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ORIGINAL ARTICLE

Atypical fibroxanthoma and pleomorphic dermal sarcoma: a 10-year retrospective study and review of the literature

Fibroxantoma atípico e sarcoma dérmico pleomórfico: estudo retrospetivo a 10 anos e revisão da literatura

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Abstract

Objectives: To characterize the demographic, clinical, histopathological, therapeutic, and prognostic features of patients diagnosed with atypical fibroxanthoma (AFX) and pleomorphic dermal sarcoma (PDS) in a tertiary center. **Methods:** Retrospective observational study conducted from January 2014 to December 2023 at the Dermatology Department of Unidade Local de Saúde Santa Maria. Patients with histological diagnoses of AFX or PDS were included. **Results:** A total of 22 male patients, all over 65 years old, were included. Most tumors were located on the head (91%), measured < 2 cm (59%), and presented as non-specific plaques, nodules, or tumors, occasionally ulcerated. Histological criteria used to distinguish AFX (73%) from PDS (27%) included adipose tissue invasion (27%), neurotropism (5%), and necrosis (23%). All patients underwent initial surgical excision, five with positive margins. Three PDS patients received adjuvant radiotherapy (RT). One patient had a local recurrence, two patients developed regional metastases, and one patient developed a distant metastasis, all in PDS cases. No disease-related deaths were reported. **Conclusions:** AFX and PDS are rare cutaneous tumors with similar clinical presentations but distinct prognostic outcomes. Immunohistochemistry plays a critical role in differential diagnosis. Surgery remains the gold-standard treatment, while RT may benefit selected PDS cases. Further studies are needed to standardize therapeutic strategies and establish clear follow-up guidelines for these patients.

Keywords: Atypical fibroxanthoma. Pleomorphic dermal sarcoma. Mesenchymal tumors. Immunohistochemistry. Wide surgical excision. Adjuvant radiotherapy.

Resumo

Objetivos: Caracterizar a demografia, manifestações clínicas, alterações histopatológicas, terapêutica instituída e prognóstico de doentes diagnosticados com fibroxantoma atípico (FXA) e sarcoma dérmico pleomórfico (SDP) num centro terciário. **Métodos:** Foi realizado um estudo observacional e retrospetivo entre janeiro 2014 e dezembro de 2023 no serviço de Dermatologia da Unidade Local de Saúde Santa Maria. Foram incluídos doentes com diagnóstico histológico de FXA e SDP. **Resultados:** Foram incluídos 22 indivíduos do sexo masculino, todos com idade superior a 65 anos. A maioria dos tumores localizavam-se na cabeça (91%), tinham uma dimensão inferior a 2cm (59%) e uma apresentação clínica inespecífica (placas,

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nódulos ou tumores, ocasionalmente ulcerados). Os critérios histológicos utilizados para diferenciação histológica entre FXA (73%) e SDP (27%) foram a invasão da hipoderme (27%), neurotropismo (5%) e necrose tumoral (23%). Todos os doentes foram submetidos a excisão cirúrgica, cinco com margens cirúrgicas positivas. Três doentes com SDP foram submetidos a radioterapia adjuvante. Verificou-se um doente com recidiva local, dois doentes com metastização ganglionar e um doente com metastização à distância em doentes com SDP. Não se verificaram mortes relacionadas com a doença. **Conclusões:** FXA e SDP são tumores cutâneos raros com apresentações clínicas semelhantes, no entanto com prognóstico distinto. A imunohistoquímica assume um papel crucial no diagnóstico diferencial. A cirurgia constitui o tratamento de eleição, podendo a radioterapia ser considerada benéfica em casos selecionados de SDP. São necessários estudos adicionais para uniformizar as estratégias terapêuticas e definir diretrizes de seguimento destes doentes.

Palavras-chave: Fibroxantoma atípico. Sarcoma dérmico pleomórfico. Imunohistoquímica. Mesenchymal tumors. Excisão cirúrgica alargada. Radioterapia adjuvante.

Introduction

Atypical fibroxanthoma (AFX) and pleomorphic dermal sarcoma (PDS) are mesenchymal tumors characterized by an unclear line of cellular differentiation[1]. In Portugal, there are no available data on incidence[2]. Historically, their classification was hampered by inconsistent and imprecise terminology. Terms such as superficial variant of malignant fibrous histiocytoma or dermal undifferentiated pleomorphic sarcoma, previously used to describe these tumors, are now considered obsolete[2]. The current consensus is that AFX and PDS represent a continuum of the same entity, as these tumors exhibit similar genetic, morphological, and clinical features[1-4]. However, the presence of subcutaneous invasion or lymphovascular/perineural invasion is not a feature of AFX and confers an increased risk of aggressive behavior, as observed in PDS[1]. Given the rarity of these tumors, there is a limited number of studies in the literature providing their characterization. The aim of this study was to assess the demographic, clinical, histological, therapeutic, and prognostic variables of patients diagnosed with AFX and PDS attending the dermatology department in a tertiary center in Lisbon, Portugal.

Methods

A retrospective, observational study was conducted to characterize the population diagnosed with AFX and PDS between January 2014 and December 2023 at the Dermatology Department of Unidade Local de Saúde de Santa Maria. Data were retrieved from the laboratory data system of the cutaneous histopathology department (Clinidata® software) using the search terms "atypical fibroxanthoma," "pleomorphic dermal sarcoma," "malignant fibrous histiocytoma," and "dermal undifferentiated sarcoma" (n = 38). Patients with incomplete records or whose follow-up was carried out in another hospital were excluded from the study (n = 16). The medical records of identified patients were reviewed to extract demographic information (gender and age at diagnosis), comorbidities, clinical characteristics (tumor location, size, progression time, and clinical diagnosis), histopathological findings (tumor margins, borders, histological patterns, cellular morphology, nuclear pleomorphism, hypodermal invasion, vascular invasion, neurotropism, necrosis, mitotic count, ulceration, and immunohistochemical [IHC] staining), therapeutic approaches, and prognostic outcomes (local recurrence, metastases, and mortality). Data collection and analysis were performed using Microsoft Excel 2021.

Results

This study included 22 patients, whose demographic, clinical, and histopathological features, treatment modalities, and outcomes are summarized in tables 1-3.

Demographic characterization

All patients included in this study were male (100%, n = 22). The mean age at diagnosis was 81 ± 6 years, with ages ranging from 68 to 95 years. Most patients were between 80 and 89 years old (59%, n = 13). Nine patients had a personal history of previous diagnosis and treatment of cutaneous neoplasms, including basal cell carcinoma (BCC) and squamous cell carcinoma (SCC). The number of BCCs per patient ranged from 1 to 5, whereas the number of SCCs ranged from 1 to 14. Only one patient with AFX was immunosuppressed due to follicular non-Hodgkin lymphoma.

Table 1. Demographic information, comorbidities, and clinical characteristics of included patients

Variables	Frequency no. (%)		
	AFX	PDS	Total
Gender			
Male	16 (73)	6 (27)	22 (100)
Female	0	0	0
Age			
60-69	1 (5)	1 (5)	2 (9)
70-79	3 (14)	1 (5)	4 (18)
80-89	10 (45)	3 (14)	13 (59)
90-99	2 (9)	1 (5)	3 (14)
Personal medical history			
Cutaneous neoplasm	8 (37)	1 (5)	9 (41)
BCC*	5 (23)	1 (5)	-
Invasive SCC*	7 (32)	0	-
In situ SCC*	2 (9)	0	-
Immunosuppression	1 (5)	0	1 (5)
Follicular non-Hodgkin lymphoma	1 (5)	0	1 (5)
Tumor location			
Scalp	7 (32)	2 (9)	9 (41)
Malar	4 (18)	1 (5)	5 (23)
Ear	4 (18)	0	4 (18)
Forehead	1 (5)	1 (5)	2 (9)
Hand	0	1 (5)	1 (5)
Shoulder	0	1 (5)	1 (5)
Size			
≤ 2 cm	13 (59)	0	13 (59)
> 2 cm	3 (14)	6 (27)	9 (41)
Mean (± SD) cm	1.5 (± 0.6)	4.5 (± 2.4)	2.4 (± 1.9)
Progression time			
≤ 6 months	6 (27)	4 (18)	10 (45)
> 6 months	1 (5)	1 (5)	2 (9)
Unknown	9 (41)	1 (5)	10 (45)

*Cumulative.
AFX: atypical fibroxanthoma; PDS: pleomorphic dermal sarcoma; BCC: basal cell carcinoma; SCC: squamous cell carcinoma; SD: standard deviation.

Table 2. Histopathological characteristics of tumors included

Variables	Frequency no. (%)		
	AFX	PDS	Total
Encapsulated			
Yes	0	0	0
No	16 (73)	6 (27)	22 (100)
Borders			
Well-defined	7 (32)	1 (5)	8 (36)
Poorly defined	9 (41)	5 (23)	14 (67)
Cell morphology			
Spindle	9 (41)	5 (23)	14 (67)
Epithelioid	4 (18)	0	4 (18)
Both	3 (14)	1 (5)	4 (18)
Nuclear pleomorphism			
Yes	16 (73)	6 (27)	22 (100)
No	0	0	0
Adipose tissue invasion			
Yes	0 (27)	6 (27)	6 (27)
No	16 (73)	0	16 (73)
Vascular invasion			
Yes	0	0	0
No	16 (100)	6 (100)	22 (100)
Neurotropism			
Yes	0	1 (5)	1 (5)
No	16 (73)	5 (23)	21 (95)
Necrosis			
Yes	0	5 (23)	5 (23)
No	16 (73)	1 (5)	17 (77)
Mitosis (per mm^2)			
< 2	0	0	0
2-4	5 (23)	0	5 (23)
> 4	11 (50)	6 (27)	17 (77)
Ulceration			
Yes	1 (5)	4 (18)	5 (23)
No	15 (77)	2 (9)	17 (77)

AFX: atypical fibroxanthoma; PDS: pleomorphic dermal sarcoma.

Clinical characterization

The tumors were predominantly located on the head (91%, n = 20); one case involved the shoulder (5%, n = 1) and another the dorsum of the hand (5%, n = 1). Most tumors measured < 2 cm (59%, n = 13), with sizes ranging from 0.5 cm to 8 cm, and had a progression time of < 6 months before diagnosis. The clinical appearance of these tumors was non-specific, typically presenting as plaques, nodules, or tumors, which may or may not have been ulcerated (Fig. 1). In most cases, the initial clinical diagnosis was SCC. Other differential diagnoses included BCC, melanoma, dermatofibrosarcoma protuberans, and hypertrophic actinic keratosis.

Histological and IHC characterization

All tumors were predominantly localized in the dermis, non-encapsulated, and, in most cases, exhibited poorly defined infiltrative borders (64%, n = 14) (Fig. 2A). Tumor cells displayed variable cytological features, including atypical spindle-shaped (64%, n = 14), epithelioid (18%, n = 4) morphologies, or both (18%, n = 4), characterized by marked nuclear pleomorphism, hyperchromasia, and irregular nuclear contours (Fig. 2B). The most common architectural pattern observed was the storiform arrangement of tumor cells (23%, n = 5) (Fig. 2C). Key distinguishing histological

Table 3. Treatment modalities and outcomes of included patients

Variables	Frequency no. (%)		
	AFX	PDS	Total
Treatment			
Surgery	16 (73)	6 (23)	22 (100)
Re-excision	1 (5)	3 (14)	4 (18)
Radiotherapy	0	3 (14)	3 (14)
Systemic treatment	0	0	0
Outcomes			
Local recurrence	0	1 (5)	1 (5)
Regional metastasis	0	2 (9)	2 (9)
Distant metastasis	0	1 (5)	1 (5)
Mortality			
Disease related	0	0	0
Disease unrelated	6 (27)	3 (14)	9 (41)

AFX: atypical fibroxanthoma; PDS: pleomorphic dermal sarcoma.

features AFX/PDS included adipose tissue invasion (27%, n = 6), absence of vascular invasion, focal neurotropism (5%, n = 1), and tumor necrosis (23%, n = 5). Most cases demonstrated high mitotic activity, with more than 4 atypical mitoses/mm^2 (77%, n = 17). Based on these histopathological findings, 16 tumors were classified as AFX and six as PDS. Due to the overlap in morphological features with other malignancies, including undifferentiated SCC, melanoma, vascular neoplasms, and other sarcomas, IHC analysis was essential to exclude differential diagnosis. CD10 was universally expressed in all tumors tested. Smooth muscle actin (SMA) was tested in 16 cases, with focal positivity in two PDS and four AFX, and vimentin showed diffuse positivity in all three cases tested (Fig. 2D-F). Negative results were obtained for a panel of other immunomarkers, including melanocytic markers, cytokeratins, epidermal proliferation markers, smooth muscle markers, vascular markers, and histiocytic markers (Table 4).

Management and follow-up

Based on initial clinical and imaging evaluation, advanced local invasion was observed in 4 patients (18%), and lymph node involvement in one patient (5%), all of which were histologically classified as PDS. No evidence of distant metastasis was found on initial staging. All tumors were initially treated with surgical excision (margin range: 0.5-2 cm), with positive margins detected in 5 patients (23%), including four PDS and one AFX. Most of these patients underwent re-excision (n = 4) after diagnosis, achieving clear margins in three cases. The patient with nodal involvement died due to an unrelated cause before re-excision (traumatic subarachnoid hemorrhage). Three patients diagnosed with PDS underwent radiotherapy (RT), including a PDS patient with positive margins after re-excision (adjuvant), and two PDS patients with minimal clear margins after re-excision (adjuvant). No patient received systemic treatment. During a median follow-up period of 25 months (range: 4-72 months), local recurrence, regional metastasis, and pulmonary metastasis were observed in one PDS case after 18 months of follow-up (previously submitted to surgery and RT). No disease-related deaths were reported. However, 9 patients (41%) died from unrelated causes.

Figure 1. Non-specific clinical appearance of atypical fibroxanthoma/pleomorphic dermal sarcoma, typically presenting as plaques, tumors (**A** and **B**), or nodules (**C**), which may or may not be ulcerated.

Discussion

AFX and PDS occur predominantly in men (up to 8 times more frequently), elderly individuals (with a peak incidence between 70 and 80 years), and are mainly located in photo-exposed areas, such as the scalp, face, and back of the hands, as observed in this population[2,5]. The diagnosis of AFX and PDS tumors remains a challenge and one of exclusion, because these tumors show no reliable discriminatory clinical,

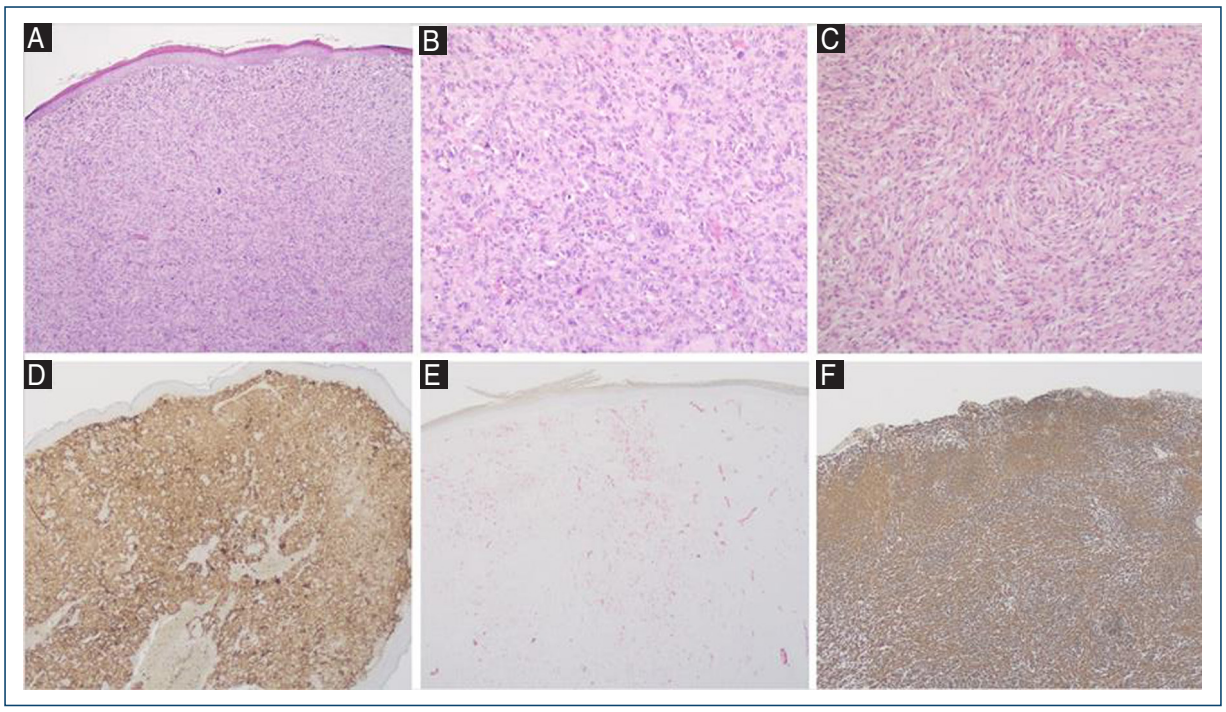

Figure 2. Most common histopathological and immunohistochemical features. **A:** tumor located within the dermis, non-encapsulated, and with poorly defined borders (HE, ×40), **B:** tumor cells displayed various morphologies, including atypical spindle-shaped and epithelioid cells, with nuclear pleomorphism (HE, ×100), **C:** storiform pattern (HE, ×100), **D:** diffuse positivity for immunohistochemical CD10 study (CD10, ×25), **E:** focal positivity immunohistochemical Smooth muscle actin (SMA) (SMA, ×25), **F:** diffuse positivity for immunohistochemical vimentin study (Vimentin, ×25).

Table 4. Immunomarkers tested in this study for differential diagnosis, including melanocytic markers, cytokeratins, muscle markers, vascular markers, and reticulohistiocytic markers

Classes	Immunomarkers
Melanocytic markers	SOX10, S100, Melan-A, PRAME, HMB45
Cytokeratins and epidermal proliferation markers	AE1/AE3, MNF116, CK34βE12, BerEP4, p63, p40, CK7, CK20
Smooth muscle markers	SMA, Desmin
Vascular markers	CD31, CD43, podoplanin
Histiocytic markers	CD34, XIIIa

morphologic, or IHC features[2-4]. The histological differentiation between AFX and PDS carries prognostic significance and relies on the extent of tissue infiltration: AFX is confined to the dermis without involvement of subcutaneous tissue, whereas PDS demonstrates invasion into the subcutaneous adipose tissue, skeletal muscle, or fascia[1]. Additional features such as perineural invasion, vascular invasion, and tumor necrosis further distinguish PDS[1]. Therefore, a deep incisional biopsy is essential for establishing a definitive diagnosis[2,4]. IHC staining plays a crucial role in the differential diagnosis of tumors with overlapping histological features, including melanoma, SCC, and other sarcomas of muscular, vascular, or reticulohistiocytic origin[2,6]. It is recommended to use at least two melanocytic markers, two cytokeratin markers, and one muscle marker[2]. Additional markers, such as vascular markers or other muscle-related markers, may be required[2,6]. Differential diagnosis between AFX/PDS and poorly differentiated SCC can be particularly challenging, with IHC markers such as p63, p40, cytokeratin MNF116, and CK34βE12 proving useful in supporting SCC[7]. CD10 has shown utility as a relatively reliable marker in AFX/PDS, with diffuse positivity observed in all evaluated tumors in this study. However, it lacks specificity and should be interpreted alongside other findings[4,8]. In the literature, a significant proportion of AFX/PDS have been reported to express CD99, procollagen-1, and CD68[2,4,8,9]. Although up to 70% of these tumors present focal positivity for SMA, the cells are negative for desmin, which can help exclude leiomyosarcoma[9,10]. Several studies found no difference in the IHC profile when comparing AFX with PDS[2,4,7,10].

Currently, there are no internationally standardized guidelines for the treatment and follow-up of AFX and PDS, apart from German guidelines and some recommendations reported in the literature[2,4,5,11]. As a result, management is still based on limited evidence. Currently, the surgical approach remains the gold standard treatment, with studies highlighting the benefits of Mohs micrographic surgery for AFX, although no similar evidence exists for PDS[5,11]. Safety margins of at least 0.5-1 cm for AFX and 2-3 cm for PDS are recommended; however, functional and cosmetic aspects should be considered[2,5,12,13]. RT appears to have a role in treating inoperable locally advanced disease or local recurrences, and may be considered in PDS cases without adequate safety margins[2,4,14,15]. In this study, two PDS patients underwent RT due to minimal surgical margins without further recurrence. To date, no systemic therapies have been approved for these tumors, but there are some case reports that PD-1/PD-L1 inhibiting antibodies may be effective[2,16,17]. Locally advanced or metastasized disease should be discussed by an interdisciplinary tumor board[2,4].

Follow-up recommendations for AFX involve examination every 6 months for 2 years, then annually, whereas PDS requires closer monitoring every 3 months for 2 years, then every 6 months for at least 5 years[2]. Surveillance should focus on the surgical site and regional lymph nodes, with ultrasound recommended in PDS[2]. Cross-sectional imaging (positron emission tomography/computed tomography or magnetic resonance imaging) is not routinely indicated, but may be appropriate in cases of clinically suspicious findings, tumors with high-risk features (e.g., positive margins, perineural or vascular invasion, large size), or in the setting of recurrence or metastatic disease[2,18].

In the literature, local recurrence rates of up to 10% for AFX and 28% for PDS have been reported[2,5,12]. Regarding regional and distant metastases, it is theoretically non-existent for AFX, but can reach up to 20% for PDS[2,5,14,19]. A recent nationwide cohort study from Denmark, including 1118 patients, reports 5-year recurrence rates of 10% for AFX and 17% for PDS, with metastatic disease occurring in 16% of PDS cases[14]. Metastases in PDS most frequently involved the lungs and occurred within the first 3 years of follow-up[14]. In our cohort, local recurrence was observed in 1 PDS patient (16%), regional lymph nodes involvement in 2 PDS patients (33%), and distant metastasis in 1 PDS patient (16%), findings consistent with previously reported data. In addition, female sex, immunosuppression, tumor necrosis, and perineural invasion were recently associated with increased risk of recurrence and disease-specific mortality in PDS[20]. In our series, the patient who developed distant metastasis had a large tumor (8 cm) located on the shoulder, with hypodermal invasion, neurotropism, and positive surgical margins after the initial excision, features that likely contributed to adverse outcomes.

Regarding mortality in AFX and PDS, several studies have reported varying outcomes[18,20,21]. A retrospective cohort including 34 patients with AFX and 10 with PDS reported an overall mortality of 60% in PDS compared to 27% in AFX[19]. In our cohort, 50% of PDS patients and 38% of AFX patients died during follow-up, although none of the deaths were disease-related. The advanced mean age (81 years) and comorbidities likely contributed to the high all-cause mortality, while effective local management (complete excision achieved in 86% of cases and adjuvant RT in patients with residual disease) may have limited disease-specific deaths.

Conclusion

AFX and PDS are rare cutaneous neoplasms that remain a diagnostic and therapeutic challenge due to overlapping clinical and histopathological features and limited standardized guidelines[2-4]. Differentiating between AFX and PDS has significant prognostic implications, emphasizing the importance of accurate histopathological evaluation and IHC analysis[1-4]. Surgical excision is the gold standard of treatment, with RT serving as an adjunct in specific cases[2,5]. Despite the rarity of these tumors, further studies are necessary to better characterize their clinical behavior, optimize management strategies, and establish evidence-based guidelines to improve patient outcomes.

Limitations

The limitations of this study include its retrospective nature, single-center design, incomplete medical records, variable nomenclature over the years, and small sample size.

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Conflicts of interest

None.

Ethical considerations

Protection of humans and animals. The authors declare that no experiments involving humans or animals were conducted for this research.

Confidentiality, informed consent, and ethical approval. The authors have followed their institution's confidentiality protocols, obtained informed consent from patients, and received approval from the Ethics Committee. The SAGER guidelines were followed according to the nature of the study.

Declaration on the use of artificial intelligence. The authors declare that no generative artificial intelligence was used in the writing of this manuscript.

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