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Title: Osteofibrous-dysplasia-like-adamantinoma and classic adamantinoma: a clinical, radiological and histological analysis of 82 cases.

Background: Osteofibrous-dysplasia-like-adamantinoma (OFD-AD) and classic-adamantinoma (AD) are rare malignant bone tumours that present mostly in the anterior tibia diaphysis before the age of thirty. They comprise between 0.1 – 0.5% of total primary bone tumours. Despite radical surgery, reported recurrence rates stand at around 30%. It is not clear what factors contributes to this high recurrence rate or if there are predictive factors that indicate to the likelihood of recurrence.

Questions/purposes:

1. A clinical, radiological and histological analysis of 82 cases.
2. To determine which factors contribute to high recurrence rates of AD and OFD-AD tumours.
3. To determine factors that predict local recurrence

Patients and Methods: Through our musculoskeletal sarcoma databases, we retrospectively identified all OFD-AD/AD patients treated at two tertiary referral centres for bone tumors and evaluated clinical, radiological and histological variables in a risk analysis for local recurrence. Statistical analysis was carried out using SPSS (22.0) software.

Results: 82 consecutive OFD-AD (n=50)/AD (n=32) cases were identified (1985-2014) (table 1). Mean follow-up and age at diagnosis were 101 months (13-207) and 23 (1-69) years respectively. There was a significant difference in mean age at diagnosis between OFD-AD and AD (18 vs 32 years ($p=0.015$)). 13 patients were treated with intralesional and 69 with a free resection margins. Surgical reconstruction was performed using allografts in 53% as singular treatment, in 7% cases both a vascularised fibula and allograft was used and in 9% a vascularized fibula was used alone, in 2% patients only curettage was performed, in the remainder (27%) prosthetic devices were used and in once case an amputation. 23 patients (28%) experienced local recurrence (LC) after 44 (mean) months (13-240). Thus far one patient died of progressive disease 44 months after initial diagnosis. One patient died of metastasized osteosarcomatous dedifferentiated adamantinoma. Both groups (OFD-AD/AD) presented with similar local recurrence rates: 13 (26%) versus 10 (31%). Surprisingly we found no significant risk (OR(range)) for local recurrence; for positive microscopic resection margins (0.9 (0.5-1.6)), size of the tumour (>5 cm) (1.4 (0.4-4.3)), multiple lesions (0.7 (0.2-2.7)) history of pathological fracture (2.8 (0.6-13.1)), or paediatric patients (2.6 (0.8-8.4)) or periosteal tumour involvement (1.1(0.5-2.5)). Surgical procedures such as resection of the biopsy tract or periosteal resection did not reduce the probability of recurrence. Tumours that recurred did show increased contrast enhancement on initial dynamic MRI, compared to none-recurrent tumours (OR 6.1 (0.9-42,8) $p = 0.045$). A trend can be demonstrated that larger resection margins around a tumour lead to longer intervals for recurrence (figure 1).

Figures and tables:

Table 1. Baseline characteristics of patient included in this study.

Characteristic	Result
n of Patients	82 (100%)
Patient demographics:	
Mean age (years) at diagnosis (range)	23 (1 – 69)
Median age (years)	18
Sex (male <i>versus</i> female)	35 <i>versus</i> 47 (43% - 57%)
Follow up time in months (range)	101 (13-207)
Tumour characteristics:	
Lesions (single <i>versus</i> multiple)	66 <i>versus</i> 16 (80% - 20%)
Mean size (cm) of lesions (range)	5.3 (0.6 – 21)
Diagnosis (OFD-ADA <i>versus</i> ADA)	50 <i>versus</i> 32 (61% - 39%)
Location:	
Tibia	98%
Diaphysis	84%
Left <i>versus</i> right	30 <i>versus</i> 52 (37% - 63%)

Treatment:

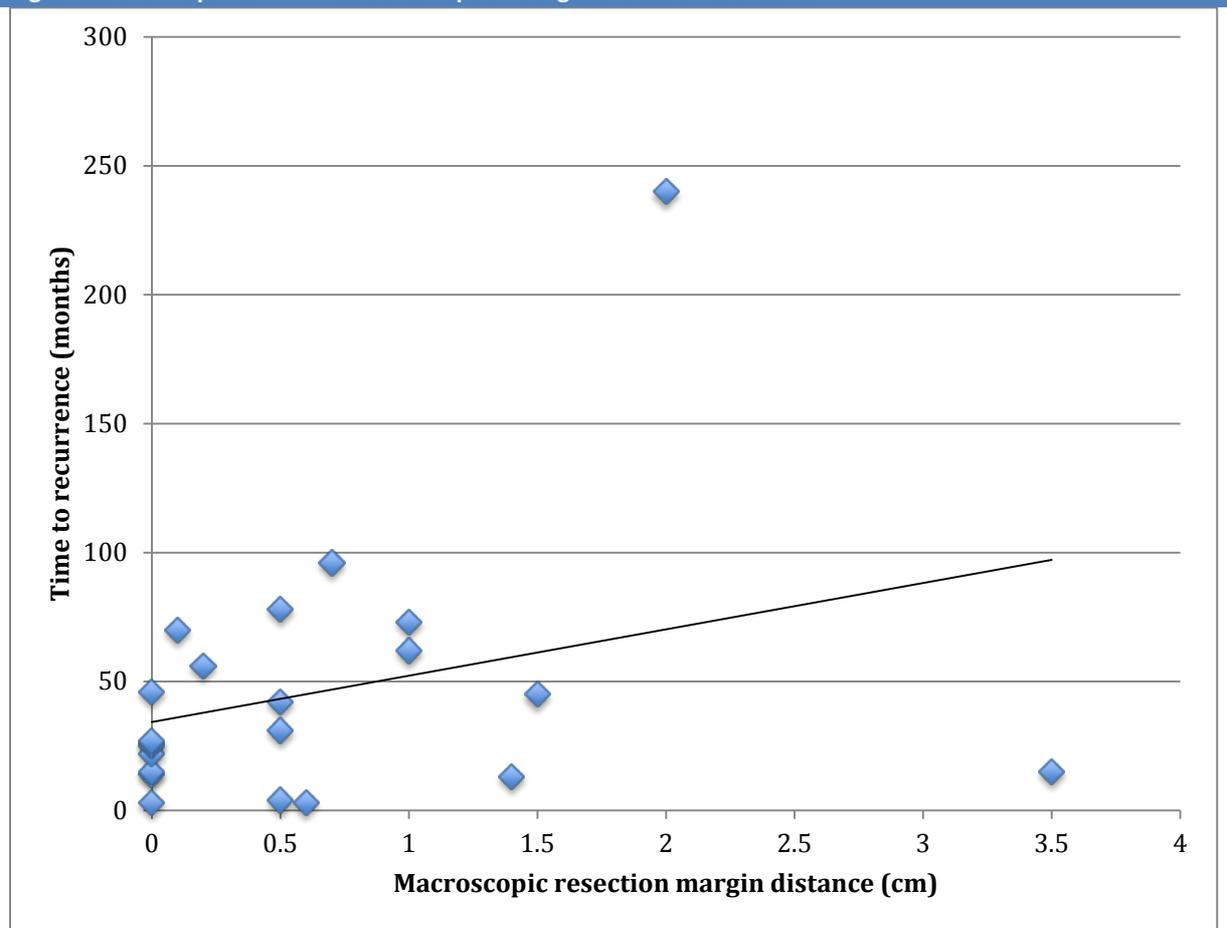
Mean narrowest macroscopic resection margin (range)	1 cm (0 – 7)
Per-operative cut into lesion	Yes 13 (16%)
Reported complications	21 (26%)

Outcome:

<i>n</i> of recurrence	23 (28%)
Mean time (months) to recurrence (range)	44 (3-240)
<i>n</i> of Fatal disease	2
<i>n</i> of metastatic disease	2

Abbreviations: ADA: adamantinoma classic type. OFD-ADA: osteofibrous dysplasia like adamantinoma.

Figure 1. Scatter plot resection distance plotted against time to recurrence.



Conclusions: Adamantinoma's are rare malignant bone tumours and thus the creation of a multicentre database including patient history, surgical-, histopathology- and radiological reports: as well as extended follow-up is mandatory in the further understanding this malignancy. We have demonstrated that gadolinium enhanced dynamic MR imaging may be a useful tool in identifying patients at risk of local recurrence and thus can be seen as a useful tool for diagnostics and follow-up. As a result of our analysis we recommend wide resection even in OFD-AD cases; in order to prevent local recurrence and especially progressive disease. Long-term follow-up is advocated as recurrence can occur up to two decades after index surgery.

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