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Atypical fibroxanthoma and pleomorphic dermal sarcoma - A 10-year retrospective study

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Introduction & Objectives: Atypical fibroxanthoma (AFX) and pleomorphic dermal sarcoma (PDS) are neoplasms whose definition has been subject to debate in recent years. Currently, it is argued that they constitute part of the same spectrum of malignant tumors with fibrohistiocytic origin, with PDS being a more aggressive variant with less favorable prognosis.

Materials & Methods: Retrospective study of patients diagnosed with AFX or PDS at the Dermatology and Venereology Department of the North Lisbon University Hospital Centre between 2013 and 2023, with description and analysis of demographic, clinical, histological, therapeutic, and evolutionary variables.

Results: Twenty-two male patients over 65 years old were identified. In 9 of the patients (39%), personal history of malignant cutaneous neoplasms was identified. Clinically, hyperkeratotic tumors and plaques were identified, with proposed clinical diagnoses of squamous cell carcinoma, basal cell carcinoma, malignant melanoma, atypical fibroxanthoma, primary cutaneous lymphoma, and hypertrophic actinic keratosis. Lesions were located on the scalp (40%), face (31%), ear (18%), shoulder (5%), and hand (5%). Overall, spindle cell neoplasms with marked nuclear pleomorphism, diffuse positivity for CD10 were observed, with several markers used for differential diagnosis with other malignant neoplasms. Subcutaneous invasion, vascular invasion, neurotropism, presence of necrosis, mitosis, and/or ulceration were the criteria used to histologically differentiate AFX/PDS. All patients underwent surgical therapy, with initial incomplete excision in 5 patients (23%), of whom 3 underwent adjuvant radiotherapy (17%). Lymph node metastasis was observed in a single patient, and no cases of distant metastasis were detected. None of the cases resulted in death directly associated with the neoplasm.

Conclusion: AFX and PDS are rare malignant neoplasms that often arise in photoexposed areas of elderly male patients. These malignant tumors lack characteristic clinical, histological, or immunohistochemical features, making their diagnosis and management a challenge in clinical practice.