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## RESEARCH ARTICLE

# MULTIPLE HEREDITARY EXOSTOSES: A CASE REPORT

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**ABSTRACT**

17 yr old male having multiple hereditary exostos is was operated for large size tumour in left Calf 2yr back as it was restricting his knee joint mobility. Recently he was operated for massive growth around right hip and right scapula. Biopsy showed benign tumour. No complications regarding surgery and recurrence.



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## INTRODUCTION

Osteochondroma is a outergrown growth plate near metaphysis of bone. Its growth increases with the growth of the child and ceases with the same. Osteochondroma can develop as single tumour or multiple tumours. It's a cartilage forming tumour which has bone also.These are stalked or sessile in nature.Multiple Osteochondroma are also known as multiple exostosis,multiple hereditary exostosis,familial Osteochondromatosis and Diaphyseal achalasia also. Common sites for occurring in body are around knee, hip and shoulder.Flat bones are also involved in multiple osteochondromas. These osteochondromas are usually bilateral, symmetrical, and directed away from the nearest joint. They are most common, numerous, and largest at the knee (70%), followed by the humerus (50%) and fibula (30%).skull is not involved. Males has preponderance than females in frequency of occurance. Common age is 10 -30 years. Osteochondromas are most commonly diagnosed incidentally on radiographs.The second most common presentation is a mass, which may or may not be associated with pain. Pain is usually caused by a direct mechanical mass effect of the osteochondroma on the overlying soft tissue. Irritation of surrounding tendons, muscles, or nerves can result in pain. Pain can also result from fracture of the stalk of the osteochondroma from direct trauma.

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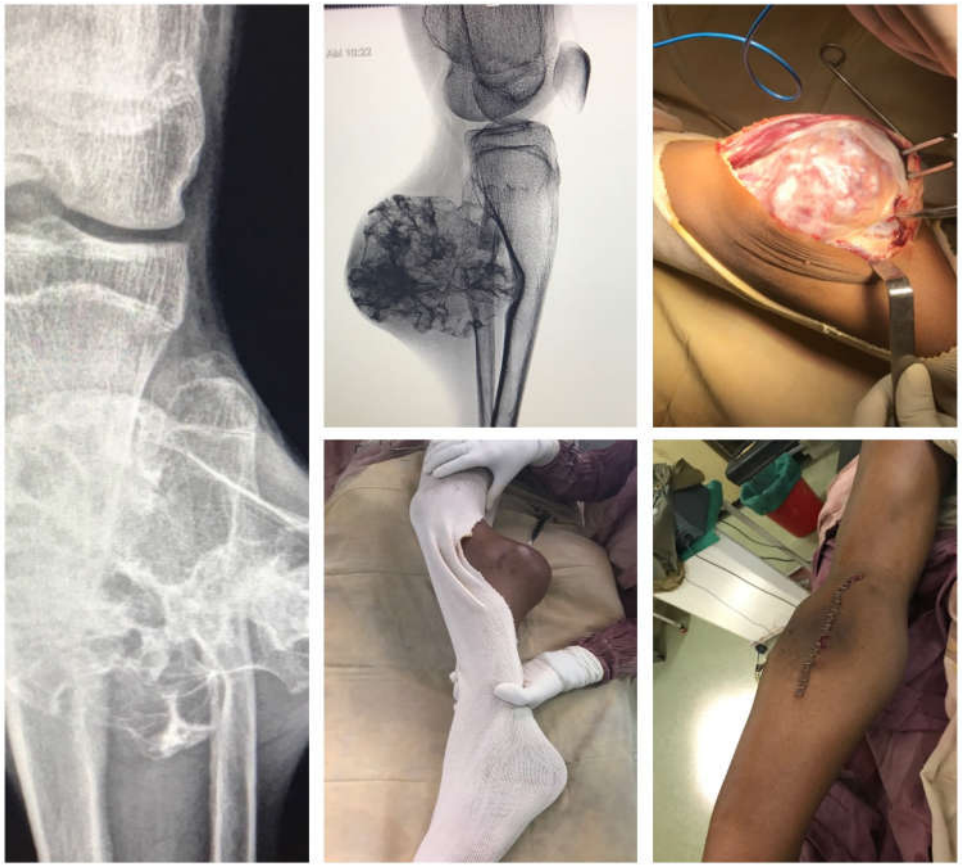
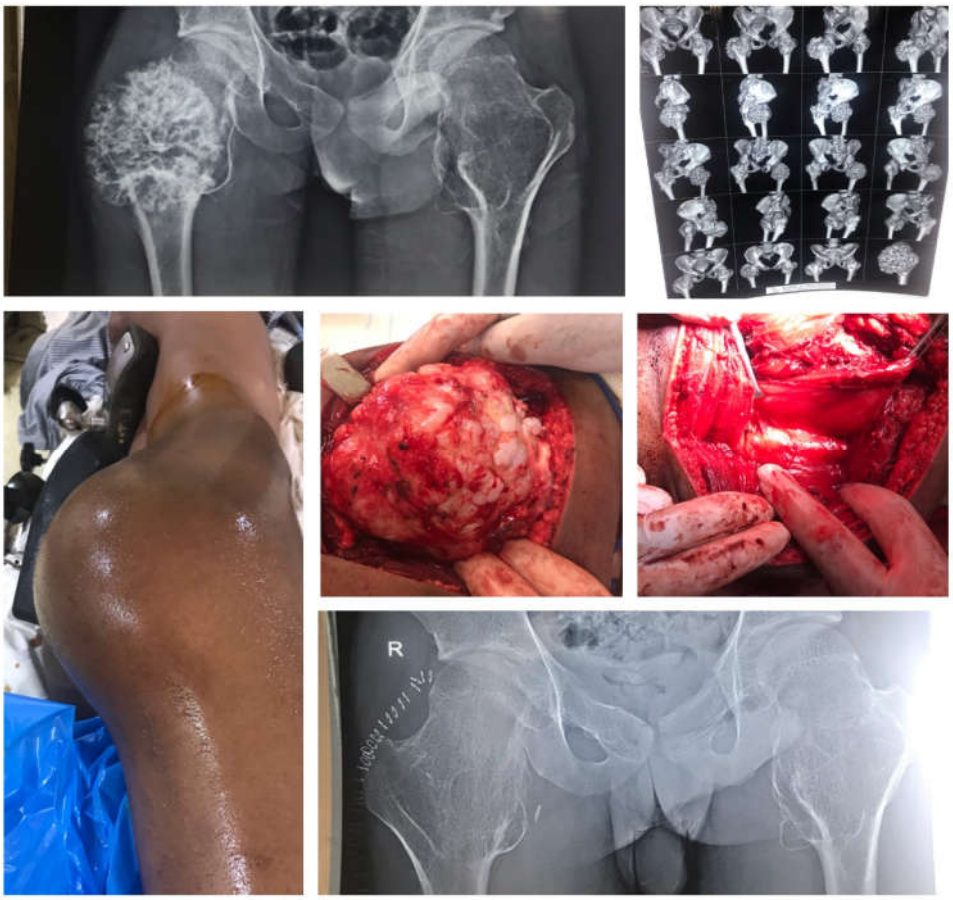
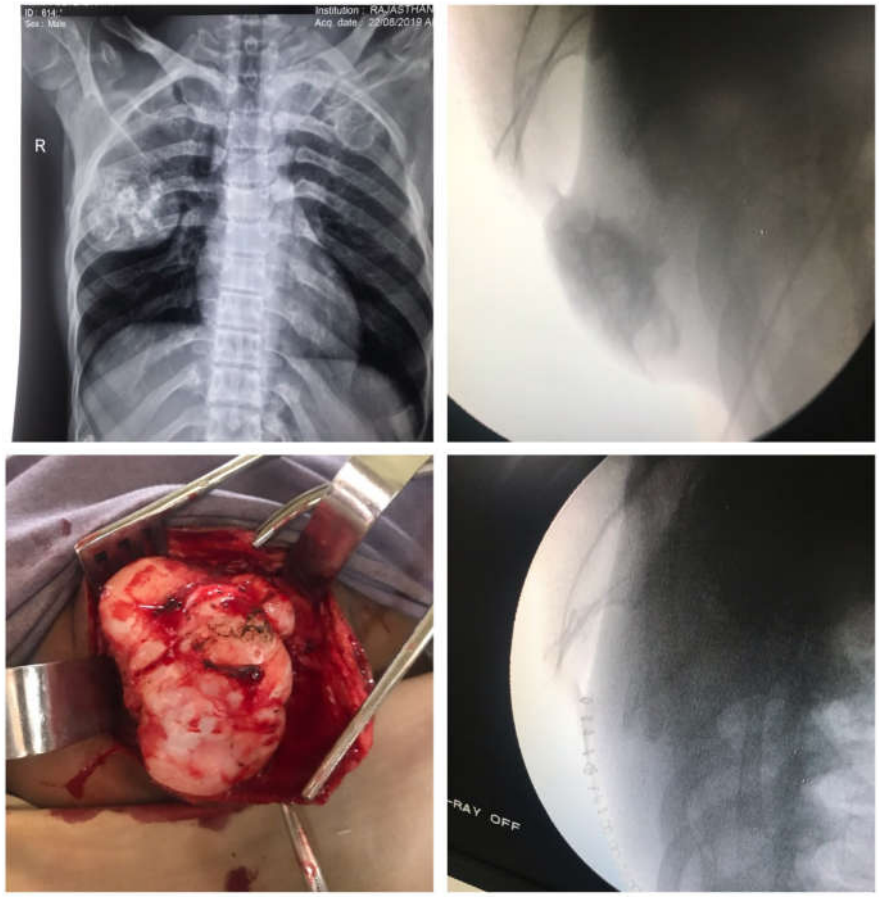
Osteochondroma may be associated with pseudoaneurysm of an extremity artery (eg, the popliteal artery), commonly evidenced by pain and swelling. Osteochondromas of the spine, though rare, can give rise to symptoms of compressive myelopathy. Asymmetric growth of paired long bones because of the presence of osteochondromas can contribute to the angular deformation leading to genu valgum, ankle tilt into valgus, dislocation of the radial head at the elbow, and wrist deformation with a dorsally displaced and shortened u lna.Radiologically tumours are easily diagnosed in X-ray.Either sessile or pedunculated.Metaphysis is broadened .CT SCAN is helpful in surgical planning and MRI is required to differentiate between the benign and malignant transformation.cartilaginous cap more than 2 cm is indicative of sarcomatous change.Biopsy confirms then. Multiple exostoses are generally autosomal dominant inheritance disorder or in 70 percent cases but 30 percent has some mutation of EXT1,EXT2,EXT3 genes affecting pre hypertrophic chondrocyte of growth plate. Indian hedgehog protein synthesis regulation mechanism's defect is one of causes identified in recent times. Solitary lesions transformation to malignancy is less than 1%but 5-10%in multiple varieties .Rare in Paediatrics but common in aged one.Pelvic lesions are more prone to malignant transformation. Histologically it resembles to normal physis.cartilage cap is surrounded by perichondrium layer. cartilage cap is formed by hyaline cartilage. chondrocytes are arranged in linear clusters. Normal primary trabecular patterns are there.



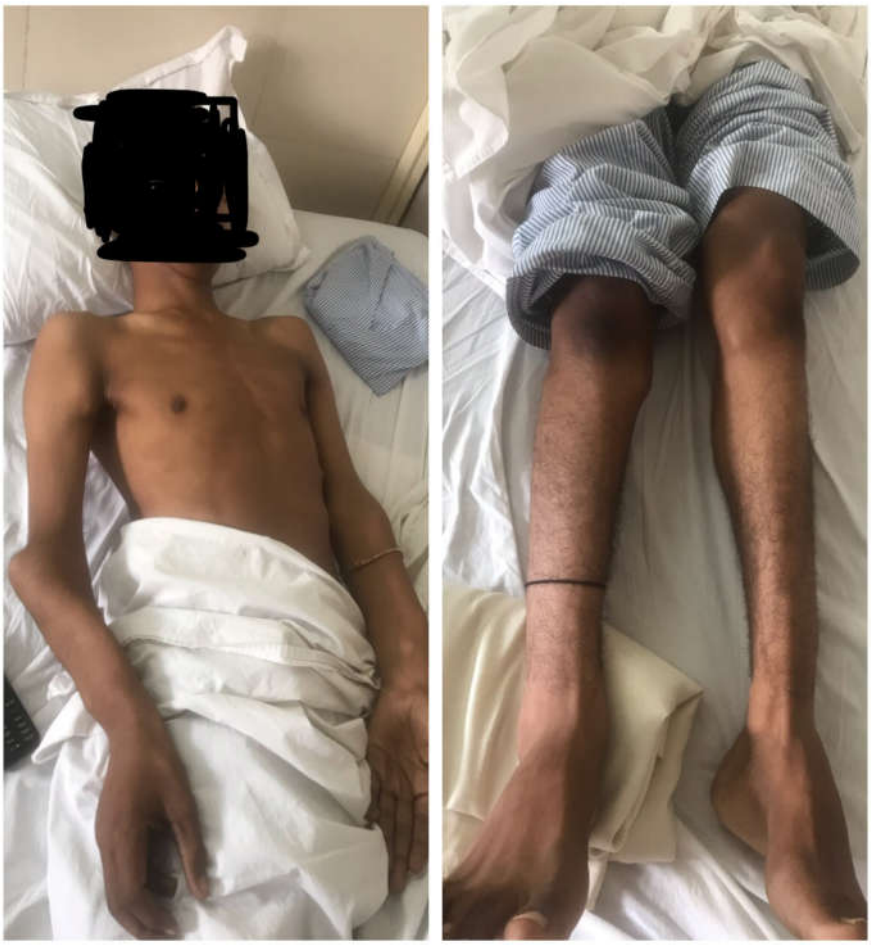
**Huge mass at Right knee posterior fibula .preop and post op X-ray.intraop pictures**




**Massive tumour around Right hip posterior aspect Preop and post op X-ray.intraop picture**




**Large tumour Rt scapula dorsal region .preop and post op X-ray.intraop picture**



**Clinical appearance of achondroplasia and growth disturbances**

Marrow elements may be present within bony stalk; marrow space / cancellous bone contiguous with that of the native bone.

## MATERIALS AND METHODS

17 years old boy presented with multiple huge swellings on all four limbs and flat bones. He had restricted left knee flexion because of massive swelling in calf. Patient had huge swelling in right hip region and right scapula also. Other findings includes dislocated right radial head and swelling on right thigh in lower thigh and reduced height and length of long bones. Patient operated for right calf swelling which was originating from upper fibula. Radiological appearance seemed to be Osteochondroma. Patient was operated in prone position and through posterior midline incision. Stretched and displaced neurological and vascular structures were dissected and tumour was excised completely and sent to histology.Report confirmed benign tumour Osteochondroma .Patient showed smooth recovery and no recurrence for 2 years.Now after two years of this surgery Patient had some trauma around right hip due to fall. So patient developed some pain and difficulty in weight bearing on right hip. Patient underwent for X-ray and The 3D CT scan and MRI SCAN suggesting non malignant transformation of mass. In lateral position the right hip area swelling was excised by protecting the sciatic nerve through posterior approach. Tumour was originating from posterior aspect of base of greater trochanter displacing the sciatic nerve and short external rotators. Intraop and histology showed benign nature of mass. After 3days of this surgery the right scapular swelling was excised in right lateral position. Histology showed the benign tumour.

## DISCUSSION

Multiple exostoses are hereditary cartilage producing tumour benign in nature may transform in malignant variety these are mainly around large joints provide mechanical difficulties in joint motions rarely Neuro vascular complications also takes place some other complications include bursitis ,tendinitis and fracture. These grows with normal bony skeletal system. Growth cessation takes place after normal growth is over.

Sudden rise in growth in adult or painful growth is indicative of further assessment to rule out malignant transformation differential diagnosis includes Dysplasia Epiphysealis Hemimelica, Turret Exostosis and Other Osteochondromatous Lesions.

**Conclusion**

Multiple exostosis are hereditary benign cartilage forming bony tumour arising near larger joints should be removed if they are obstructing the joint function or creating angular deformities or growth disturbance or any Neuro vascular complications should be removed. Malignant transformation should be ruled out by MRI scan and tissue biopsy if sudden rise in pain and size.

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