Unusual presentation of Chordoma in distal radius

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Abstract

Chordoma is rarely seen in appendicular skeleton. Here we describe a rare case of chordoma occuring in distal radius. Radiograph showed ill defined lytic distal radius, meta diaphyseal lesion. IHC marker for Chordoma Brachyury was done (TMH , Mumbai), which came diffuse strongly positive- confirming the pre op diagnosis of Extra Axial Chordoma. He was treated with Wide resection of the distal radius and one bone forearm wrist arthrodesis

Keywords: Chordoma, extraxial, radius

Introduction

Chordoma is a rare malignant bone tumor derived from notochord remnants mainly occurring in axial skeleton, namely the sacral region, followed by spine and base of skull , rarely described in appendicular skeleton [1]. Until now only 11 cases have been objectively confirmed by brachyury immunostaining diagnosed as Appendicular Chordoma [2-8]. In this case report, we highlight a case of distal radius Chordoma with IHC confirmed positive brachyury marker. Brachyury, is a nuclear transcription factor, which is a recently described specific immunohistochemical marker for diagnosing chordoma [8]. Chordomas are slow growing painless, locally aggressive mass . Here in, we have discussed the case report and literature review of similar cases. Despite its distinct histopathological feature , at times a chordoma mimics certain other malignant as well as benign tumour containing variable amount of myxochondroid stroma . We will discuss the various differentials, diagnostic dilemmas and management protocol for this patient

Case Report

40 year old male patient with complains of Pain and swelling of left distal forearm, progressively increasing for 3 months. In the past he had a history of fall, 1 year back and sprained left distal forearm treated with analgesics and he was back to daily routines by 10 days. He Presented with insidious onset Distal forearm swelling with pain and open biopsy done elsewhere. No similar complains elsewhere, no constitutional feature of fever, weight loss. X ray Forearm showed - ill defined lytic distal radius, meta diaphyseal lesion, eccentric , with periosteal reaction and narrow zone of transition with cortical breech , internal septations and soft tissue component. MRI Showed heterogenously enhancing Soft tissue component – (6.5x 4.4x 2.9 cm) along the distal radius (7 cm radius erosion and periosteal elevation) extending to both the compartment of forearm displacing the tendons outward. Medially abutting the ulna with no erosions. Lesion encasing the anterior neurovascular bundle however , radial and ulnar arteries appears free. He was being evaluated in our institution and Evaluated for Secondaries (Lung , GI , Prostate), Giant cell tumour, Myeloma, Brown Tumour which came as negative. Tissue diagnosis Reported "Physaliferous cells" in myxoid mucinous background - Chordoma . IHC marker for Chordoma Brachyury was done (TMH , Mumbai), which came diffuse strongly positive- confirming the pre op diagnosis of Extra Axial Chordoma . MRI screening of the whole spine were negative for chordoma. He was treated with Wide resection of the distal radius and one bone forearm wrist arthrodesis Grossly , the excised specimen was in form of multiple , grey white glistening sift tissue fragment with myxoid areas ,measuring 7 cm x 4cm x 2 cm . Histopathology examination confirmed diagnosis of Chordoma with surgical margins free – featuring Tumour cells composed of polygonal cells with moderate to abundant eosinophillic to vaculated "bubbly" cytoplasm (Physaliphorous cells) arranged in nodules with myxoid matrix . IHC – Diffusely positive for Pan cytokeratin , EMA (Epithelial membrane antigen) , S100 P.

Discussion

Extra axial Chordoma are rare tumours . Extra axial chordoma are also called chordoma peripheriсum . Very few documented cases of the Chordoma are documented in the world literature , precisely 11 in number [Table 1] with IHC confirmation and single largest study has been the Tirabosco et al [4], Rizzoli Orthopaedic institute , Italy featuring 8 cases , with mean age 42 years and tibia as the commonest site and the largest size documented is 11 cm with mean size 4.6 cm. However, none of the study in literature has to the best of our knowledge, documented a extra axial chordoma in Distal radius . Histopathologically, the differential diagnosis of an extra axial chordoma include malignant tumours , such

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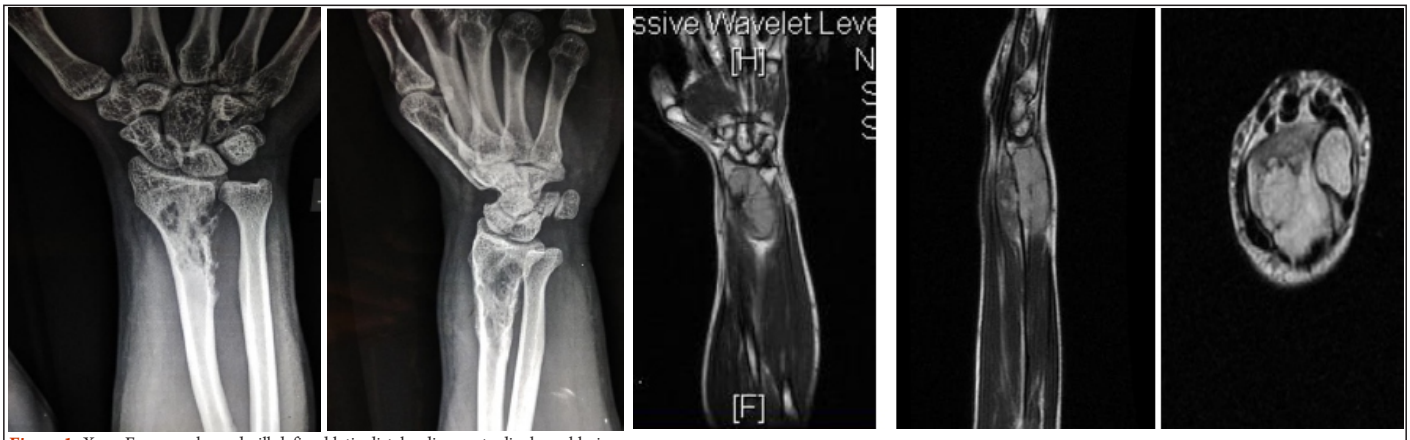

Figure 1: X ray Forearm showed - ill defined lytic distal radius, meta diaphyseal lesion, internal septations and soft tissue component

Figure 2: MRI Showed heterogenously enhancing Soft tissue component

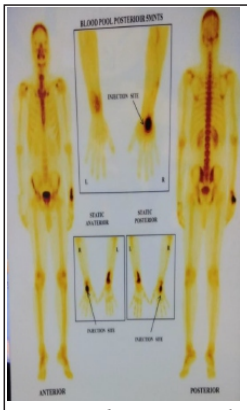

Figure 3: uptake primary site only

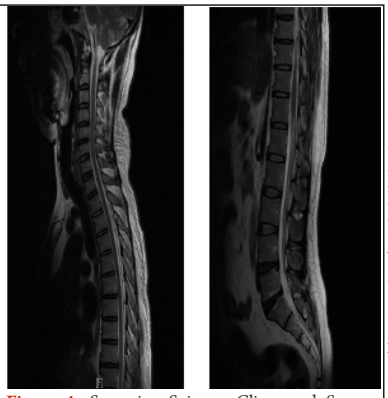

Figure 4: Screening Spine – Clivus and Sacrum Negative

as conventional and extraskeletal osteosarcoma, a myoepithelial carcinoma , chondroblastoma , chondromyxoid fibroma. Histopathological confirmation with IHC marker with Brachyury, Epithelial markers such as cytokeratin and S100 P are used to differentiate an extra axial chordoma from an extra skeletal chondrosarcoma , as these are expressed by the former tumour .Brachyury , a nuclear transcription factor involved in mesodermal differentiation- notochord , is a valuable marker as it is extremely sensitive and specific for differentiating Chordomas from myoepithelial /mixed tumors of soft tissues. An exact diagnosis has the most important therapeutic relevance. All the documented literature shows most extra axial chordoma can be treated with Wide

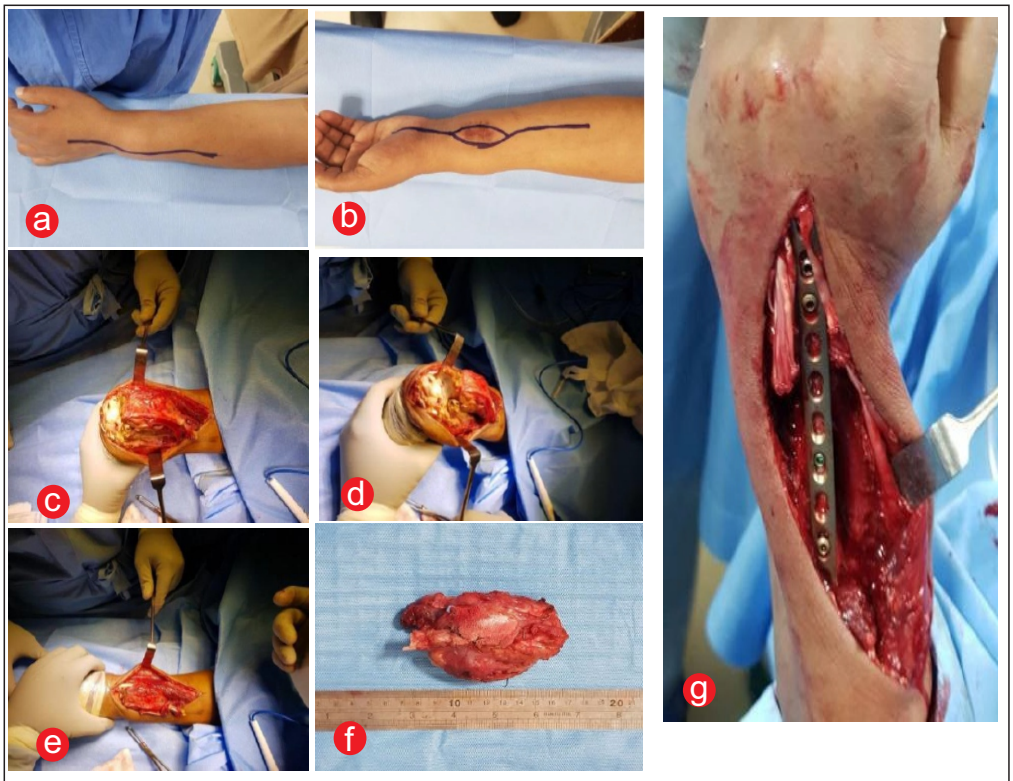

Figure 5: a,b - Planning the incision with dorsal,volar approach for R0 resection and incorporating previous biopsy scar. c,d-Wrist capsule exposed for distal margin. e,f - Resection of 10 cm bone lesion with 3 cm medullary margin distally and proximally margin was articular surface of distal radius. g- Reconstruction of the hand with one bone forearm Wrist arthrodesis and tendon repair .

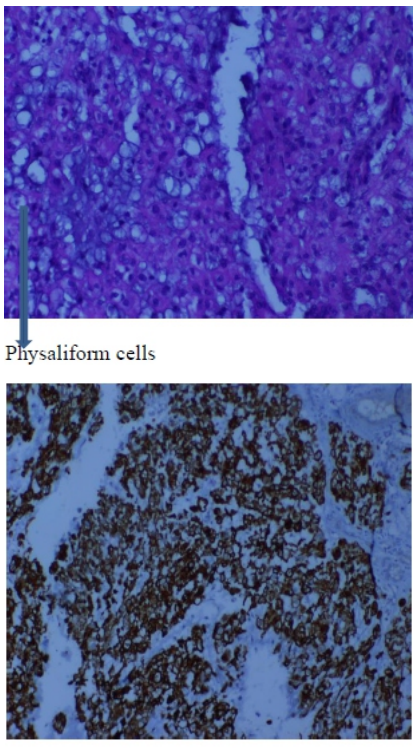

Figure 6: H&E 200X densly packed epitheliod cells with in been tumor cells showing vacuolated bubbly cytoplasm (physaliphorous cells)

1	Nielsen	36/male	Distal Ulna	8 cm	CK +	EMA +	S 100 +
2	Donell Et al	21/male	Poximal tibia	Not Menti oned	CK +	Not mentioned	+
3	Tribass co et al	36/m 56/f 41/f 27/m 35/m 55/m 68/male 18/female	Distal ulna 10th rib Pubis Prox tibia Mid tibia Metatarsal Dist. Femur Prox Tibia	NK NK NK NK NK NK 11cm NK	CK+ CK+ CK+ CK+ CK+ CK+ CK+ CK+	NOT DONE FOR ANY SAMPLE	+ + + + + + + +
4 Present Study	Subin Et al	42/male	Distal radius	+	+	+	+

Table 1: Study characteristics of important similar cases reported in Literature

Surgical Resection, as those treated with curettage are known to recur. The classical teaching for chordoma is the embryological remnant mesodermal tissue occurring along the vertebral column especially at Sacrum , skull base but extra axial Chordoma is something which challenges this notion , as some extra axial chordoma even occurring at distal appendicular skeletal systems . Identification and confirmation of this rare tumour has a bearing on the treatment protocol as wide resection and reconstruction can be the best treatment option . In our case , patient underwent wide resection and reconstruction with one bone forearm wrist arthrodesis . At the end of 1 year of follow up , he is doing well with no recurrence and is back to his daily routine activities .

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