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Axial osteosarcoma: a 25 year monoinstitutional experience in patients younger than 19 years.

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Background. The survival of patients with pelvic or axial osteosarcoma (OS) remains poor and the management of these patients is particularly challenging.
Patients and methods. Object of this study is a cohort of unselected patients aged Results. Twenty patients between 3-19 years (median age 14) were included. Five patients were metastatic. The most frequent histological subtype was chondroblastic OS (45%) followed by osteoblastic OS (30%). Eight patients had pelvic OS, 8 axial OS and 4 mandible/maxilla OS. All patients received chemotherapy. Necrosis post chemotherapy was evaluable in 9 patients (≥ 90% in 3 cases). Surgery was performed in 12 patients (3 amputations). Radiotherapy was delivered in 7 patients (total dose 24-60 Gy). Median follow-up was 35 months (8-276), 5-year overall survival was 40% and 5-year event free survival was 37%.
Six patients are alive: 2 with pelvic OS (both had a good response to chemotherapy, one underwent hemipelvectomy and the other received radiotherapy); 1 with axial and multicentric OS (good histological response and radical surgery); 3 with mandible/maxilla OS (a fourth patient died of sepsis during chemotherapy). One patient with axial OS died because of a second bone tumor and another one for breast cancer. We highlight that 6 patients had a p53 mutation: 2 are alive, 2 died for OS, 1 for breast cancer, 1 for glioblastoma.
Conclusion. Adequate local treatment and good pathological response are relevant for prognosis of axial OS, that remains dismal. An impressive number of p53 mutations are reported in our series.

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