

AGGRESSIVE JUXTACORTICAL CHONDROMYXOID FIBROMA OF THE PROXIMAL THIGH REQUIRING COMPLETE PROXIMAL FEMORAL RESECTION

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Sarcoma Service

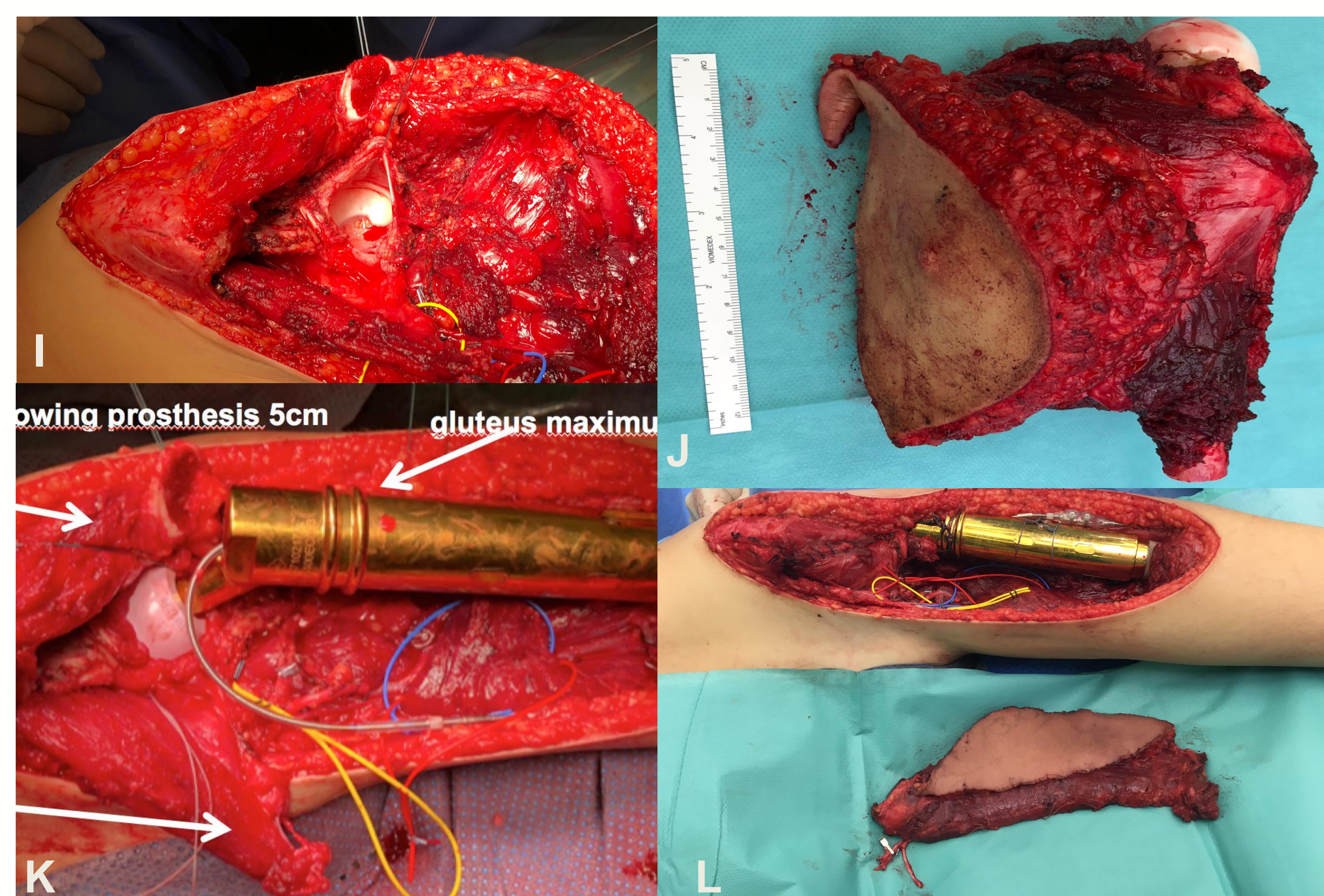
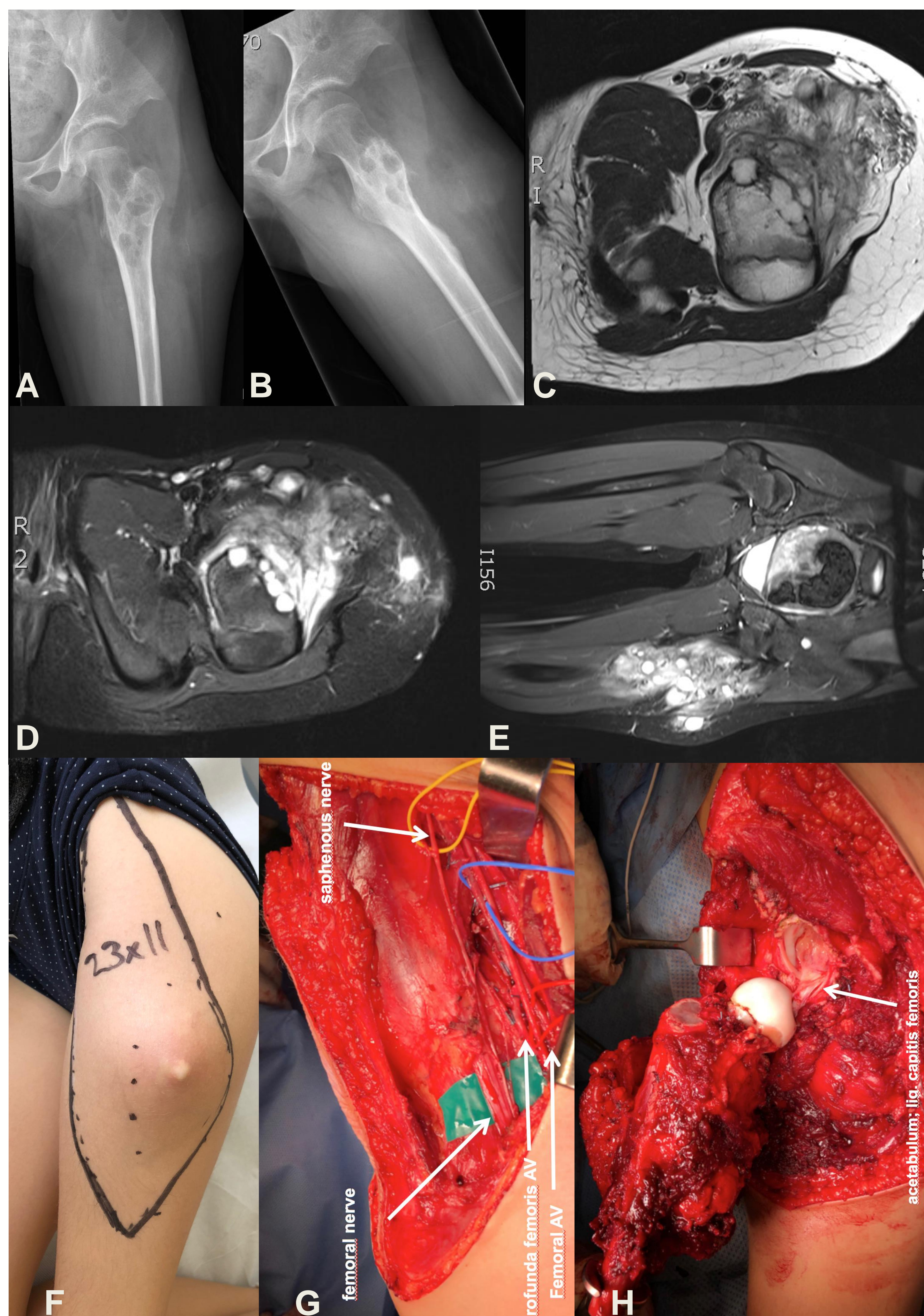
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INTRODUCTION

Chondromyxoid fibroma (CMF) is a very rare, benign bone tumor of cartilaginous origin, which has the tendency to locally recur in up to 15% after curettage. The WHO classifies it therefore as intermediate malignancy (local recurrence, no metastasis). Juxtacortical localisation is an extremely rare variant of CMF, whereas malignant behaviour has been noticed but not reported so far.

PATIENT & METHODS

A 10 year old girl was diagnosed with a juxtacortical CMF of the proximal anterior femur (A-E). The tumor's soft tissue mass was 5cm, with cortical penetration of 1cm. To keep the morbidity as low as possible, it was elected to resect the soft tissue part en bloc, and to curett the bony part. The initial postoperative course was uneventful.



RESULTS

Three months post surgery, a 10cm diffuse recurrence was noted, mainly in the soft tissues, as well as within the biopsy tract (A-E). The case was internationally reviewed both pathologically and surgically, and another curettage for the confirmed CMF was suggested. The parents postponed this for religious reasons. 10 months postoperatively. The tumor mass was ever increasing, involving now the entire subcutaneous tissues as well as the entire quadriceps muscle origin, with impending fracture of the proximal femur. We elected to resect the proximal femur together with the entire origin of the quadriceps muscle as well as a soft tissue island of 23x11cm (F-L). For reconstruction we used a growing prosthesis, and a functional, neurotized latissimus dorsi muscle with a generous skin island. Four months post surgery, the patient is pain free, walks without crutches and is free of tumor.

CONCLUSION

Although CMF is considered benign or locally of intermediate grade, it may show an aggressive behaviour like a malignant tumor and needs to be treated as such. NextGen sequencing did not detect any genetic abnormalities which may allow to distinguish such more aggressive subtype from benign forms.

HIGHLIGHTS:

A functional latissimus dorsi transfer may be used to address a large soft tissue defect in the proximal femur, the loss of the femoral nerve as well as (in part) the quadriceps motor function.