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Case Report

Primary Penoscrotal Myxoid Chondrosarcomas

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Abstract

Introduction: Extraskeletal myxoid chondrosarcoma (EMC) is a rare malignancy that is classified as a tumor of uncertain differentiation by the World Health Organization. It usually presents as a slow growing mass in men in their fifth to sixth decades.

Presentation of the case: Here we report 4-year follow up of a man with penoscrotal EMC that had been managed by complete tumor excision.

Conclusion: Due to such a low incidence of EMC with primary origin of scrotum, a therapeutic approach is very challenging but wide excision with definite free margins seems to be the best treatment.

Keywords: Extraskeletal myxoid chondrosarcoma; MRI; Histopathology; Scrotum

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Consent: Consent was taken from the patient for publication of this case report.

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Introduction

Chondrosarcoma which was first defined by Stout and Verner is a malignant neoplasm of cartilage that most commonly involves the extremities. The histologic appearances of the tumors described ranged from relatively well-differentiated chondrosarcoma to that of satellite or rounded chondroblasts lying in a myxoid matrix having a distinctly nodular pattern. Extraskeletal myxoid chondrosarcoma (EMC) is a rare malignancy that is classified as a tumor of uncertain differentiation by the World Health Organization. It usually presents as a slow growing mass in men in their fifth to sixth decades. Genitourinary involvement is extremely rare and just less than 10 cases have been reported. Here we report 4-year follow up of a man with penoscrotal EMC that had been managed by complete tumor excision.

Case Presentation

A thirty-eight-year-old man was admitted with complaint of a non-painful penoscrotal mass of seven months' duration with sudden growth in the last 20 days. The patient's medical history was unremarkable. Physical examination revealed a firm, non-tender moveable penoscrotal mass which was discrete from the both testicles and penile shaft (Figure 1).

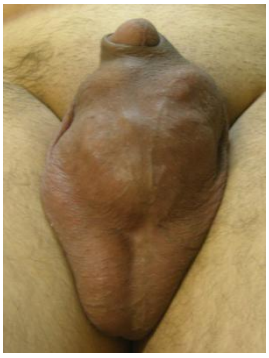

Figure 1 Large lobulated penoscrotal mass

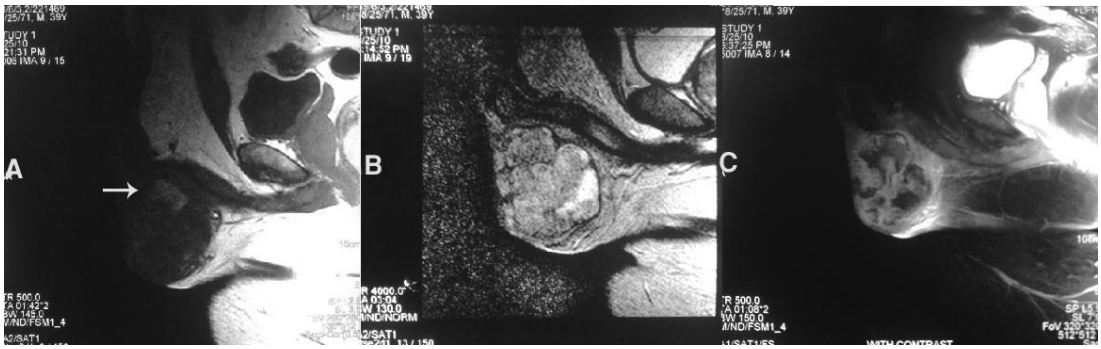

Figure 2 A: T1-weighted MR image reveals a well-defined penoscrotal mass of low signal intensity containing a high signal focus (arrow) implying hemorrhage. B: On T2-weighted image the mass shows multi-nodular architecture with intermediate to high signal intensity. C: After contrast administration the mass enhances in a heterogeneous pattern.

In ultrasonographic evaluation, the mass was of intermediate, heterogeneous echo with internal small cystic lacunae. On T1 and T2 weighted magnetic resonance (MR) images, the mass was of low and high signal intensity, respectively. T1-weighted magnetic resonance (MR) images revealed a lobulated well defined homogeneous mass of low signal intensity except for a high signal focus implying hemorrhage in its superior portion. On T2-weighted images the mass showed multi-nodular architecture with intermediate to high signal intensity and contained internal low signal septations. After administration of gadolinium-based contrast agent the mass shows moderate to high enhancement in a heterogeneous pattern. No extension to corpora cavernosa or corpus spongiosum was detected and testes were intact (Figure 2).

The patient underwent a midline penoscrotal exploration. The mass was obviously extratesticular, encapsulatd, without any adhesion to surrounding tissues. The tumor was completely removed (Figure 3).

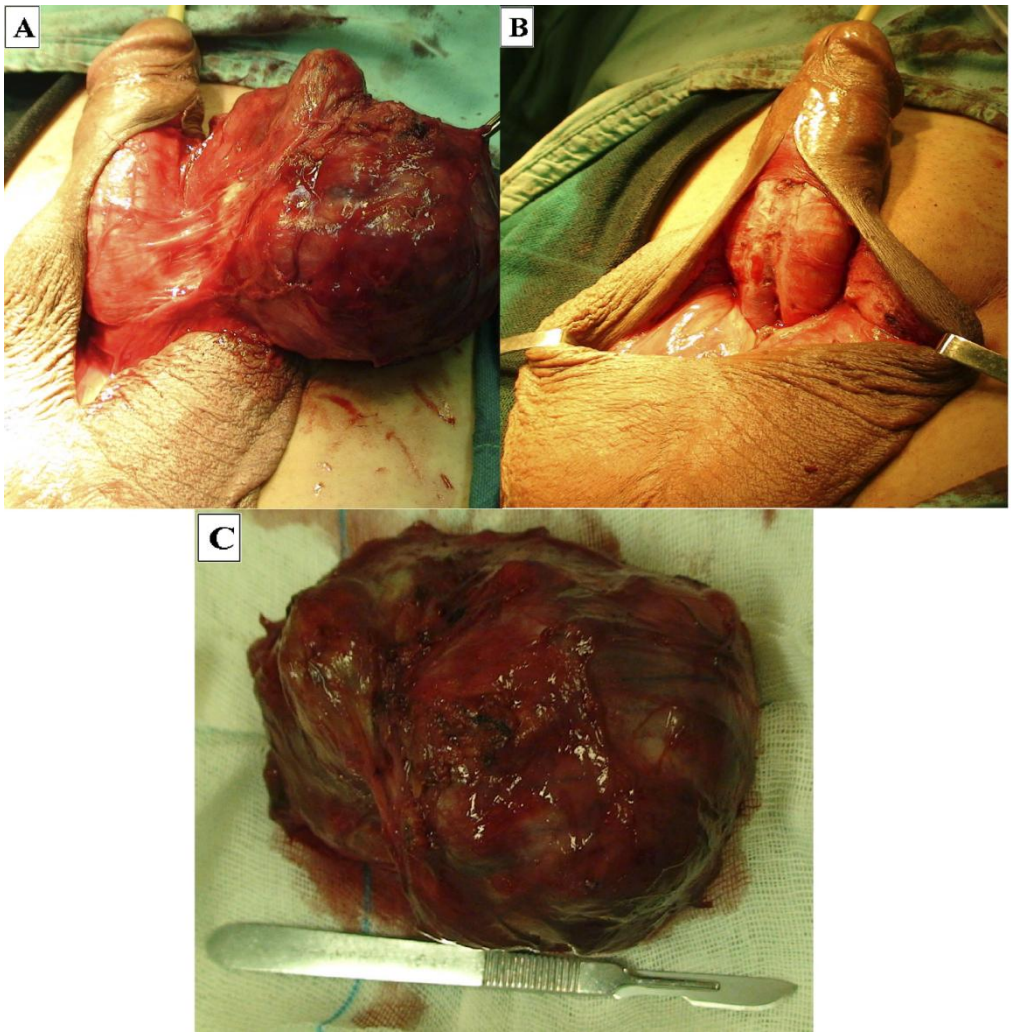

Figure 2 Intra-operative appearance of a well-circumscribed tumor; A: The mass was discrete from the corpora cavernosa and corpus spongiosum. B: Penoscrotal area after en bloc resection of the tumor. C: Gross specimen reveals encapsulated mass measuring 14*8*5 cm with a creamy external surface

In histopathology, the mass the mass was consisted of myxoid stroma bearing clusters and sheets of round tumor cells having eccentric atypical vesicular and mitotically active nuclei with prominent nucleoli and deeply eosinophilic cytoplasm. (Figure 4) With the use of immunohistochemistry, the neoplastic cells were shown to express CD99, NSE, synaptophysin, S-100 proteins. Expression of cytokeratin, myo-D1 and desmin was not observed.

Computed tomography (CT) scans of the chest, abdomen and pelvis ascertained no metastasis. After 4 years of post-operation follow up, annual clinical examination and follow-up CT scan detected no recurrence, metastasis or any unfavorable behavior.

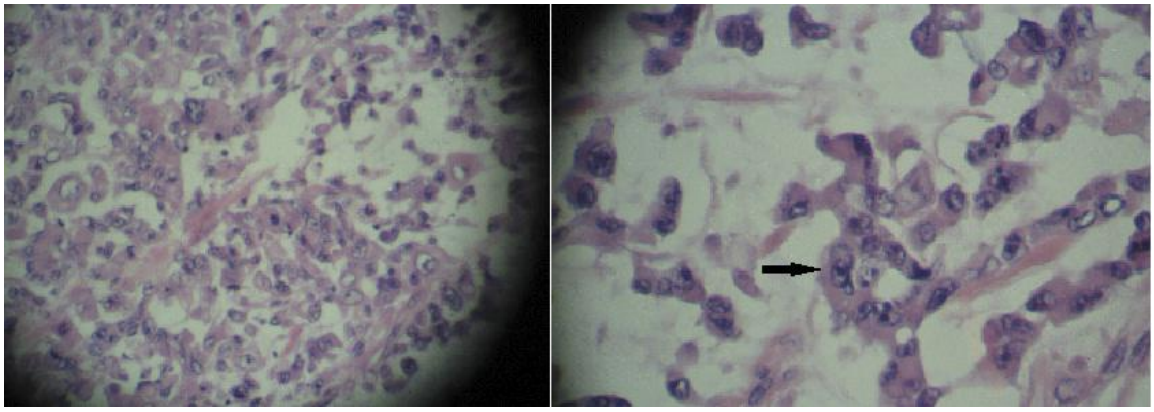

Figure 3 Histopathology of the mass was consisted of myxoid stroma bearing clusters and sheets of mitotically active round tumor cells

Discussion

EMC which was first defined clinicopathologically by Enzinger and Shirak in 1972 accounted for 2.5% of all soft tissue sarcomas [1]. It commonly affects patients aged 50–60 years but is also known to occur in younger people with male to female ratio of 2:1 and it mostly involves proximal extremities and limb girdles. Genitourinary involvement is a relatively uncommon manifestation of EMC, with only a handful of reported cases in the literature since the first report of a tunica vaginalis EMC in 1973 [2]. Patients usually present with non-specific symptoms, including tenderness and palpable mass. In compare to literature our case was a male with EMC in his genitalia and like other reported case of genital EMCs it had a rapid growth. The mass also had the same characterisites of an EMC, described in literature: a lobulated and encapsulated mass with no tenderness. However our patient was younger than the range described in literature.

Apart from revealing the signal characteristics of soft tissue masses, evaluating the extracompartmental extension is one of the most important reasons for performing MRI before surgical planning. High signal focus in T1-weighted images of our case was suggestive of hemorrhage. Although intralesional hemorrhage is common pathologic finding, it is uncommon by MRI. Tateishi et al. found this finding in only one case out of nineteen [3]. The extensive myxoid stroma of myxoid chondrosarcoma has exceptionally high water content with low attenuation by CT and very high signal intensity by T2-weighted MR as with our case [4]. Heterogeneous enhancement pattern of myxoid chondrosarcoma by MRI images is in good accordance with the heterogeneous pathologic features including cystic, hemorrhagic and necrotic areas and has been reported in most case series [1, 2, 5].

EMC has a tendency to reoccur locally with an occasional distant metastasis in the lungs. The behavior of EMC is that the potential of metastatic disease is greater in children than in adults, while local reoccurrence is same in both. However unlike chondrosarcoma of bone, EMC behaves in a less aggressive fashion. As with other soft -tissue sarcomas in adults, surgical resection is

recommended in EMC. EMC typically has a prolonged clinical course with complete resection [1]. However, the behavior of EMC has been reported to be poor, with frequent local recurrences and distant metastases [3-5]. Sixteen percent of our patients developed local recurrences, and metastases occurred in 16% of our cases. EMC seems to have frequent recurrences

On the basis of the clinical, radiological and laboratory findings did not allow us to characterize histologic type of the tumor so the patient underwent a marginal en bloc resection of the mass.

Conclusion

Due to such a low incidence of EMC with primary origin of scrotum, a therapeutic approach is very challenging but wide excision with definite free margins seems to be the best treatment.

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