Giant Chondrosarcoma of the Clavicle - A Case Report and Literature Review.

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ABSTRACT

The most common tumours of the clavicle are Myeloma and Ewing's sarcomas while chondrosarcomas being a relative rarity at this location. However, a clavicular chondrosarcoma is considered favourable compared to the traditional more challenging locations like pelvis, femur and humerus. The subcutaneous location of the clavicle facilitates early detection. A further favourable consideration is that a complete claviculectomy without incurring significant morbidity can be performed. We reported an adult male patient who presented to our unit with a massive clavicular chondrosarcoma with radiological spread to the retro-pectoral lymph nodes. A total claviculectomy was performed with en bloc removal in the first stage followed by local lymph node clearance and adjuvant chemotherapy and radiotherapy. At one year follow up no recurrence was noted and the patient had good functions of the shoulder. We recommend total claviculectomy as the procedure of choice for clavicular chondrosarcomas.

Keywords: Chondrosarcoma, Clavicle, Claviculectomy, Tumour.

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INTRODUCTION

Although clavicle tumours are rare, an enlarging clavicular mass should be regarded as a primary malignancy until proven otherwise.[1] Chondrosarcomas have a predilection for males over the age of 40 years and occur most frequently in the pelvis, femur, and humerus[2], their occurrence in the scapula, and even less so in the clavicle is a rarity.[3] More frequently occurring tumours of the clavicle described in one series of 12 patients were Ewing's sarcoma and multiple myeloma.[4] Another study considered 23 dedifferentiated chondrosarcomas and confirmed that only one tumour was originated from clavicle.[5] Besides several distinct osseous peculiarities of the clavicle like its horizontal orientation and the first bone in the foetus to ossify, the most relevant defining feature however is the fact that it can be resected entirely without causing any significant disability.[6] This final point is taken an advantage in clavicular chondrosarcomas because due to poor response to both chemotherapy and radiotherapy a total claviculectomy is the most common definitive surgical procedure performed.[3,7] We presented a case report of a patient with giant chondrosarcoma of the clavicle which was treated successfully with total clavicle excision.

CASE REPORT

A 57 years old right hand dominant, manual labourer presented to Orthopaedic unit Helen Joseph Hospital University of the Witwatersrand, Johannesburg, South Africa with a progressively enlarging, painless right clavicular mass of one year duration. The patient was a known hypertensive and chronic

smoker. On systemic enquiry he confirmed associated loss of weight for the preceding year. On general examination he appeared healthy and without lymphadenopathy. Examination of the right clavicle revealed a large 15cm by 10cm firm mass with its long axis orientated along the long axis of the bone. The mass was firm, non-tender, fixed to the underlying tissue, had a discreet edge and there were no overlying skin changes (Fig 1A, IB). Neurological examination of his right upper limb was normal. Vascular examination revealed normal distal pulses and no signs of venous engorgement.

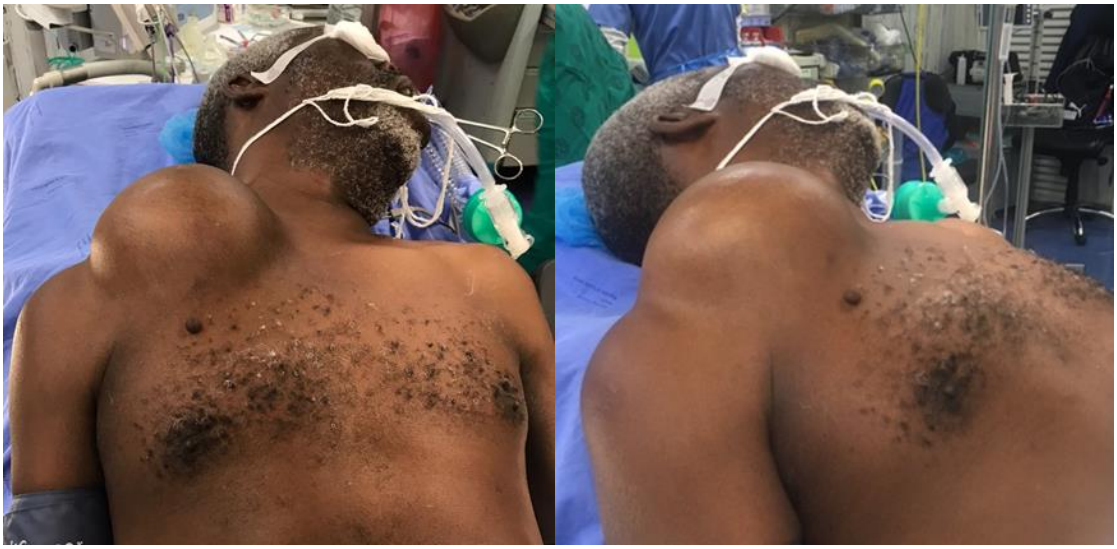

Figure IA, IB: Pre-operative patient photograph showing the large painless mass centred over the right clavicle. The alarming size of the mass is appreciable with lack of overlying skin changes.

After haematological investigations we proceeded with a chest X-ray which although excluding any obvious lung metastases confirmed that the origin was from right clavicle and had been completely replaced by tumour (Fig. II).

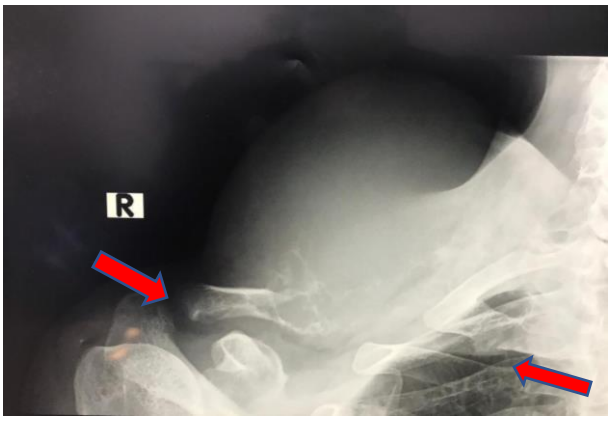

Figure II: Pre-operative antero-posterior right shoulder X-ray confirming the right clavicle which could not be visualized except for osteolytic and expanded tumour mass with medial and lateral clavicle remnants (red arrows).

A CT chest was performed which confirmed that 15cm by 10cm soft tissue mass replacing the right clavicle. Unfortunately the right retro-pectoral lymphadenopathy was also noted (Fig. III) but lung fields were clear of metastatic disease. A subsequent abdominopelvic CT scan was performed and excluded gross involvement.

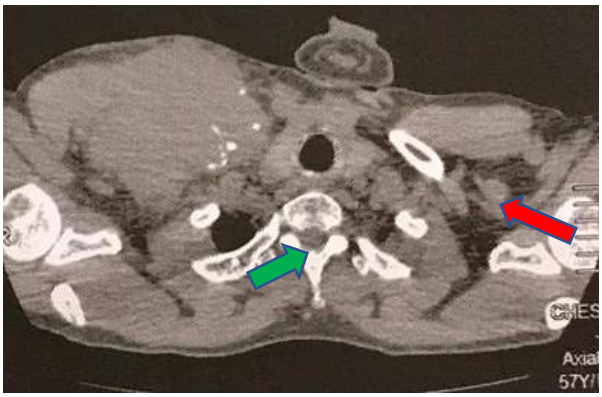

Figure III: Pre-operative CT chest showing the iso-dense soft tissue mass completely replacing the right clavicle. Only a few scattered hyperdense (osseous) remnants of what was the right clavicle can be seen contained within the tumor substance on its medial aspect (red arrow). The hypodense border surrounding the tumor eluded to a non-infiltrative edge which predicted a favourable plane facilitating excision and was found intra-operatively. In this view an enlarged retro-pectoral lymph node is visualized (green arrow).

An MRI of his right shoulder was subsequently performed which confirmed the hyperintense lesion to be easily discernible from the surrounding tissues. The well-circumscribed nature of the mass again eluded to a probable plane of excision that would be found intra-operatively.(Fig. IV) Due to the absence of any vascular involvement clinically, a CT angiogram was not performed. However due to the obvious proximity of the lesion to major vascular structures we acknowledged that CT angiogram should have been performed by an orthopaedic surgeons who come to face with a tumour of this size in this location. Technetium bone scan revealed single retro-pectoral lymph nodes in the axilla.

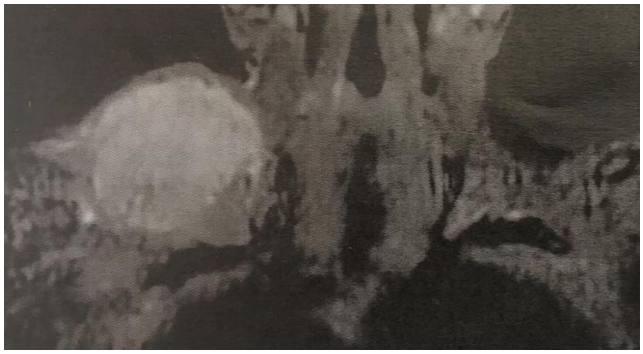

Figure IV: Pre-operative MRI T2W coronal image of the right shoulder showing the hyper-intense tumour. The mass is heterogenous in appearance suggestive of areas of central necrosis. The lesion is furthermore well-circumscribed and x clearly distinguishable by its hyperintensity from the surrounding tissues.

A fine needle biopsy of the lesion confirmed the tumour to be a chondrosarcoma and the decision was taken that the best way forward would be to perform a total claviculectomy with subsequent referral to oncologest for adjuvant therapy and general surgeon for lymph node clearance.

Operative procedure

The patient was taken to the operating room and placed in supine position with the operating table adjusted to the beach chair position. His head was rotated to the left side away from the right clavicle. The acromioclavicular and sternoclavicular ends of the clavicular remnants were marked on the skin with a cutaneous marker and joined with a planned incision line that passed over the maximal convexity of the tumor (Fig. V)

The skin incision was made and the skin was sharply dissected superiorly and inferiorly off from the capsule of the tumour to which it was unattached. The dissection was continued over and then posterior to the superior pole of the tumour. The same procedure was performed inferiorly over and then under the inferior pole of the tumour. The remaining attachment directly beneath the tumour from the lateral to the medial end of the clavicle along its length was left intact at this point for fear of involvement of the subclavian vein (Fig. VI).

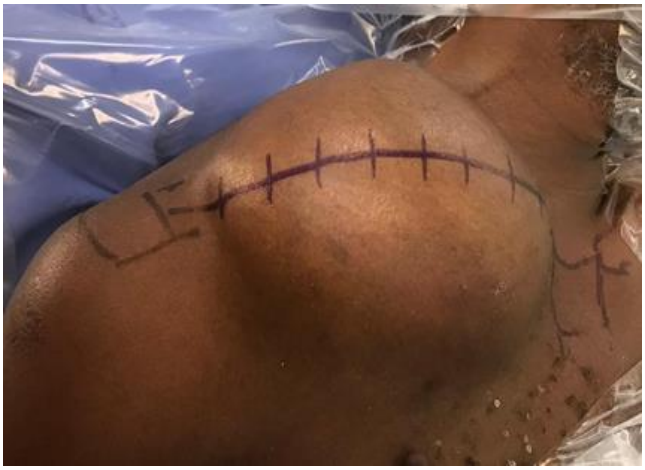

Figure V: Intra-operative photograph of the medial and lateral ends of the clavicle marked and joined by a line that passes over the maximal convexity of the tumor.

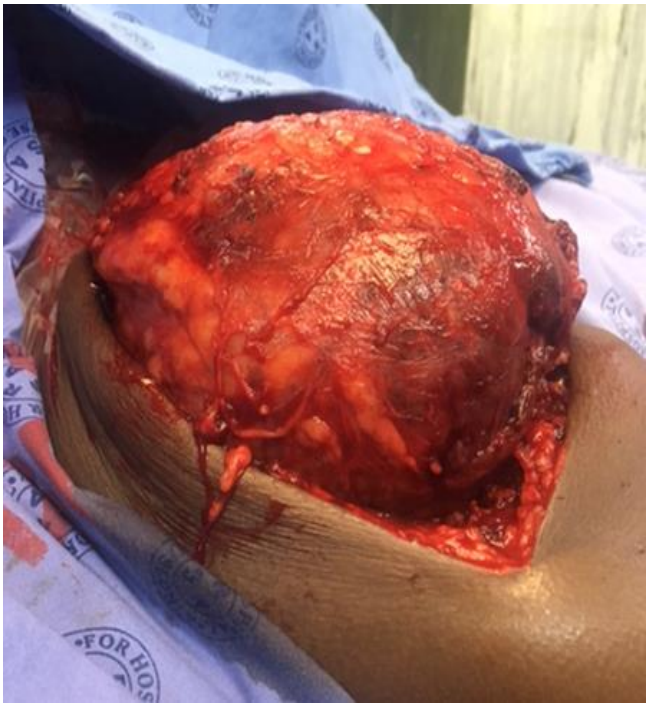

Figure VI: Lateral intra-operative photograph of the exposed tumour. The skin has been reflected from its superior and inferior poles which have been undermined. The tumour remains attached laterally at the acromioclavicular joint along the length of the clavicle and medially at the sternoclavicular joint.

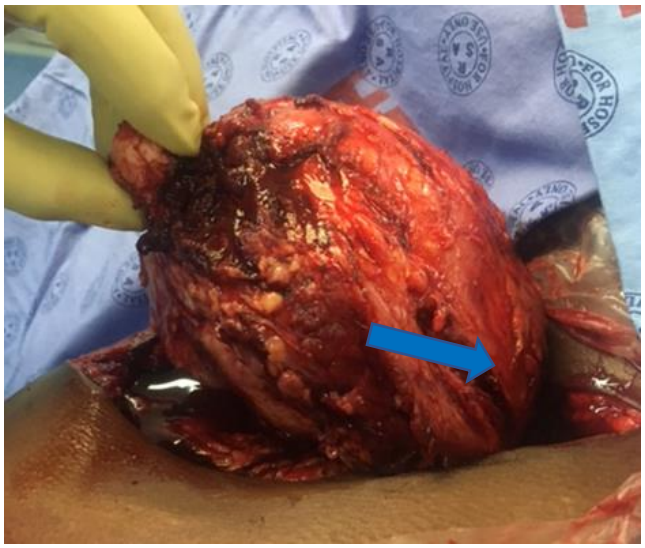

Figure VII: Intra-operative photograph of the surgeon holding up the lateral end of the clavicle which has been disarticulated from the acromioclavicular joint. By using this to lift the tumour a subperiosteal dissection was carried out from lateral to medial (blue arrow).

Once the superior and inferior poles of the tumour had been undermined the acromioclavicular joint was identified and disarticulated. This allowed us to use this lateral bony remnant of the clavicle to elevate the tumour to expose its remaining inferior aspect. Using a combination of diathermy and blunt dissection the bed of the tumour was freed from lateral to medial working strictly in the sub-periosteal plane on the posterior clavicular border (Fig. VII).

Once reaching the medial border the sternoclavicular joint was disarticulated and the claviculectomy was completed. The area of the operative bed was noted to pulsate due to the very close proximity of the subclavian artery beneath the subclavian vein. Just lateral to the sternoclavicular joint the parietal pleura of the lung was noted below and behind the 1st rib. Fortunately none of these structures were encountered or breached during the dissection. The tumour was removed in its entirety with no breach of its overlying capsule (Fig. 8).

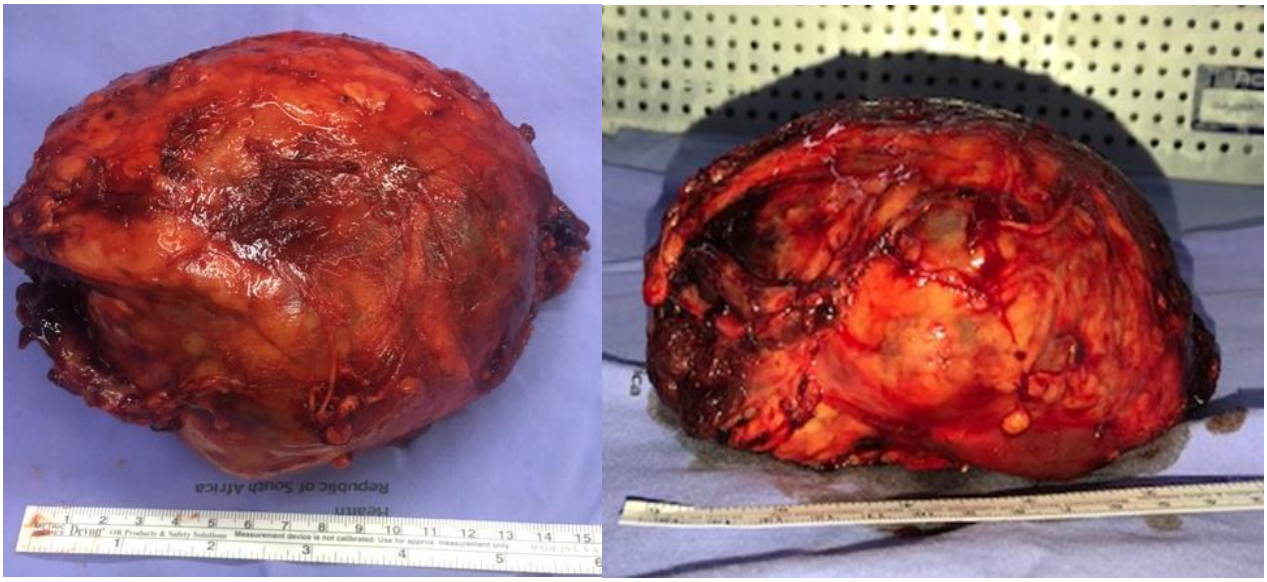

Figure VIII: Per-operative photographs of the chondrosarcoma on a sterile drape. In these photos its truly massive size is appreciated measuring 15×12×10cm.

After removal of the tumour the surgical site was irrigated thoroughly, haemostasis ensured and closed in layers with a Portovac drain in situ. Post-operatively the patient had no neurological deficit with normal power and sensation in his right upper limb being demonstrated. A post-operative Xray was taken, which on the anteroposterior view confirmed that complete claviculectomy had been performed (Fig. IX).

The patient's wound healed well and the histology report confirmed the tumour to be a chondrosarcoma of grade II with clear margins. He was referred to the general surgery department for dissection of his involved regional node as well as to the oncology department for consideration for adjuvant therapy. At one year follow up no recurrence was noted and the patient had good functions of the shoulder.

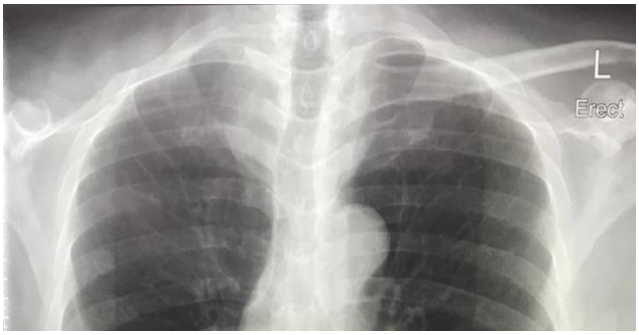

Figure IX: Post-operative anteroposterior chest X-ray confirming a complete claviculectomy. The disarticulation through the acromioclavicular and sternoclavicular joints ensured no remaining bone would be left.

DISCUSSION

Clavicular chondrosarcomas are characteristically painless due to the clavicle being a non-weight bearing bone.[8] Radiologically clavicular chondrosarcomas are commonly radiolucent on Xray imaging, iso-dense to surrounding muscles on CT imaging and on MRI imaging their cartilaginous matrix makes them hypo-intense on T1W and hyper-intense on T2W, imaging.[9] Our case report demonstrated all these typical radiological features (Fig. II-IV). Regarding the yield from fine needle biopsy in clavicular chondrosarcomas, confirmation of chondrosarcoma in 26% to 94% cases had been reported.[8] However other studies recommended that needle biopsy should be avoided altogether in clavicular tumour due to an unacceptably high risk of neurovascular injury as the clavicular cortex is softened by the tumorous process and advocated incisional biopsy as a preferred and safer method for diagnosis.[10] The risk of tumour seeding along an incisional biopsy tract and the large size of our patient's tumour made the risk of neurovascular injury small and supported our decision to perform a fine needle biopsy. The pre operative diagnosis of chondrosarcoma helped us to perform an enbloc claviculectomy but ensuring that no breach of the capsule occurred.

Generally chondrosarcoma exhibits poor response to both chemotherapy and radiotherapy. However the disease progression had been slowed in some patients.[7] Our case report had a large tumour size of 15 ×12×10cm and were able to achieved clear margins**.** One study reported 3 years survival rate of chondrosarcoma patients 73% with tumour size and adequacy of resection being independent predicators of survival.[11]

The presentation of clavicular chondrosarcoma was a painless mass of huge size due to delayed presentation in our case report. However if entrapment of the middle suprascapular nerve occurs pain usually compel the patient to present earlier.[4]

Abdehgah and colleagues[12] presented a case report of a chondrosarcoma clavicle in a 22 year old lady. The tumour size was 10×6 cm and histologic grade was II just like our case report. They treated the tumour with claviculectomy and partial sternum resection. At one year follow up their patient had no recurrence. It has been reported that histologic grade I chondrosarcoma had 5 year survival rate of 90% while grade II and grade III had 60% survival rate.[13]

Fine needle aspiration cytology (FNAC) confirmed the diagnosis in Abdehgah case report as well as in our case report. Some authors however, recommended repeat FNAC or open biopsy if negative results are obtained.[8]

When we searched the literature we found some interesting case reports of chondrosarcoma clavicle associated with unique signs and symptoms. Kapoor[14] reported a case report of a chondrosarcoma medial clavicle in a 28 year old soldier with a three year history of pain in shoulder and ptosis. The chondrosarcoma was arising from osteochondroma (size 50×45×25 mm) and had caused Horner's syndrome. The patient was treated by resection of medial clavicle. The patient had no recurrence at one year follow up but ptosis had persisted. Kobayashi[15] reported chondrosarcoma arising from osteochondroma of left clavicle in a 26 year old man. The tumour had caused thoracic outlet syndrome. The patient was treated with claviculectomy and symptoms of thoracic outlet syndrome were relieved. The patient had no functional deficit and no recurrence was noted at three years follow up.

We evaluated our patient at one year follow up and no recurrence or metastasis was noted. However longer follow up is mandatory.

CONCLUSION

Complete surgical resection in the form of total claviculectomy with clear margins offers the best chance of cure in patients with chondrosarcoma clavicle. We noted the procedure to be safe and recommend that a step-wise dissection in the manner we described should be performed. Pre operative work up must be complete and the approach must be multidisciplinary involving the Orthopaedic surgeon, general surgeon, thoracic surgeon, vascular surgeon, oncologist, radiologist, physiotherapist and rehabilitation expert.

Conflict of Interest: None

Grants/Funding: None

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