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A Case of a Malignant Cutaneous Mixed Tumor (Chondroid Syringoma) of the Scapula with Spinal Cord Compression Secondary to Metastasis: A Case Report

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Abstract **Case Report**

Malignant chondroid syringoma is a rare cutaneous tumor of mixed epithelial and mesenchymal origin. It is the malignant counterpart of benign chondroid syringoma and has a potential for local invasion and distant metastasis, although such progression is extremely uncommon. A 60-year-old Mediterranean man presented with a painful, ulcerative 6-cm lesion on his right scapula, progressively enlarging over one year. Biopsy confirmed malignant chondroid syringoma. Imaging revealed local tissue invasion, pulmonary nodules, bone metastases, and spinal cord compression due to vertebral metastases (T1–T4), leading to lower limb weakness and paralysis. The patient was referred for radiotherapy and neurosurgical evaluation. Malignant chondroid syringoma is difficult to diagnose clinically due to nonspecific features. Histological evaluation, especially with hematoxylin and eosin staining, is essential for accurate diagnosis. Immunohistochemistry may support but does not definitively distinguish malignant from benign forms. Treatment involves wide excision with negative margins; however, adjuvant radiotherapy or chemotherapy may be necessary in metastatic cases. The tumor's unpredictable behavior necessitates long-term surveillance. This case represents the first report from Africa, and only the second globally, of malignant chondroid syringoma presenting with spinal cord compression. Given its aggressive potential, early diagnosis, complete excision, and long-term follow-up are critical. Radiotherapy may be considered for unresectable or metastatic lesions.

Keywords : Chondroid syringoma, Aggressive, radiotherapy.

INTRODUCTION

Cutaneous mixed tumor (chondroid syringoma) is the cutaneous counterpart of pleomorphic adenoma of the salivary glands, comprised of both epithelial and mesenchymal components. Malignant transformation is exceptionally rare, with only a few cases reported. We report a case of a malignant cutaneous mixed tumor occurring on the scapula of an 60-year-old man.

CASE REPORT

An 60-year-old mediterranean man presented to our plastic surgery department for a 6-cm ulcerative budding indurated plaque on his right scapula, Slightly mobile, painful and bleeding on contact, which he reported had been present and evolving in size for 1 year. He had no medical history. A shave biopsy was performed. Histologic examination revealed malignant chondroid syringoma.

Computed tomography was performed showedsubcutaneous lesional process, locally infiltrating right parascapular with necrotic center, to be compared with histological data. Pulmonary nodules and micronodules associated with secondary mixed bone lesions.

While being admitted at the hospital he showed a progressive weakness and numbness in the lower limbs. he had been experiencing difficulty with walking, and her neurological deficit had increasing worsened over the preceding 15 days. Without being able to walk and MRI signal abnormality in the dorsolumbosacral somatic area of secondary appearance. appearance of

vertebral metastases, with epidural and intra-canal extension involving the T1, T2, T3 and T4 stages.

He was referred to oncology with concern for these metastasis No less / Decompressive radiotherapy; dosimetric scanner performed and neurosuregry was contacted telling that a Laminectomy (decompression surgery) or by radiotherapy then coverage would be necessary.

Based on the morphologic features and immunoprofile, the patient was diagnosed with a malignant chondroid syringoma.

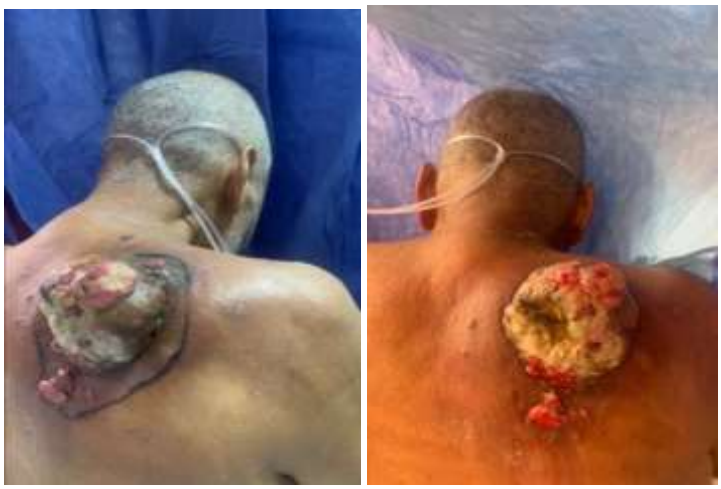

Figure 1: chondroid syringoma of the scapula

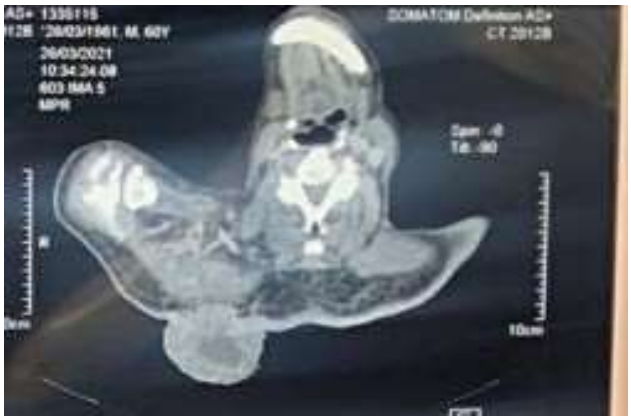

Figure 2: CT SCAN

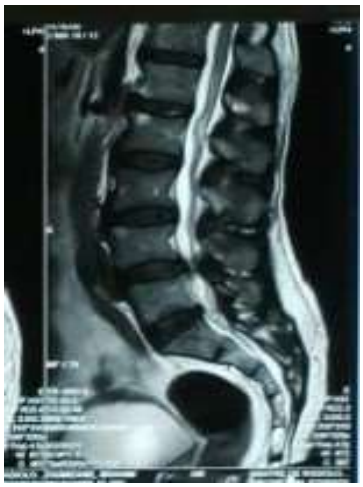

Figure 3: Admission sagittal T2-weighted MR images of the thoracic spine revealing an extradural mass projecting into the spinal canal and causing anterior displacement of the spinal cord to the right

DISCUSSION

Malignant chondroid syringoma is a rare tumor of epithelial and mesenchymal origin with nonspecific clinical features. It has been reported to occur de novo or in a preexisting benign chondroid syringoma. Benign mixed cutaneous tumor of the skin was first described in 1859.

Further specification in 1961 delineated the tumor as eccrine in origin with a cartilaginous component, reidentifying the tumor as a chondroid syringoma, which later was determined to have a malignant counterpart [1–3]. Reversing the distribution of benign chondroid syringomata, malignant tumors are more prevalent on the trunk and extremities, and relatively unusual on the head and neck. Although these tumors most commonly present in middle age, they have been reported in patients as young as 13 years [4].

The differential diagnosis of these clinically nondescript lesions is broad. The patient's age, location, and the prevalence of NMSCs led us to suspect an NMSC in our patient. Histopathologic assessment often demonstrates both an epithelial component with eccrine or apocrine differentiation and a mesenchymal component consisting of osteoid, mucinous, and/or cartilaginous differentiation [5]. Benign chondroid syringomata may exhibit atypical features, and thus, deep biopsies including the entire lesion are often required to assess for features of a malignant diagnosis such as infiltration [5]. Cellular features on hematoxylin and eosin (H&E) that favors malignant tumors include large size, increased mitotic activity, atypical mitoses, infiltrative margins, cytologic atypia, necrosis and perineural, and/or lymphovascular invasion [6]. Immunohistochemistry alone has not been found helpful in clarifying the diagnosis. In a study of 50 benign chondroid syringomata, all samples with available tissue were diffusely positive for S100 and cytokeratin [7] with uniform negativity for p63, epithelial membrane antigen, cytokeratin 5/6, and calponin [7]. This immunoprofile of benign lesions matches that of our malignant case. As other authors have noted, it is the H&E features that suggest a malignant process [6]. Treatment of these tumors consists of wide excision with confirmation of negative margins. There is no clearly defined role for adjuvant treatment, although chemotherapy and adjuvant radiation have been used [2,6]. A case of malignant chondroid syringoma treated by Mohs micrographic surgery has been published with the authors presenting the benefit of tissue preservation. This has to be weighed against the fact that these tumors have an unpredictable course, with a potential for local and distant metastasis. In a case series of 39 patients, 50% had local recurrences, 24 had nodal and distant metastases, and 10 died of metastatic disease [6]. Late distant or lymph node metastasis or delayed malignant transformation of a benign chondroid syringoma is possible, with one such transformation reported 20 years after initial diagnosis [5]. Commonly reported sites of metastases include lymph nodes, lung, and bone [2]. Invasive spread has involved the central nervous system [8]. With possible metastatic potential in mind, extended surveillance for metastasis to multiple systems is prudent.

This case highlights the importance of obtaining adequate tissue for histologic evaluation, as partial biopsies can lead to confusion with metastatic adenocarcinoma or other skin neoplasms because of their clinically ambiguous presentations. In this case, immunohistochemistry was consistent with an adnexal tumor. However, the H&E morphology rather than immunoprofile distinguished the malignant tumor from its benign counterpart. Staged margin-controlled excision was used in this instance to evaluate en face margins on excision. Long-term follow-up is still recommended because of the risk of local recurrence and metastasis to lymph nodes, bone, and lung.

CONCLUSIONS

To the best of our knowledge, this is the first report of spinal cord compression caused by a metastasis of a malignant chondroid syringoma, diagnosed in Africa. The second in the world [9] Given the rarity of this tumor and the lack of published experience in the therapeutic management of spinal secondary lesions, Resection followed by radiotherapy may be an acceptable approach to achieve short-term, progression-free survival. Radiotherapy should be tried in future cases of incomplete excision or as a palliative therapy. Because disease recurrence including metastasis will occur in a high percentage of cases, long-term followup is required.

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