SKIN CANCER (OTHER THAN MELANOMA)

DERMATOFIBROSARCOMA ROTUBERANS (REPORT OF 50 CASES)

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Introduction: Dermatofibrosarcoma protuberans (DFSP) is a slow growing tumor with a very low metastatic potential but with significant subclinical extension and great capacity for local destruction.

Objective: elucidate the epidemio-clinical and pathological characteristics, management and evolutional aspects of DFSP with comparison with the data of the literature.

Patients and methods: we report a retrospective study of all histologically confirmed cases of DFSP, during 37 years.

Results: Fifty cases of DFSP (23 males and 27 females) were diagnosed. Mean age at diagnosis was 48.5 years (range 17-84 years). The tumor appeared before the age of 15 years in five patients (10%). This was a recurrence in 11,7% of cases. Six patients reported a history of trauma. The average delay before consultation was 5.6 years (range 1month-30 years). Solitary nodule (58%) and multinodular mass (34%) were the main clinical aspects of DFSP. Mean size of the tumor was 4.8 cm (range 1-20 cm). The most affected areas were the trunk (56%), extremities (26%) and the cephalic region (18%). The diagnosis of DFSP was histologically confirmed in all cases. Immunohistochemical study was achieved in 35 cases and showed positive staining for CD34. No metastases were identified. The treatment consisted of large surgical excision in all patients. Four patients had revision surgery because of tumor margins. Eight cases (17%) presented a local recurrence after a mean delay of 12.5 months.

Conclusion: DFSP is a rare disease and an improved awareness and understanding of this condition is required by dermatologists, surgeons and pathologists to allow its early diagnosis and treatment. Pathological examinations are required in patients with suspected DFSP, with the aim of minimizing the misdiagnosis rate. The main risk is tumor relapse. Then, DFSP requires prompt treatment by extended tumor resection, followed by an increased follow-up frequency.