

Desmoplastic Fibroma of bone

Desmoplastic Fibroma of bone is a rare, locally aggressive bone tumor. Once it was called the osseous counterpart of extra-abdominal desmoid tumor, because of the histological aspects. In 2013, the WHO described the microscopic appearance of desmoplastic fibroma as being composed of slender, spindle to stellate cells with minimal cytological atypia and abundant collagenous matrix.



The true incidence of desmoplastic fibroma of bone is unknown but it is estimated with ~0.1% of all primary bone tumors. It usually occurs in adolescence and young adults. The typical location seems to be below elbow or knee, but some cases are reported with a presentation in the pelvis. The treatment of choice range from intra-lesional curettage to wide surgical excision and also adjuvant radiotherapy has been reported to be a therapeutic option. However, marginal or intralesional excisions, leading to a high rate of local recurrence 15%-72%.

Up to date only view case reports and a small number of case series regarding desmoplastic fibroma have been published. A multi-institutional analysis of a larger series would provide valuable information regarding the natural disease course and help sharpen the therapeutic decisions.

We would therefore like to ask you to contribute your cases to an EMSOS-wide study.

Goals:

We hope to include a total of at least 60 patients with desmoplastic fibroma of bone treated between 1990 and 2016 with the aim to:

1. collect data regarding clinical characteristics, treatment, and outcome from multiple centers
2. evaluate the probability of local recurrence, metastasis and dedifferentiation
3. determine possible prognostic factors

Inclusion criteria:

- histologically proven desmoplastic fibroma of bone
- surgical or other treatment at your center

Data collection is planned from February 2018 to December 2018.

When including patients in the database:

1. Please be as thorough as possible, don't hesitate to contact us should you have any questions.
2. As the combined dataset will not include personal hospital identifiers from each institution please label your patients as 1,2,3 etc. Please keep a personal record of which number corresponds to which personal hospital identifier.

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We thank you for your help and participation.