male and female equally over a wide age range, with a peak incidence between the third and fifth decades; approximately 20% of cases occur in pediatric patients, in whom they are frequently malignant.[12] Approximately 70% of cases are distributed in the soft tissue region, while the remainder of cases are found in the cutaneous areas, viscera and bones.[13] Very few studies are available on demographic and clinical data of this tumour. Studies have shown that status of margin very well equates to the incidence of local recurrence. As much as 50% of cases develop metastasis to distant organs usually at a median interval of around 2 years after initial diagnosis.

Myoepithelial carcinoma of soft tissue is morphologically identical to that of salivary gland. Although myoepithelial carcinoma in the salivary gland is a well-known tumour, only few cases of myoepithelial carcinoma of soft tissue have ever been reported because of their rarity.[3]

Myoepithelial tumours with no to mild cytologic atypia (low-grade) can be classified as myoepithelioma, whereas tumours with moderate to severe atypia (high-grade) can be classified as myoepithelial carcinoma (vesicular or coarse chromatin, prominent, often large nucleoli, or nuclear pleomorphism).[4] High-grade cytologic atypia in myoepithelial tumour is a clear indication of malignancy. But the diagnosis of malignancy is much more difficult if there is a lack of cytologic atypia, since cytologic atypia are not required for the diagnosis of myoepithelial carcinoma.[13]

The heterogeneity of immunophenotype and various histologic patterns and cell types of myoepithelial carcinomas of soft tissue make the differential diagnoses broad, including metastatic carcinoma, malignant melanoma, proximal-type epithelioid sarcoma (ES), extraskeletal myxoid chondrosarcoma (EMC), epithelioid malignant peripheral nerve sheath tumor (MPNST), and poorly differentiated synovial sarcoma.[11] Immunohistochemistry is absolutely important for diagnosis of myoepithelial carcinoma of soft tissue because of its morphological similarity to other neoplasms.

Complete excision with clear free margins is the mainstay of treatment. In case of histopathologically involved margin status and/or nodal deposits, post-operative radiotherapy is advocated.

CONFLICTS OF INTEREST:

No potential conflict of interest relevant to this article was reported

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