Title: Teleangiectatic osteosarcoma: an analysis of 87 cases.

**Abstract:** Introduction: Telangiectatic osteosarcoma (TOS) is a rare subtype of osteosarcoma. We review our experience to characterize its prevalence, treatment, relapse and survivorship at long term follow-up.

Methods: Eighty-seven patients aged from 4 to 60 years (mean 20 years), were treated from 1985 to 2008. Lesions affected the femur (38), humerus (20), tibia (19), fibula (4), pelvis (3), foot (2) and radius (1). Eight patients had metastatic disease at diagnosis. Seventy-eight patients were treated with neoadjuvant chemotherapy with three or more drugs according to different protocols, nine had surgery as first treatment. Limb salvage surgery was performed in 71 cases, amputation in 14 and rotationplasty in one. One patient died before surgery. Prognostic factors were evaluated with Kaplan-Meier analysis.

Results: At a mean follow-up of 8 years, overall survival was 81%, 65% and 65% at 2, 5 and 10 years respectively. Fifty-two patients were disease-free, three were alive with disease, twenty-nine died with disease and three died of other causes. Thirteen local recurrences were observed. Twenty-three patients developed lung (20) or bone (3) metastases. Pathologic fracture did not significantly influence survivorship. Prognostic influence of age of the patients was evaluated at three different cut-off (15, 20 and 25 years-old): younger patients had better survivorship, without statistical significance. Induced necrosis according to Huvos' classification was significant at both univariate and multivariate regression Cox analysis (p=0.0001).

Conclusion: TOS does not have a poor prognosis as previously reported in the literature. A high percentage of patients can be cured with neoadjuvant chemotherapy and surgery. In most patients, limb sparing surgery is possible and safe.