2019 WSLS Poster 015

Secondary Angiosarcoma: a fatal complication of chronic lymphedema

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Abstract: Secondary Angiosarcoma (SA, Stewert Treves Syndrome) is a rare malignant cutaneous lesion, which arises in chronic lymphedema of the extremity, often observed after breast cancer treatment. We reviewed the history and the oncological outcome of two patients with SA treated with multimodal therapy. Both patients were suffering more than 5 years from massive lymphedema after oncological treatment of breast cancer with mastectomy and radical lymph nodes dissection followed by radiation therapy. The lymphedema extended from the upper arm to the forearm and the dorsal parts of the hand. Both patients were referred to our cancer center after developing the multiple reddish blue macules and palpable subcutaneous nodules on the skin affected by lymphedema. Biopsies of the lesions revealed the diagnosis of SA in both cases. Patients were discussed in our interdisciplinary tumor board and received multimodal therapy including radical epifascial skin excision, hyperthermic isolated limb perfusion with TNF-alpha and Melphalan. The first patient developed local recurrence and metastatic disease 12 months afterwards. She received palliative chemotherapy and died of disease. The second patient had no evidence of disease 18 month after therapy with sufficient hand function. Though the incidence of lymphedema has decreased in the last decades, possibly due to improvements in operative techniques and improvements in radiation therapy, SA is still a severe diagnosis with high morbidity and high mortality. Multimodal therapy including hyperthermic isolated limb perfusion with TNF-alpha and Melphalan, combined with radical resection of the affected skin, subcutaneous tissue including the fascia, with large safety margins may lead to a better local control and thus possibly less metastatic disease. Extensive use of lymphatic supermicrosurgery for the treatment of chronic lymphedema can probably further decrease the incidence of SA.