Dear EMSOS members,

Within the framework of the EMSOS I would like to propose a study addressing a very rare bone tumor entity in order to optimize ongoing treatment modalities.

Primary leiomyosarcoma of bone (LMSoB) makes up only <0.7% of primary malignant bone tumors. In a multicenter study from 4 referral centers in Germany (PD Dr. Tunn and Dr. Niethard - Clinic for Tumor Orthopedics, Sarcoma Center Helios Clinic Berlin-Buch; PD Dr. Knebel - Department of Orthopaedics and Sports Orthopaedics, Technical University Munich; and PD Andreou – Dept. of Tumor Orthopedics, Helios Clinic Bad Saarow) and Austria (Prof. Dr. Leithner - Medical University of Graz) between 1993 and 2018 we could compile 35 cases in order to investigate the optimal treatment options. LMSoB shows similar histopathological properties as soft tissue leiomyosarcoma. Current treatment approaches include besides the surgical wide resection the use of perioperative chemotherapy, in analogy to the treatment of osteosarcoma or highly malignant soft tissue leiomyosarcoma. Data on the effectiveness of these therapeutic protocols in LMSoB is rare. One of the largest published multicenter studies from the Japanese musculoskeletal oncology group recruited 48 cases showing no benefit for patients receiving a cisplatin based chemotherapy. A published poster at the ESMO 2019 presenting data from the EURO-B.O.S.S. study group included 20 patients showing that LMSoB had a worse overall survival than high grade osteosarcoma treated in the protocol.

We hypothesize that chemotherapy analogous to the therapy of soft tissue leiomyosarcoma might be superior to chemotherapy protocol for bone sarcomas.

Our preliminary results with a median follow-up of 49 months showed an event-free survival (EFS) after 2/5 years of 60%/45%. Disease-specific survival (DSS) after 2/5 years was 91%/87%. 64% developed distant metastases after a median of 20 months (IQR 12-63 months). At last follow-up 31% of the patients were alive with their disease, 40% with no evidence of disease and 20% died from their disease. 84% underwent surgical treatment. 94% had an R0 resection. 53% of patients with a high-grade tumor treated perioperative chemotherapy. Survival improved significantly for patients with surgery (DSS: p=0.011) and for high-grade tumors located on the extremities (DSS: p=0.003, EFS: p<0.001). Perioperative chemotherapy for high-grade tumors was not associated with an improvement in DSS (p=0.719) or EFS (p=0.858).

We could conclude that surgery remains the most important treatment option for LMSoB. Patients with extremity localization had a better prognosis. Potential for distant metastasis is high (64%). Therefore the need for effective systemic therapy is high. In the reported 35 patients the use of perioperative chemotherapy had no effect on EFS or DSS. We therefore seek to get a much larger number of patients through EMSOS in order to evaluate the hypothesis that LMSoB should be treated as its soft tissue equivalent.

I have attached a corresponding Excel table and am available at any time for any questions.

Thank you for your participation,
Best regards from Berlin,
Maya Niethard

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