West Midlands Cancer Alliance

Atypical Fibroxanthoma (AFX) /Pleomorphic Dermal Sarcoma (PDS) Guidelines

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EAG name	Skin Expert Advisory Group (EAG)	
Document Title	West Midlands Cancer Alliance Atypical Fibroxanthoma (AFX) /Pleomorphic Dermal Sarcoma (PDS) Guidelines	
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Document Purpose	The AFX/PDS guidance has been developed to support consistent, high-quality care by addressing gaps in clinical pathways and ensuring equitable access to timely diagnosis and treatment for patients with rare skin cancers.	
Authors	Dr. Agustin Martin-Clavijo - Dermatology Consultant Professor Joseph Hardwicke PhD FRCS (Plast.) - Consultant Plastic & Reconstructive Surgeon Mr. Kamal Bisarya - Plastic Surgeon	
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Approval Signatures:	**Chair Skin EAG** 	**Clinical Director, West Midlands Cancer Alliance** 

Document name:	**Terms of reference for West Midlands Cancer Alliance Expert Advisory Groups**		
Version	**Date**	**Amendment**	**By**
V1.0	06/1/25	Changes made to guidelines table following discussions within rare cancer group	Dr. Agustin Martin-Clavijo
V2.0	31/1/25	Refinements made to guidelines table following discussions within UHB consultant colleagues.	Dr. Agustin Martin-Clavijo
V3.0	27/2/25	Further refinements were made following presentation at skin EAG and feedback from members.	Dr. Agustin Martin-Clavijo
V4.0	17/10/25	Final refinements made to document following discussion within September skin EAG and placed on WMCA branding template	Dr. Agustin Martin-Clavijo / Angela Heathcote

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1. Background

Atypical Fibroxanthoma (AFX) and Pleomorphic Dermal Sarcoma (PDS) are rare skin cancers that tend to happen in the head and neck of older people. The main risk factors are chronic sun exposure and immunosuppression. Although categorised as 2 different diagnoses, they are now considered as part of the same spectrum, with AFX representing the more superficial and less aggressive form, and PDS the more aggressive and deeper invasion type. The incidence of both has been increasing with an estimated incidence of 0.5 cases per 100,000 people.

2. Purpose

The purpose of this document is to provide guidance for members of the Multi-Disciplinary Team (MDT) to support their decision making and management of cases of AFX/PDS.

3. Guidance

These guidelines are for the management of patients with AFX/PDS across the West Midlands. Evidence in the management of these rare tumours is limited. These guidelines try to follow current best practice and have been written as a consensus document between the Expert Reference Group (EAG) membership and the sarcoma MDT. A guideline is not a rigid constraint on clinical practice, but a concept of good practice against which the needs of the individual patient should be considered. It therefore remains the responsibility of the individual clinician to interpret the application of these guidelines, considering local service constraints and the needs and wishes of the patient. It is not intended that these guidelines are applied as rigid clinical protocols.

All AFX/PDS cases should be managed by a specialist skin multi-disciplinary team (SSMDT). The 2019 Sarcoma Service Specification also require that all cases of suspected or proven sarcoma have to be discussed at a sarcoma MDT

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High risk factors

>4cm diameter	Perineural invasion
Deep invasion past subcutis	Incomplete excision
Lymphovascular invasion	Recurrence

Primary treatment -

AFX	PDS low risk	PDS high risk
Excision 5-10mm down to deep fascia/pericraneum	Excision with at least 1cm down to deep fascia/pericraneum	Excision with at least 1cm down to deep fascia/pericranium
Histological clearance of at least 1mm	Histological clearance of at least 1mm	Histological clearance of at least 2mm

- Radiotherapy can be considered on
  - High risk primary
  - Narrow or incomplete excision
  - Recurrent tumours
  - Surgery not possible or not appropriate
- In-transit metastasis- Aim for complete surgical excision with at least 1mm histological margin
- All patients to be discussed by the SSMDT and the sarcoma MDT. This is especially important in cases with extensive disease, lymph node metastasis or distant metastasis where the sarcoma MDT may take the lead

Imaging

AFX	PDS low risk	PDS high risk
Not required	Baseline CT (Head and Neck TAP)	Baseline CT (Head and Neck TAP)
		Consider imaging 6 monthly for2 years, then yearly for 3 years

Follow up

AFX	PDS low risk	PDS high risk
6 monthly for 1 to 3 years or PIFU	Has per very high risk SCC or PIFU	3 monthly for 2 years, then 6 monthly for 3 years

4. References

- https://www.bad.org.uk/pils/atypical-fibroxanthoma/ (accessed 10.12.2024)
- Overview | Sarcoma | Quality standards | NICE (accessed 10.12.2024)
- The incidence of atypical fibroxanthoma and pleomorphic dermal sarcoma in Denmark from 2002 to 2022 O Fruergaard, M Ørholt et al Surgical Oncology, 2024-12-01, Volume 57
- 54853 Trends in Incidence and Mortality of Pleomorphic Dermal Sarcoma: A Retrospective Cohort Study Using SEER Data. Jafry, MustufaPowers, Molly et al. Journal of the American Academy of Dermatology, Volume 91, Issue 3, AB347

5. Resources

These guidelines have been developed by consensus of the EAG membership and the Rare Skin Cancers Task Group along with Dr Agustin Martin-Clavijo -Dermatology Consultant (UHB), Professor Joseph Hardwicke PhD FRCS (Plast.), Consultant Plastic & Reconstructive Surgeon (UHCW), Mr. Kamal Bisarya, Plastic Surgeon, (UHNM) and Niall O'Hara, Plastic surgery NTN registrar (West Midlands deanery).