28th Annual Meeting of the
European Musculo-Skeletal Oncology Society
\& 16th EMSOS Nurse and Allied Professions Group Meeting

April 29th - May Ist 2015
Athens, Greece

PP-189
Multifocal pseudomyogenic haemangioendothelioma of bone: a case report of a young man with lung metastases
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Introduction: Pseudomyogenic haemangioendothelioma (also called epitheliod sarcoma-like haemangioendothelioma) is a new entity firstly described in 2011 by Fletcher and Hornick. It is a rare intermediate vascular tumor which can be multicentric, rarely metastasizing. It predominantly occurs in lower and upper limb of young male patients.
Case: We report the case of a 22-years-old male patient who referred to his doctor with swelling and pain 20 days after a trauma on his right foot. The X-Ray (Figure 1), MRI and CT examinations detected multiple aggressive osteolytic lesions in several bones of the foot (distal phalanx of first and second finger, head of fourth and fifth metatarsal bones). Open biopsy of the fifth metatarsus showed pseudomyogenic haemangioendothelioma. Immunohistochemically the tumor cells expressed ERG, CK AE1/AE3, CK CAM5.2, INI1, Smooth M Actin, CD31 whereas MS Actin, Desmin and Podoplanin were negative. Bone scan and chest CT showed asymptomatic intramedullary lesions in the right posterior acetabular column and in D10 and multiple lung nodules (Figures 2-4). The treatment has been different in the different anatomic sites: we performed amputation of the fifth finger and of the first and second distal phalanges, and curettage of the fourth metatarsus (Figure 5). In agreement with the oncologist we decided to follow-up the lung lesions and the other bone lesions and to treat the patient with denosumab ( 120 mg sc per month).
Conclusion: Pseudomyogenic haemangioendothelioma of bone is an unpredictable lesion and the treatment, still unclear, requires a multidisciplinary approach.


Figure 1


Figure 2

Figure 3



Figure 4


Figure 5

