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MULTICENTRIC EPITHELIOID HEMANGIOENDOTHELIOME OF BONE CASE REPORT WITH LIMB SALVAGE AND LONG TERM FOLLOW UP

Introduction :

Multicentric epithelioid hemangioendothelioma of bone is a rare tumour classified as malignant. Our observation addresses his real nature and question the necessity of mutilating treatment.

Case report :

A 13 year old girl was referred to us from another orthopaedic department for a 18 months pain in the right leg spreading down from knee to foot while walking. She has no history o f infection or trauma. Physical examination was normal except a painful limited flexion of knee and a discrete swelling of the foot. Plain radiographs revealed multiple osteolytic lesions in the distal part of the right femur, proximal and diaphyseal tibia and foot.

The definitive diagnosis of multicentric epithelioid hemangioendothelioma of bone Grade 1 was established by open biopsy of femur and confirmed by subsequent histological examinations of material curetted of all sites.

To avoid mutilation in this young girl we performed in intralesionnal curettage with cryotherapy by liquid nitrogen followed by osteosynthesis and cementation of both distal femur and proximal tibia. Distal tibia and foot lesions were curetted and cemented without cryotherapy to avoid skin complications. Neither chemotherapy nor radiotherapy was delivered.

Post operative screening included physical examination, plain radiographs, and MRI every 3 months for 2 years, every 6 months for 2 years and every year thereafter. At last screening, with 14 years follow up the patient enjoys a normal life (only heavy sports are prohibited) with no pain, normal flexion of the knee and no evolution on any site. No new lesion appeared.

Discussion :

Multicentric epithelioid hemangioendothelioma of bone is generally considered as a sarcoma with unpredictable course. The 5 year disease free survival of the 89 patients reported by Srinivasan was only 24% but many authors emphasise the grading of the tumour as main prognosis factor.

Intra lesionnal treatment without adjuvant of real malignant lesion should be followed by recurrence. In this case with long follow up all lesions remain stable. For femoral and tibia lesion it could be explained by cryotherapy but not for the foot locations were only curettage and cementation was applied. Therefore le real malignant potential of grade 1 Multicentric epithelioid hemangioendothelioma of bone of our case and the mutilating procedure usually advocated as only therapeutic with intend to cure appeared questionable.

Conclusion :

The favourable evolution of this grade 1 multicentric epithelioid hemangioendothelioma of bone confirms the prognostic value of grading of this tumour and pleas for individualized surgical treatment of patient avoiding mutilating surgery when feasible.
The hemimelic presentation of lesion also suggests a strong genotypic etiologic factor and may justify further study.