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OSTEOSARCOMA IN PATIENTS WITH ROTHMUND-THOMSON SYNDROME

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Background: Rothmund-Thomson syndrome (RTS) is associated with an increased risk of osteosarcoma, but information about affected patients is limited. This retrospective analysis explored the clinical features of high-grade osteosarcomas in patients with RTS and outcome after multimodal therapy.

Methods: The Cooperative Osteosarcoma Study Group (COSS) database was searched for eligible patients. Seven patients with high-grade osteosarcoma had a diagnosis of RTS and their patient-, tumor- and treatment-related variables and outcome were reviewed.

Results: Median age at diagnosis was 13 years (range 7-16), 5 were female, 2 male. Tumors involved proximal tibia (4), distal tibia (1), distal fibula (1) and proximal ulna (1). Most frequent subtypes were osteoblastic (3) and malignant fibrous-histiocytoma-like osteosarcoma (2). Three patients had metastatic disease at diagnosis. All patients were treated with surgery and chemotherapy according to COSS-protocols. All but one were underweight at start of treatment, at least four needed nutrition as support during therapy. Four patients received chemotherapy as scheduled, the other three required dose modifications and terminated treatment prematurely. A wide resection of the primary tumor was achieved in all individuals. Two of three patients failed to achieve surgical clearance of their primary metastases and died, the third relapsed with multiple metastases and also died. Two of the four patients with localized disease remained alive in 1st complete remission (CR), 10.5 and 17.6 years after diagnosis, a third patient was in 2nd CR after surgery and chemotherapy for recurrence (solitary lung metastasis), with a follow-up of 13.0 years. The fourth patient, for whom osteosarcoma was already the third primary malignancy, died of acute leukemia, 7.0 years after diagnosis and while still in 1st CR of osteosarcoma.

Conclusion: Patients with osteosarcoma in RTS may be cured with appropriate multimodal therapy. They should be treated like other osteosarcoma patients but individual features and special support have to be considered.

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