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Extra Skeletal Mesenchymal Chondrosarcoma - A Rare Case Report

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Abstract: ***Extra skeletal Mesenchymal Chondrosarcoma (EMC) is a rare neoplasm of soft tissues. A number of case reports have been published in literature from time to time, yet it reveals difficulties in treatment and prognostication. Here is a rare case report of an elderly female who presented with a wrist swelling. X-ray revealed a extra osseous soft tissue lesion with specks of calcification. Patient underwent a local excision, which on histopathological examination revealed mesenchymal chondrosarcoma. This case report is an attempt to emphasize the rarity of the lesion, to stress the need for extensive sampling and to include it in the differential diagnosis of soft tissue lesions with calcification, especially in young people. The treatment includes aggressive surgery with chemotherapy.***

Keywords: Mesenchymal chondrosarcoma, aggressive treatment, extensive sampling

1. Introduction

Mesenchymal chondrosarcoma is a high grade neoplasm accounting for 2-10% of all Chondrosarcomas [1-6]. It was first described by Lichtenstein and Bernstein in 1959 [7] . It is commonly said to originate from within the skeleton,[1] though some studies reveal extra osseous sites more common[2,8] . Fifty years had rolled over since its inception as a recognized entity, yet it poses challenge in diagnosis and management. Here we report a case of extra skeletal mesenchymal chondrosarcoma, which is quite rare.

2. Case Report

80 year female presented to the surgery OPD with swelling in the right wrist for 3 months. The swelling was gradually increasing in size. She had no complaints of pain/ loss of appetite /loss of weight, except for mild restriction of movements owing to the size of swelling.

On examination, the swelling measured 10x7.5x7cm. Skin was thinned out and stretched over the swelling and mirrored the bossylated surface of the underlying lesion. No warmth or tenderness noted when palpated. Investigations started with routine blood tests followed by X-ray. Xray revealed a large soft tissue mass in the distal third of forearm with multiple dense chalky white opacities. The underying bone was free of lesion. Patient underwent a wide local excision. Macroscopically, the lesion was large, thinly encapsulated measuring 7.5x7x6cm with a bosselated surface (**Fig 1)**.

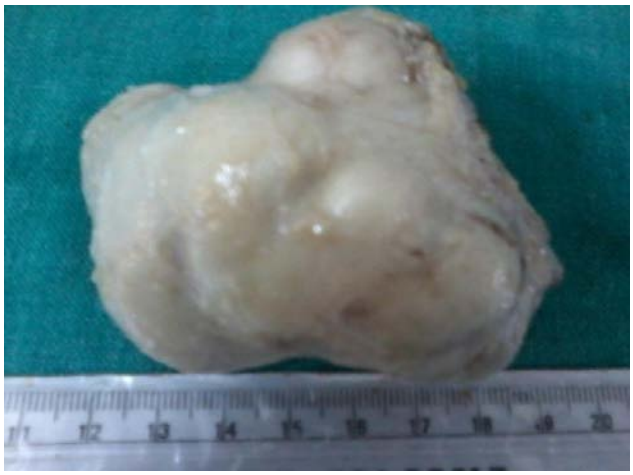

Figure 1: Photograph showing the bossylated outer surface of the tumor.

Cut section revealed a tumor which was grey white, solid, firm with a few glistening nodules (**Fig 2)**.

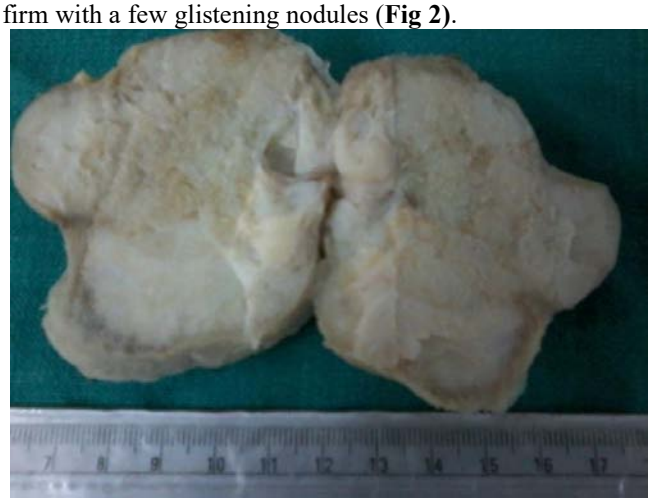

Figure 2: Photograph showing the cut surface of the tumor which is gray white solid with tiny glistening nodules

Microscopically, the lesion was biphasic with malignant chondroid nodules surrounded by undifferentiated small round cells (**Fig 3,4).**

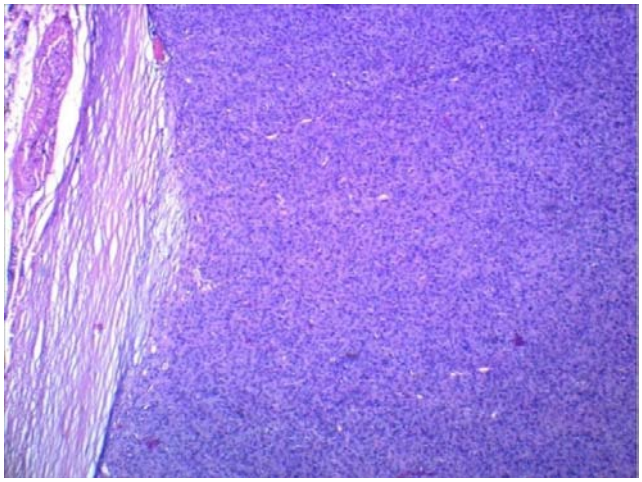

Figure 3: Microphotograph showing partly encapsulated lesion with undifferentiated small cells(H&E, 40x).

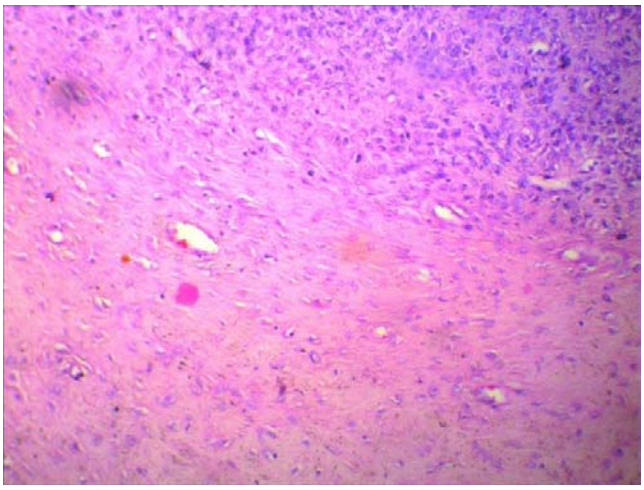

Figure 4: Microphotograph showing the transition between well differentiated chondroid nodules and small cell areas(H& E, 100x).

The nodules had lacunae harboring malignant chondrocytes (some had binucleation) exhibiting moderate to severe nuclear atypia with multiple foci of ossification (**Fig 5)**.These nodules were surrounded by highly cellular, densely packed compact zones of small round to spindly undifferentiated cells arranged in sheets, exhibiting a focal hemangiopericytomatous pattern **(Fig 6)**. Atypical mitotic figures were observed both in chondroid and small cell zones with a mitotic rate of 35/10 hpf. Hence a diagnosis of extra skeletal mesenchymal chondrosarcoma was offered.

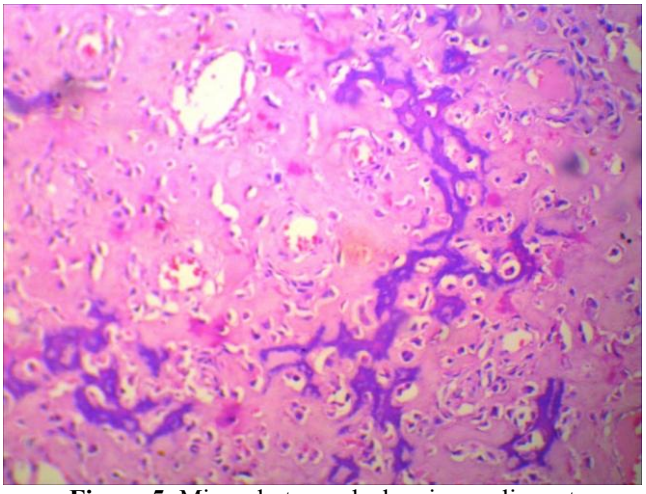

Figure 5: Microphotograph showing malignant chondrocytes with areas of ossification(H&E, 100x).

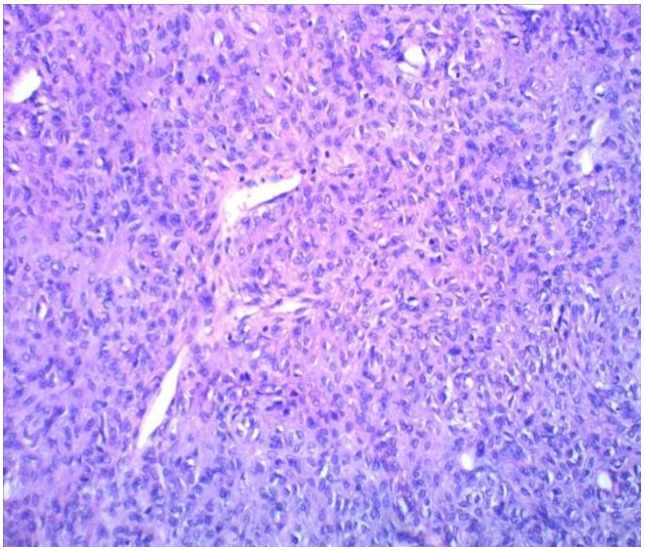

Figure 6: Microphotograph of small round cells showing focal hemangiopericytomatous pattern(H&E, 10x).

3. Discussion

Mesenchymal chondrosarcoma is one among the rarest subtypes of chondrosarcomas.[1] The rarity of the entity has made it difficult to understand its natural history and treatment outcome. Hence, analysis of these rare lesions will give an insight about the knowledge deficient zones of the disease process.

Mesenchymal chondrosarcoma accounts for 25% of all chondrosarcomas occurring in children and young adults, [2,8] where it is most common, in contrast to the subject in our study .Many studies show a female preponderance.[1,2] The most common site is the head and neck (especially orbit) [1,8] followed by chest wall and pelvic bones. In the appendiceal skeleton, the lesion commonly involves the lower extremities.[1] Review of literature shows a number of case reports and few case series involving various sites in head and neck like sino nasal tract and jaw bones. [9,10] Extra skeletal mesenchymal chondrosarcoma arising in the central nervous system,[11] is even rarer. Most of those described in literature are intra cranially located, with a very few cases being reported intraspinally [12] The visceral organs (eg:kidney) too are not totally spared of the tumor.[8,13]

Radiologically, osseous lesions present as a locally destructive, osteolytic lesion with calcified spots and extra osseous extension. Microscopically, the lesion is biphasic with chondroid nodules surrounded by undifferentiated mesenchymal component. [1,2,8] The mesenchymal component is composed of either round or spindle cells or can be a mixture of both. Literature reveals both these subtypes occur in fairly equal proportion.[1] The mesenchymal component is known to exhibit other patterns, most common of which is the hemangiopericytomatous pattern. Other changes include osteoid formation, ossification and calcification. [1]

Diagnosis of mesenchymal chondrosarcoma becomes difficult when the cartilaginous component is absent(due to poor sampling), when it reveals diagnostic challenge masquerading as pure mesenchymal lesions eg. Ewing'ssarcoma, neuroblastoma, leukemia and desmoplastic small round cell tumor.

Immunohistochemically, the undifferentiated cells are positive for vimentin, CD99 and chondrocytes for S100.[1] CD99 is nonspecific as it is positive in Ewing's sarcoma too. New immunohistochemical markers proved useful in distinguishing these lesions include FLI-1, collagen type 2 and SOX 9.[14, 15, 16, 17] In studies conducted by Wehrili et al and Fanburg Smith et al, SOX9 showed positivity in 95.5% of cases in both cartilaginous and mesenchymal components making SOX9 a highly sensitive and specific marker.[15,16]

FISH analysis reveal HEY1-NCOA2 rearrangement,characteristic of mesenchymal chondrosarcoma. Studies highlight the importance of *HEY1-NCOA2* rearrangement in mesenchymal chondrosarcoma not only as a diagnostic marker but also for its potential in therapeutic advances. [18,19]

The mainstay of treatment includes aggressive surgery.[1,3]The addition of radiotherapy and chemotherapy shows variable results in different studies with some studies emphasizing on neo-adjuvant chemotherapy too. [1,2,3,8] This is proved in CWS/COSS study, where only two of seven patients demonstrated tumor reduction greater than 66% by adding chemotherapy.

Studies have tried to correlate histopathological parameters with survival outcomes but the results were not fruitful.[20] Although there is no necrosis and the mitotic rate in these tumors is generally very low(in contrast to that in our study), mesenchymal chondrosarcoma is classified as grade 3 according to FNCLCC and NCI grading systems because of the inherent nature of the disease (locally aggressive, late recurrence and widespread dissemination) and hence the prognosis is generally poor.[21] Yet, the 10 year survival rates in various studies range from 20% in 1980s to 60% in 2010. This could have been because of combined treatment modalities and aggressive approach followed these years in handling these tumors. [1]

4. Conclusion

We conclude saying mesenchymal chondrosarcomas is one of the rare tumors requiring aggressive treatment for overall disease free survival. Sampling defect can bring about diagnostic issues, especially with other pure mesenchymal lesions. Hence a combination of radiological, histopathological, immunohistochemical and molecular methods are required for the accurate diagnosis.

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