Review Article

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Malignant Mixed Tumour Chondroid Syringoma of the Skin: A Case Report and Literature Review

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Abstract

Malignant mixed tumour of the skin is a rare adnexal tumour seen more commonly in females and has propensity to recur locally and present with delayed metastasis. Wide excision is the treatment of choice and final diagnosis is made after histopathological report. We present a case of malignant mixed tumour of forearm in a male.

Keywords: Malignant mixed tumour; Skin; Recurrence.

Introduction

Worldwide, a considerable morbidity is caused by malignancies of skin.[1] Dermal malignancies can be of epidermal origin (squamous cell carcinoma, basal cell carcinoma and malignant melanoma) or of adnexal origin; Basal cell carcinoma and squamous cell carcinoma are known to account for approximately 98% of these.[1,2] Adnexal tumours are rarer than epidermal tumours and can originate from follicular, sebaceous, apocrine or eccrine lineage.[3] Mixed tumour (chondroid syringoma) is a follicular and folliculosebaceous apocrine hamartoma which is benign and Malignant Mixed Tumour (MMT) is its malignant counterpart.

MMT is exceedingly rare and accounts for less than 0,005% of all dermal malignant epithelial neoplasms with less than 50 case reports in literature.[4,5] Synonyms include – malignant chondroid syringoma, cutaneous malignant mixed tumour, metastasising chondroid syringoma, aggressive chondroid syringoma and atypical mixed tumour of skin.[6] The diagnosis is confusing and it has propensity for recurrence.

We present such a case of MMT in upper limb in an adult male.

Case Summary

A 57 year-old male presented with complaints of swelling over left forearm since last 15 years. At examination, the patient was stable and a swelling of 4x4 cm was present over medial aspect of left forearm, soft in consistency, mobile, non-tender, not fixed to underlying structure or overlying skin; prominent dilated veins could be seen (Fig. 1). He had history of similar swelling at same site 15 years back for which excision was done. So, further evaluation was done keeping the clinical diagnosis of lipoma or hemangioma in mind.

Local ultrasonography suggested a well-defined, heterogenous, hypoechoic lesion in subcutaneous plane with increased internal vascularity with few cystic areas of degeneration. Fine Needle Aspiration Cytology suggested adnexal tumour. Wide local excision with 2 cm margins was done.

Histopathology revealed presence of benign stratified squamous epithelial lining with

subepithelial tissue showing biphasic tumour composed of myxoid and spindle shaped stromal cells along with glands invading into the subcutaneous adipose tissue suggesting malignant mixed tumour of skin (Fig. 2). Immunohistochemistry confirmed low grade Malignant Adnexal tumour, favouring malignant mixed tumour with CK7 positive and SOX 10 Positive (Fig. 3).

The patient, though asymptomatic on follow–up, was advised detailed investigation.

Computed Tomography (CT) of thorax, abdomen and pelvis was done which suggested lytic lesion in left ischium with enhancing soft tissue components within (size: 2.5x3.5x4.5cm) eroding the bony acetabulum and preserved femoral head (Fig. 4). Small nodules were also noted in lungs. CT guided biopsy was done from the posterior acetabular column which suggested metastasis from malignant mixed tumour.

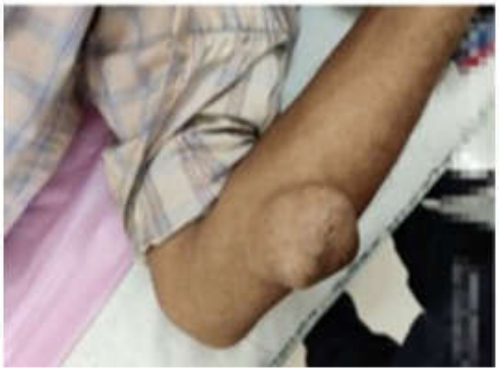

Fig. 1: Clinical image of the forearm swelling

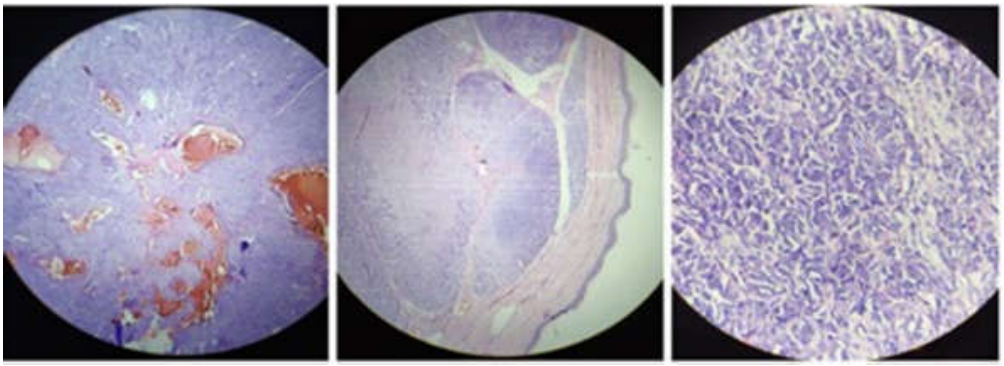

Fig. 2: Histopathology images showing presence of epithelial cells set within a myxoid and/or chondroid stroma

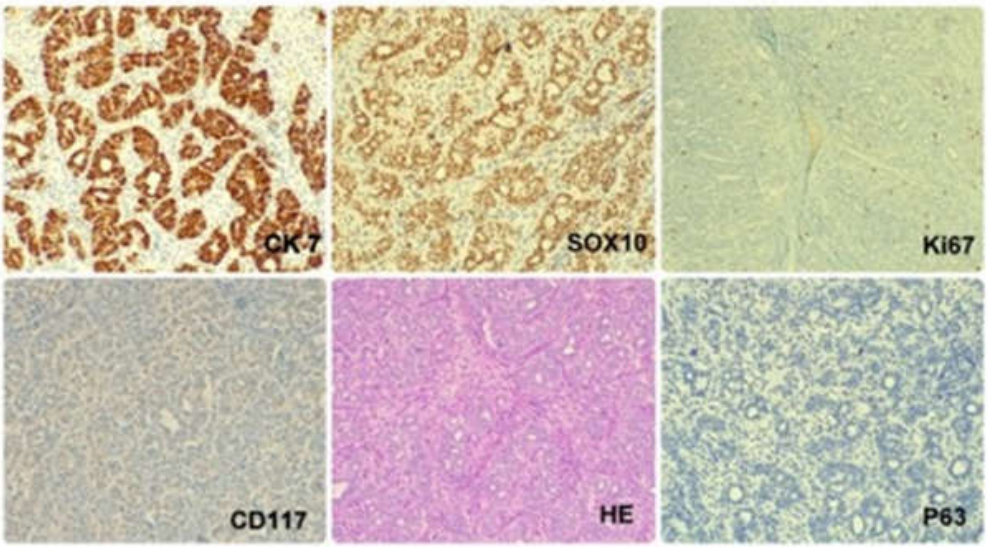

Fig. 3: Immunohistochemistry confirmed low grade Malignant Adnexal tumour, favouring malignant mixed tumour with CK7 positive and SOX 10 Positive

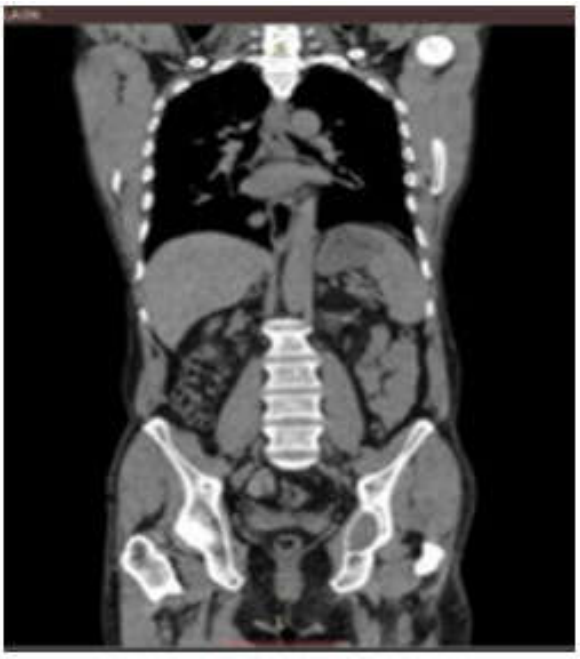

Fig. 4: CT suggesting lytic lesion in left ischium eroding the bony acetabulum with preserved femoral head

Discussion

Malignant mixed tumour (MMT) is an extremely rare cutaneous adnexal malignancy with a significant risk for aggressive behaviour, local recurrence and a propensity for delayed metastasis.[1,4] It occurs in a wide age-range and is twice as common in females than in males.[1,4] MMT shows a predilection for the trunk and the extremities. MMT may be confused clinically with many benign and malignant lesions. Therefore, the histopathological and immune-histochemical (IHC) examination is required for accurate diagnosis and management.

First described by Hirsch and Helwig in 1961, Malignant mixed tumour may arise de novo or rarely develop in benign mixed tumour.[1,7] The benign counterpart is commoner in the head and neck region and had no sex predilection.[8,9] It is an uncommon neoplasm of sweat gland origin and has histological similarity to salivary gland mixed tumours (pleomorphic adenomas) – there is epithelial glandular component admixed with myoepithelial/mesenchymal stromal component.[4]

MMT can occur at any age; however, the majority of cases have been reported in adults.[4] It clinically presents as a non-specific nodule or mass of size ranging from 2 to 10 cm in greatest dimension.[4]

Clinically, MMT may be confused with many benign and malignant lesions.[1] Hence, histopathological and immunohistochemical (IHC) examination is essential to establish accurate diagnosis.[1] This ifferentiation of MMT from other skin malignancies is important clinically because of the difference in its prognosis and treatment from the more commonly encountered dermal cancers.[10]

In 1961, Headington divided mixed tumours of skin into two groups based on histopathology - apocrine and eccrine type.[11] A plausible origin of MMT can be myoepithelial.[12] MMT displays histopathological features similar to its benign counterpart - composed of epithelial cells arranged in nests, cords, and glands set within a myxoid and/or chondroid stroma – with presence of atypia.[4] Some depict a deceptively bland histologically (such are mostly aggressive) and some depict markedly atypical histology; diagnosis of MMT being then made based on the focal presence of myxoid or chondroid stroma.[4] Features suggesting malignancy include infiltrative growth pattern, nuclear pleomorphism, increased and/or atypical mitotic activity, tumour necrosis and lympho-vascular invasion.[4] At IHC, the epithelial component of often expresses cytokeratins, epithelial membrane antigen (EMA) and carcinoembryonic antigen (CEA).[4] The myoepithelial cells in the stromal component typically co-express cytokeratin and S100 protein and may also express other myoepithelial markers such as glial fibrillary acidic protein (GFAP).[4]

Differential diagnosis of MMT includes mainly extra-skeletal myxoid chondrosacroma and mucinous carcinoma.[1] Extraskeletal myxoid chondrosarcoma consists of non-cohesive elongated tumour nests without ductal or tubular structures; the cells being cytokeratin negative.[12] Mucinous carcinoma has distinct PAS positivity of the extracellular myxoid stroma.[12] Other differential diagnoses include, sarcomatoid carcinoma (carcinosarcoma) with chondrosarcomatous areas, matrix producing melanoma, metastatic chondrosarcoma, chondroma of soft parts, ossifying fibromyxoidtumor of soft parts, and extra-axial soft tissue chordoma.[4] In the present case, the histopathology and IHC showed CK7 positive and SOX 10 Positive suggesting MMT.

The clinical course of MMT is unpredictable. The available literature reports local recurrences, regional metastasis and distant metastasis in 50% of cases.[13] Nodal metastases and distant metastases have been reported in 39% and 36% of the cases, respectively.[4] The commonest site for distance metastasis has been reported to be lung, followed by bone andbrain; but widely disseminated metastases have been reported in the end stage of

the disease.[4,13,14] MMT are known to have a slow progression and prolonged course.[4] The overall mortality has been reported to be 25%.[4]

Complete excision of the tumour with wide margins remains the mainstay of the treatment.[1] Subsequent clinical and radiologic follow-up is required to detect local recurrence or nodal and/or distant metastases.[4] Skeletal metastases have been reported torespond to radiotherapy.[1] Combination chemotherapy has not been reported to be beneficial in metastasis.[13]

Literature suggests only few case reports of this entity. Two cases were reported by Trown et al. in 1994[9] and by Nel et al. in 2016.[6] Famulski et al. have reported a similar case in a 56 year old lady with MMT with emphasis differential diagnosis.[1] Recently, two cases have been reported by Kothiya et al., one of whom developed late metastasis to lungs.[15]

Conclusion

MMT, being rare, complete excision with wide disease-free margins, correct histopathological diagnosis and adequate follow-up are required for disease-free survival.

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Permissions: Nil

References

1. Famulski W, Kanczuga–Koda L, Niewiarowska K, et al. Malignant mixed tumour of the skin: a case report and review of the literature. Prog Health Sci 2013;3(1):154–7.
2. Galadari E, Mehregan AH, Lee KC. Malignant transformation of eccrine tumours. J Cutan Pathol 1987 Feb;14(1):15–22.
3. Alhumidi AA. Simple approach to histological diagnosis of common skin adnexal tumours. Path Lab Med Int 2017;9:37–47. Doi: https://doi.org/10.2147/PLMI.S139767
4. Gardner JM, Smoller BR. Malignant mixed tumour of skin. In: F Rongioletti et al. (eds), Rare Malignant Skin Tumours, Springer Science+Business Media New York 2015;77–80.
5. Tulenko JF, Conway H. An analysis of sweat gland tumors. SurgGynecol Obstet. 1965 Aug; 21:343–8.
6. Nel CE, Byl DVD, Grayson W. Malignant Chondroid Syringoma. Dermatopathology 2019;6:77–84.
7. Hirsch P, Helwig EB. Chondroid Syringoma: Mixed tumor of skin, salivary gland type. Arch Dermatol 1961;84:835–47.
8. Shvili D, Rothem A. Fulminant metastasizing chondroid syringoma of the skin. Am J Dermatopathol 1986 Aug;8(4):321–5.
9. Trown K, Heenan PJ. Malignant mixed tumour of the skin (malignant chondroid syringoma) Pathology. 1994 Jul;26(3):237–43.
10. Steinmetz JC, Russo BA, Ginsburg RE. Malignant chondroidsyringoma with widespread metastasis. J Am AcadDermatol 1990 May;22(5 Pt 1):845–7.
11. Headington JT. Mixed tumors of skin: Eccrine and apocrine types. Arch Dermatol 1961 Dec;84:989–96.
12. Aleida LM, Sangueza OP, Kutzner H. Cutaneous myoepithelial neoplasms: Clinicopathologic and immunohistochemical study of 20 cases suggesting a continuous spectrum ranging from benign mixed tumor of the skin to cutaneous myoepithelioma and myoepithelial carcinoma. JCutanPathol 2003 May;30(5):294–302.
13. Takahashi H, Ishiko A, Kobayashi M, et al. Malignant chondroidsyringoma with bone invasion: A case report and review of the literature. Am J Dermatopathol 2004 Oct;26(5):403–6.
14. Araujo JL, de Aguiar GB, do Prado Aguiar U et al. Malignant chondroidsyringoma with central nervous system involvement. J Craniofac Surg 2012 Mar;23(2):514–5.
15. Kothiya M, Mittal N, Kumar R et al. Malignant chondroidsyringoma of thigh with late metastasis to lung: A very very rare case report. Indian J PatholMicrobiol 2017;60:418–20.