

# Haemangioendothelioma of olecranon process- Limb salvage surgery with custom total elbow prosthesis

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## Citation

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## Abstract

Haemangioendothelioma of the bone is a very rare vascular tumour which accounts for less than 1% of the malignant bone tumours. Here we present this uncommon tumour arising from the olecranon of the right ulna and discuss on the complexities and challenges of elbow resections and its reconstructions

## INTRODUCTION

Bone tumours arising from the elbow region is a challenging situation. We report a case of haemangioendothelioma, a rare malignant vascular tumour, arising in the olecranon of the ulna, which was treated with total elbow resection and reconstructed with custom made total elbow prosthesis (Waldemar Link, Hamburg, Germany). A brief review of the literature regarding haemangioendothelioma of the bone and the treatment strategies for elbow tumours with respect to the challenges involved in the reconstruction is reviewed and discussed herein.

## CASE REPORT

A 51 year old gentleman working in Middle East sustained a pathological fracture of the right olecranon. He underwent open reduction and internal fixation with curettage and tension band wiring apart from biopsy of the fractured site(Figure 1).

## Figure 1

Figure 1: Tension band wiring of pathological fracture

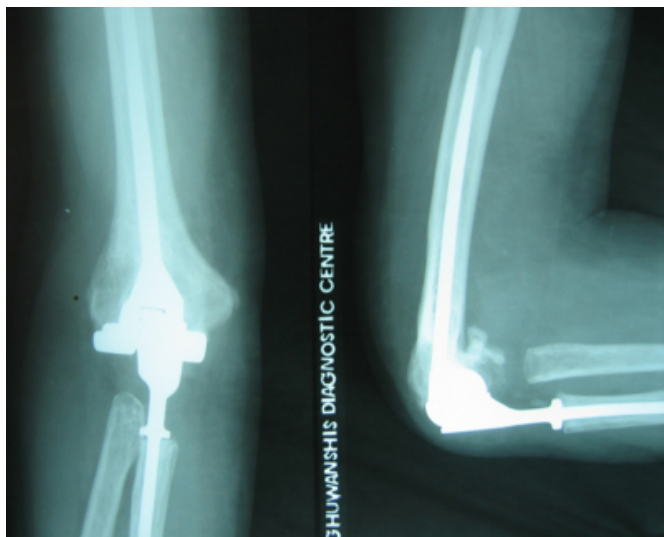


The histopathology was suggestive of epitheloid haemangioendothelioma, grade II, which was confirmed with immuno-histochemistry studies( CD 34 and smooth muscle actin were positive). He was subsequently referred to

our centre for further management. Metastatic workup included computed tomographic scan (CT scan) of the chest and bone scan which were negative. The axillary lymph node biopsy for an ipsilateral enlarged axillary node also did not reveal any metastatic disease. Based on the X- ray scannogram customized titanium prosthesis was imported from Germany (Waldemar Link, Hamburg). He underwent total elbow resection and replacement with distal humerus proximal ulna customized prosthesis (Figure 2). Extensor (triceps) of the elbow was sutured to the prosthesis and the soft tissue in the forearm. The skin was closed after putting subcutaneous vicryl sutures. He had an uneventful postoperative period, and was started on supervised graded elbow exercises, and had a mean Mayo Elbow Performance (MEP) score of 70 (max – 100 points) at the time of discharge. At one year follow up he has excellent MEP score of 90 points with an elbow extension of 70 degrees.

### Figure 2

Figure 2: Custom made prosthesis in place



### DISCUSSION

Epithelioid haemangioendothelioma of the bone is a very rare malignant vascular tumour which shows intermediate biological appearance between haemangioma and conventional angiosarcoma. They account for less than 1% of malignant bone tumours. They are composed of tumour cells which show endothelial differentiation (1). It has been reported in the liver, lung, gastrointestinal tract, head and neck, heart, central nervous system and bone. Lesions of the liver and lung are more common in females, whereas tumours of the bone and soft tissues have an equal sex distribution. They have a wide skeletal distribution commonly involving the skull, vertebra and the long bones

(2, 3). They show multicentricity in 30% of cases. They tend to occur during the second and third decades of life. On microscopy epithelioid haemangioendothelioma is composed of anastomosing cords, solid nests, and strands of endothelial cells that may sometimes form narrow vascular channels. The endothelial component uniformly express vimentin and many cells stain with antibodies to Factor VIII, CD31, CD34, and Ulex Europaeus and the epithelioid type may also express cytokeratins and EMA (1). The histological degree of differentiation is the most significant factor in the prognosis of patients with these types of tumours. The radiological findings are non specific. But these lesions are predominantly osteolytic. They may be solitary or multifocal and /or polyostotic (4). These tumours characteristically lack periosteal reaction. Surgical resection is the mainstay of treatment, and there is no definite role for adjuvant therapy.

With the advent of the custom made elbow prosthesis, limb salvage surgeries for bone tumours of the elbow region have become feasible. Total elbow arthroplasty is a challenging procedure especially when performed for malignant bone tumours. Soft tissue cover for the prosthesis a major challenge. These prostheses due to its sub cutaneous location are prone to infectious complications. Extensor reconstruction is another technical challenge. These prostheses may be rigid hinged, semi constrained or non constrained types. Elbow joint allograft (EJA) or the composite allograft-implant reconstruction involving the entire joint (distal humerus, proximal radius and ulna, capsuloligament structures) is also a technique proposed, especially in young subjects when total elbow prosthesis is contraindicated and in salvage of failed total elbow arthroplasty.

### CONCLUSION

Haemangioendothelioma of the bone is a very rare malignant vascular tumour which shows intermediate biological behaviour. Primary bone tumours of the elbow region should be considered for total elbow arthroplasty when feasible as it provides a good functional elbow joint.

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