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Significant progress in the diagnosis and treatment of rare solid tumours such as soft tissue and bone sarcomas, which occurred in recent years, is the result of the concentration of patients with these cancers in specialised reference centres, international cooperation, and the achievements of molecular diagnostics. The key in the management of primary cancer as well as in the case of recurrence of the disease is planned multidisciplinary treatment — a combination of surgery (basic method) with radiotherapy (RTH) and/or chemotherapy (CTH) and physiotherapy [1–4]. Thanks to this, the chances of limiting the scope of resection and obtaining long-term survival or cure are significantly increased.

Modern, individualised combined therapy, including reconstructive procedures, saves the limb in the majority of patients with sarcomas of the aforementioned location (in reference centres amputations are currently performed in less than 10% of patients). There is a steady but slow increase in the percentage of patients with sarcomas with long-term survival (the current five-year survival rate for sarcomas with a limb localisation is 55–78%). The prognosis at the metastatic stage is still poor (median survival: about 12–18 months). Favourable local results are achieved only by patients with sarcoma after scheduled (previous biopsy and comprehensive imaging multidisciplinary evaluation) complete excision of the primary lesion within the microscopically tumour-free margins (resection R0).

Following correct diagnosis, the majority of patients after complete resection require supplemental RTH (soft tissue sarcomas) or perioperative chemotherapy (bone sarcomas) and many weeks' rehabilitation, and control for at least five years in the centre that carried out the treatment. The introduction of drugs targeting molecular or genetic alterations in cells that participate in aetiopathogenesis (e.g. denosumab in giant cell tumour of bone, kinase inhibitors in gastrointestinal stromal tumours, or imatinib in dermatofibrosarcoma protuberans [DFSP]) has also contributed to the improved prognosis of patients [3–5].

References

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