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## DERMATOFIBROSARCOMA OF DARIER AND FERRAND LOCALLY ADVANCED AND METASTATIC: ABOUT 7 CASES.

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**Introduction:** Darier and Ferrand dermatofibrosarcoma is a rare soft tissue sarcoma characterized by a slow evolution with a major risk of recurrence in case of non-cancerous resection. Although large surgical excision remains the standard treatment, imatinib has a place in locally advanced and metastatic forms. The objective of this study is to determine the epidemiological, diagnostic, therapeutic and evolutionary characteristics collected from the clinical records of patients for dermatofibrosarcoma of Darier and Ferrand.

**Materials and methods:** This is a retrospective study conducted from January 2013 to December 2016 in the department of medical oncology at CHU Hassan II in Fez, including patients with Darier and Ferrand dermatofibrosarcoma locally advanced or metastatic.

**Results:** 7 cases were reported, 3 women and 4 men, the mean age was 46.2 years (range: 33-65). The tumor was located in the scalp in 3 cases, 1 case in the peri-umbilical, 1 case at the level of the chest wall and 1 case at the level of the scapular region. In all cases, there was histological evidence of the diagnosis of dermatofibrosarcoma. Immunohistochemical examination with CD34 antibody was performed in all cases showing positivity with cytoplasmic and membrane CD34 labeling. The tumor stage was

locally advanced in 5 cases and metastatic at the pulmonary level in 2 cases. Five patients had local recurrence in the initial site of resection. Imatinib was administered at a dose of 400mgx2 / day in all patients. The outcome was marked by stability in 4 patients and a partial clinical response in 2 patients after an average duration of 11 months, while 1 patient was lost sight of. Regarding tolerance to imatinib, 3 patients presented with a hand-grade I syndrome, 2 patients with thrombocytopenia and 1 patient with severe neutropenia, which required stopping treatment temporarily with reintroduction after resolution of neutropenia.

**Conclusion:** The dermatofibrosarcoma of Darier and Ferrand is a rare cutaneous tumor with slow local evolution taking place over several years. It is distinguished by its diagnostic difficulty, its tendency to recurrence and the rarity of its metastases which are essentially pulmonary. Imatinib is an effective therapeutic option with a good safety profile in the treatment of locally advanced or metastatic dermatofibrosarcoma.

### Biography

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