The EMSOS+ Adamantinoma study: a clinical, radiological and histopathological analysis of 192 cases.

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Introduction and Objectives
Adamantinoma of the long bones is a low-grade, slow-growing, primary malignant bone tumour composed of epithelial cells in a fibrous or osteofibrous stroma. Current studies show that adamantinomas appear to be of epithelial nature. Histologically there is heterogeneity in appearance of the epithelial (tumour) cells within this lesion. This has led to the distinction of osteofibrous dysplasia-like adamantinoma (OFD-AD) and classic adamantinoma (AD). In the first lesion cells are often thinly spread throughout and osteofibrous dysplastic stroma while in the latter the tumour consists largely of lesional cells.

Adamantinomas account for 0.3%-1% of malignant bone tumours, which makes this an orphan disease. Pooling of data from multiple specialised bone tumour centres becomes essential for research. This paper is the mid-term update to this EMSOS+ adamantinoma study.

Methods
A global multicenter retrospective database was created in which patient demographics details, clinical and radiological details as well as histological and surgical treatment details were collected. Only centers with experience in treating rare bone tumors were asked to participate. The questionnaire was surgeon completed. Patients were included if > 24 months follow-up (FU), inclusion between 1975-2014. Cox regression was used to assess the risk of several factors for local recurrence in OFD-AD and AD.

Results
192 patients (95OFD-AD) with a minimum FU of 24 months were included in this study. Mean age at presentation for the tumors combined was 24 years (median 18). 47% of patients are male. OFD-AD was diagnosed 16 years earlier than classic AD (16yr vs 32yr). Patients had a mean FU of nearly 9 years (2-32yrs range). 53 patients (28%) experienced local recurrence (LC) (mean) 51months (3-240) after initial diagnosis. Recurrences were spread equally between OFD-AD and classic AD(25 vs 28). 15 patients (15%) experienced metastatic disease and 11(13%) patients suffered fatal disease, all cases were considered classic AD cases.

Multivariate Cox regression (covariates; size>+- 5cm<, pathological fracture >Y/N<, free resection margins>Y/N<) showed that free microscopic resection margins are significantly protective for the risk of LR (HR0.2 CI95% 0.1-0.4) in both OFD and AD. Pathological fracture increased the risk for LR in AD cases only (HR3.4 CI95% 1.1-10.2). There were no recorded cases of progression of diagnosis from OFD-AD to AD after initial recurrence.

Discussion/Conclusion
Orphan diseases such as adamantinomas require collaborative international studies to advance research. Therefore, we have set up a global EMSOS+ study/partnership with many renowned specialist bone tumor centers globally. OFD-AD tumors behave locally aggressive and may be considered as borderline tumors, while classic adamantinomas can metastasize and result in a fatal outcome. Both tumors have shown a substantial risk for LR and recur at similar rates. In all patients with adamantinomas we advocate wide resection margins and extended FU.

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