Mini Review

BIOMEDICAL Journal of Scientific & Technical Research

ISSN: 2574 -1241

DOI: 10.26717/BJSTR.2024.56.008839

Advances in Understanding and Management of Atypical Fibroxanthoma

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ARTICLE INFO

Received: April 19, 2024

Published: April 26, 2024

Citation: Abdel Selim and Ghada Elayat. Advances in Understanding and Management of Atypical Fibroxanthoma. Biomed J Sci & Tech Res 56(2)-2024. BJSTR. MS.ID.008839.

ABSTRACT

Atypical Fibroxanthoma (AFX) presents as a rare but clinically significant spindle cell tumour of the skin, primarily affecting elderly individuals with a history of chronic sun exposure [1]. Recent advancements in diagnostic techniques and therapeutic modalities have enhanced our understanding and management of this neoplasm [2,3]. This review provides a thorough examination of the current knowledge regarding AFX, encompassing epidemiology, pathogenesis, clinical presentation, histopathological characteristics, immunohistochemical findings, molecular pathology, differential diagnosis, treatment options, and prognosis. Additionally, emerging research areas and future directions in the field of AFX are discussed.

Keywords: Atypical Fibroxanthoma; Cutaneous Malignancy; Spindle Cell Tumour; Histopathology; Immunohistochemistry; Molecular Pathology; Treatment Options; Prognosis; Emerging Research

Introduction

Atypical Fibroxanthoma (AFX) is an infrequent, yet diagnostically challenging cutaneous malignancy characterised by spindle to epithelioid cells [4]. Its clinical behaviour ranges from indolent to locally aggressive, necessitating prompt and accurate diagnosis for optimal management [5]. While histopathological assessment remains fundamental, recent molecular studies have shed light on the underlying genetic alterations driving AFX tumorigenesis [6].

Epidemiology

AFX represents approximately 1% of all cutaneous tumours, predominantly affecting elderly individuals with a predilection for sun-exposed areas [7]. Its incidence is rising, likely due to increased awareness and improved diagnostic techniques. Epidemiological studies have also highlighted associations with immunosuppression and previous radiation exposure [8].

Pathogenesis

Chronic sun exposure and immunosuppression are implicated in AFX pathogenesis [9]. Molecular studies have identified genetic alterations, including TP53 mutations, chromosomal abnormalities, PDGFRB rearrangements, and CDKN2A loss, contributing to dysregulated cell proliferation and tumour growth [10,11]. Additionally, dysregulation of signalling pathways such as the PI3K/AKT/mTOR pathway has been implicated in AFX pathogenesis [12].

Clinical Presentation

AFX typically manifests as a solitary, rapidly growing nodule or plaque on sun- damaged skin, with variable clinical features such as erythema, ulceration, and bleeding [13]. Rarely, regional lymph node involvement or distant metastasis may occur. Clinicians should be vigilant in recognising AFX, particularly in patients with a history of chronic sun exposure or immunosuppression [14].

Histopathological Characteristics

Histologically, AFX exhibits a dermal proliferation of spindle to epithelioid cells with marked cytological atypia and brisk stromal inflammation [15]. Immunohistochemical staining aids in diagnosis, with positive staining for vimentin and CD10 [16]. Additional markers, including p16, p53, and Ki-67, may also be used to characterise AFX and assess its proliferative activity [17].

Molecular Pathology

Genetic alterations, including TP53 mutations, chromosomal abnormalities, PDGFRB rearrangements, and CDKN2A loss, have been identified in AFX [18,19]. These molecular findings provide insights into the underlying mechanisms of tumorigenesis and may offer potential therapeutic targets in the future. Furthermore, gene expression profiling studies have revealed distinct molecular signatures associated with AFX subtypes, which may have prognostic implications and guide personalised treatment approaches [20].

Differential Diagnosis

AFX can mimic other spindle cell tumours, necessitating careful histopathological evaluation and immunohistochemical staining to differentiate it from its mimics, such as dermatofibrosarcoma protuberans, spindle cell squamous cell carcinoma, and leiomyosarcoma [21]. Ancillary techniques such as fluorescence in situ hybridisation (FISH) and next- generation sequencing (NGS) may also be utilised to aid in the differential diagnosis of AFX [22].

Treatment Options

Surgical excision with clear margins remains the mainstay of treatment for AFX, with Mohs micrographic surgery offering precise tumour removal and preservation of surrounding healthy tissue [23]. Adjuvant therapies, including radiation therapy and topical imiquimod, may be considered in select cases, particularly in unresectable or recurrent lesions [24]. Emerging targeted therapies, such as tyrosine kinase inhibitors and immunotherapy, hold promise for the management of advanced or metastatic AFX and warrant further investigation in clinical trials [25].

Prognosis

AFX generally carries a favourable prognosis, with low rates of metastasis and disease- specific mortality [26]. However, local recurrence rates vary depending on factors such as tumour size, depth of invasion, and adequacy of surgical excision. Long-term follow-up is recommended to monitor for recurrence and assess treatment outcomes [27].

Conclusion

Atypical Fibroxanthoma is a rare but clinically significant cutaneous malignancy characterised by spindle to epithelioid cells [28]. Recent advancements in molecular pathology have improved our understanding of its pathogenesis and may facilitate the development of targeted therapeutic strategies [29]. Further research is warranted to elucidate the molecular mechanisms underlying AFX tumorigenesis and to optimise treatment approaches for this challenging neoplasm.

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ISSN: 2574-1241

DOI: 10.26717/BJSTR.2024.56.008839

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