

Case Report

Extraskelatal Soft Tissue Chondromas of Head and Neck Region

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Abstract: Chondromas are benign cartilage producing tumors which are composed of mature hyaline cartilage. We report two cases of extra skeletal soft tissue chondromas. Our first case was a 30 year old male who presented with swelling on the right pinna, while the other case was a 17 years old female with a nodule on the right cheek. The right pinna growth was surgically removed and the specimen was sent to Armed Forces Institute of Pathology (A.F.I.P) for anatomical diagnosis. The right cheek nodule was removed and the specimen was sent to a local laboratory and the histopathological diagnosis of non-keratinizing squamous cell carcinoma was made. Blocks of this case were sent to A.F.I.P for review. On the basis of morphological features both the cases were diagnosed as extra skeletal soft tissue chondromas.

Key Words: Soft tissue, extra skeletal chondromas, head & neck

Introduction

In 1883, Baumuller first reported an ossifying chondroma of skin on the dorsum of foot.¹ Soft tissue chondromas are benign, extra-skeletal and extra-synovial, mesenchymal tumors composed almost exclusively of mature hyaline cartilage. Cartilaginous tumours occurring at extra skeletal sites are rare lesions. Three types of extra osseous chondromas are intra-articular chondromas, juxtacortical chondromas and chondromas of soft parts. Fewer than 200 cases have been described in the literature. No local data is available on these tumors.

Most of the cases occur in the distal phalanges, the toes and fingers. Only few cases occurring in the head and neck area have been described. The age of occurrence so far reported spans from 30 years to 60 years. Clinically, they present as a slow growing mass, occasionally painful and tender. They are usually less than 3 cm in size, and on palpation, firm or rubbery, well defined, and often mobile. Histologically majority of the cases are composed entirely of lobules of mature hyaline cartilage.

The histological differential diagnosis includes benign tumors like ganglion cyst, myositis ossificans, synovial chondromatosis, osteochondroma and malignant tumors like chondrosarcoma, extra skeletal osteosarcoma and synovial sarcoma. Local excision is curative in almost all of the cases.

Radiologically they appear as ring like, cotton-ball or "popcorn" calcifications of various sizes. A bone scan may show increased uptake while MRI shows increased signal intensity on T-2 weighted images. The treatment relies mainly on surgical excision with a small amount of normal tissue around the tumor, as recurrence rate of 18 % has been reported in literature. This case series aims to highlight the importance of this uncommonly diagnosed entity.

Case

Case 1: First case was a 30 years old male who presented with 20 days history of swelling and pain in left pinna. On physical examination there was a 2 cm, firm and tender mass in upper part of left pinna. The overlying skin was erythematous. The swelling was surgically excised and the specimen was sent to Armed forces Institute of Pathology (A.F.I.P) Rawalpindi for histopathological examination. On gross examination, the specimen consisted of multiple dark brown, firm pieces of tissue collectively measuring 2 cm. The specimen was embedded as such in one block. Histologically the sections revealed mature hyaline cartilage composed of chondroblasts and with variable sized chondrocytic lacunae. Few fragments showed epithelioid cells and foreign body type multinucleated giant cells surrounded by mixed inflammatory infiltrate. No evidence of malignancy was seen in the material examined. **Figure 1**

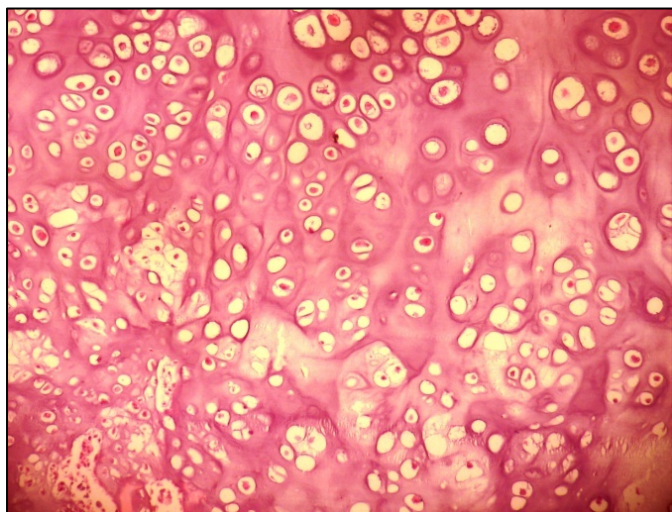


Figure I: Medium power photomicrograph (original magnification, 200; H-E stain) showing mature hyaline cartilage with chondrocytic lacunae.

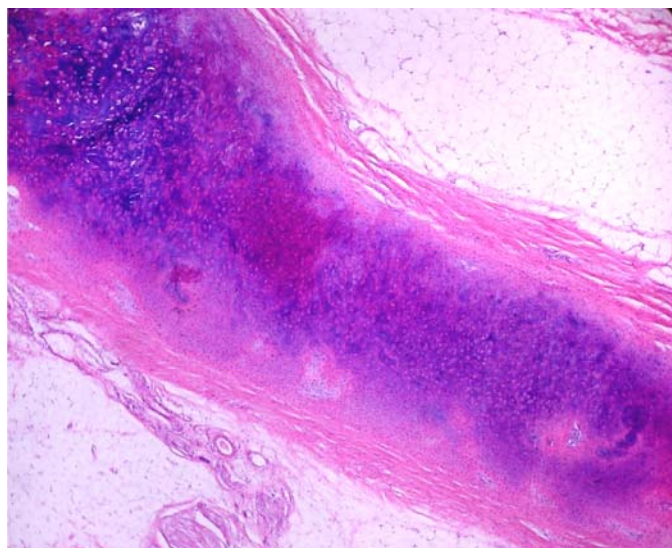


Figure II: Photomicrograph from the case 2 showing mature hyaline cartilage surrounded by adjacent fibro adipose tissue.

Case 2: A 17 years old female presented with a mass arising from right cheek. On clinical examination the mass was lobulated and solid. It measured 2.5 x 1.5 cm. Rest of the clinical examination was unremarkable. The lesion was excised and sent to a peripheral laboratory for histopathological examination. It was reported as non keratinizing squamous cell carcinoma. The treating surgeon requested reviewing of the blocks

from A.F.I.P. Microscopic examination revealed skin with underlying tissue showing a well defined mass comprising of confluent lobules of hyaline cartilage composed of chondroblasts with variable degree of cellularity. Calcification was not present. The lesion was reported as extra skeletal (soft tissue) chondroma. The excision was curative in this case. **Figure II**

Discussion

Soft tissue chondromas are benign tumours of cartilaginous tissue that may arise adjacent to normal bony or cartilaginous structures. These benign tumors can be seen in any age group but are commonly seen in patients between 30 to 60 years of age. There is slight male preponderance (1.2:1).² Soft tissues chondromas arise principally in extremities (96%) with 72% in the upper limb, 24% in the lower limb, 2% in the head and neck and 2% in the trunk.³ Rare sites such as fallopian tubes, parotid gland have been reported. One case of soft tissue chondroma of the cheek like our case has been reported in the literature.⁴ There are other terms used to describe these benign solitary cartilaginous masses such as: extra skeletal or soft tissue chondroma, extra osseous chondroma, tenosynovial chondroma, chondroma of soft parts, and cartilaginous tumor of the soft tissue. Lesions showing evidence of ossification are called Extra skeletal osteochondromas.

The tumor is typically associated with a tendon, tendon sheath or joint capsule. There are several theories explaining their origin: Sood et al concluded that they arise from metaplastic change from adipose tissue. Dahlin and Salvador suggested a synovial origin.⁵ While Uehara,⁶ Rosenfeld, Kurzer and Becker postulated that they arise from the activation of heterotopic cartilaginous tissue. Recently, nonrandom clonal alterations of chromosomes 6, 11, and 12 have been reported in soft tissue chondromas. In another study molecular analysis showed that a gene called HMGA2 located at 12q15 appears to be involved.⁷

Patients usually present with a solitary slowly growing soft-tissue mass which may or may not be associated with pain or tenderness. In one of our case the swelling was tender on palpation. Lesions are typically well circumscribed and lobulated and rarely exceed more than 2 cm in size. In both of our cases the swellings were less than 5 cm in greatest diameter. The radiographic findings of extra skeletal chondromas typically consist of well-circumscribed, extra skeletal lobulated soft tissue mass with dense, central calcifications or areas of ossification.⁸ On MRI (Magnetic Resonance Imaging) they show high signal intensity on T2-weighted images and intermediate signal intensity on T1-weighted images. Due to the peculiar site of both of our cases radiological investigations were of little value.

Differential diagnosis for a discrete soft tissue mass containing mature ossification includes myositis ossificans, extraskelatal chondroma with enchondral ossification, synovial osteochondromatosis, tumoral calcinosis, synovial sarcoma, and extraskelatal osteosarcoma. In our cases the clinical differential diagnosis included fibroepithelial polyp and myositis ossificans.

Fine needle aspiration cytology (FNAC) of soft tissue chondromas is rarely practiced; only one case diagnosed on FNAC has been reported in the literature. FNAC of a typical soft tissue chondroma reveals chondrocytes present in a cartilaginous matrix.⁹ On histopathological examination these tumors grossly appear as well circumscribed, often encapsulated and have a shiny, myxoid or calcified cut surface. On light microscopy most tumors are composed of mature hyaline cartilage, often showing foci of dystrophic calcification or metaplastic ossification.¹⁰ Cellularity of the tumor is quite variable and chondrocytic lacunae also tend to vary in size. In some cases marked nuclear atypia, pleomorphism, binucleated or multinucleated lacunae are present but no malignant transformation has been reported. Dystrophic calcification, metaplastic ossification and cytological atypia were not seen in our cases. The differential diagnosis includes well-differentiated extra skeletal chondrosarcoma, periosteal chondroma, extra skeletal myxoid chondrosarcoma, and mesenchymal chondrosarcoma. The differential diagnosis in both of our cases included chondroma of bone and chondrosarcoma. They were excluded due to lack of bony component and cytological features of malignancy. The differential diagnosis in case 2 also included benign mixed tumour of salivary gland, which was excluded owing to absence of any epithelial component.

The usual treatment for extra skeletal chondroma is surgical excision with wide margins. Few cases of recurrent chondromas have been reported in the international literature. This case series aims to highlight the importance of diagnosing soft tissue

chondromas as they are benign conditions but may exhibit increased cellularity and mitoses which should not be mistaken for a malignant condition. Surgical excision in both of our cases was curative and there was no recurrence after 6 months of follow-up.

Conclusion

Extra skeletal chondromas are rare soft tissue benign tumors that should be considered in the differential of a soft tissue mass. Histopathological evaluation is required for the final diagnosis.

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