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Tibial periosteal Ewing sarcoma

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Introduction: Ewing sarcoma is classically known as "big simulator". Reconstruction after resection is sometimes difficult, there is not a perfect option. We present a case of an periosteal Ewing sarcoma, an extremely rare entity.

Method and Results: A young sport man(21 years old) refered a postraumatic pain in right tibia since 3 years. He had an increasing pain and swelling mainly in the last 6 months. No relevant antecedents exist. Physical exploration was normal, except to a 5x5 cm mass in medial border of distal third of the right tibia. An important periosteal reaction at the distal third of right tibia was seen by X-Ray. MRI and CT informed the presence of periosteal mass compatible with periosteal post-traumatic haematoma. Percutaneous biopsy was done and the pathology report suggested round cell malignant tumour of soft tissue, Ewing's sarcoma as the first possibility. Bone marrow biopsy revealed no infiltration by the tumour. The sarcoma conference decided a neoadjuvant chemotherapy treatment and after that, surgery and adjuvant chemotherapy. After neodajuvant chemotherapy and new staging, the soft tissue mass decreased and we decided a limb-salvage surgery. Wide intercalary resection of distal tibia (14,5cm) was done, continued by reconstruction with free combined anterolateral thigh flap (ipsilateral) and vascularized fibula bone graft (contralateral). The complex was synthetized with a locking compression plate One week later, thigh flap failed and was substituted by a free vascularized latissimus dorsi flap. Definitive pathologic study revealed a periosteal Ewing's sarcoma from the right tibia which is a very rare malignant tumour presentation. Fourteen months later, the patient walks in partial weight bearing with two canes, without any signs of local recurrence and no pain. X-ray shows a union of the distal fibula graft. However, the proximal junction is still without clear union. The patient has not metastatic disease at the last revision.

Conclusions: Periosteal Ewing sarcoma is an extremely rare entity. Few cases have been reported. Large intercalary bone defects are still challenging and difficult to manage, even when a good surgical technique is performed.