Case report

An extremely rare case of atypical fibroxanthoma – case report

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ABSTRACT

The first clinical case of atypical fibroxanthoma was documented in 1958, and to date, approximately 3,000 patients have been described in the literature with this diagnosis. Furthermore, the recurrence rate or potential for metastasis in atypical fibroxanthoma is estimated to be as low as 2%. It is noteworthy that fibroxanthoma most commonly arises in the head and neck region. Therefore, the clinical case presented here of a patient diagnosed with atypical fibroxanthoma on the left earlobe margin, with suspected disease recurrence after approximately one year, is extremely rare, so we believed it is noticeable to report.

Key words: atypical fibroxanthoma, immunohistochemistry, head and neck neoplasms, neoplasms, therapeutic approach, case report

INTRODUCTION

Atypical fibroxanthoma (AFX) is a rare, primary, dermal neoplasm of uncertain histogenesis, typically arising from sun-damaged skin on the heads and necks of elderly individuals. Since its initial description in 1958, approximately 3,000 cases have been reported in the literature. AFX is characterized by a low degree of malignancy, with rare recurrences and a metastatic potential of less than 2% [1]. Diagnosing AFX involves a combination of clinical, histopathological, and immunohistochemical evaluations. Clinically, AFX often presents [2] as a rapidly growing, solitary nodule with variable coloration, including flesh-colored, pink, or erythematous hues [3]. AFX most commonly arises on the skin of the head and neck, predominantly affecting elderly patients [2]. Histopathologically, the tumor is characterized by the presence of spindle or epithelioid cells with marked pleomorphism, including prominent nucleoli, and is accompanied by a mixed inflammatory infiltrate [4]. Epithelioid cells tend to adopt a more rounded cellular morphology, a characteristic also observed in certain cases of melanoma. Immunohistochemical staining, particularly for markers such as CD68 and vimentin, is widely utilized in the diagnosis of AFX [5]. The challenge in diagnosing AFX lies in its resemblance to other cutaneous lesions and skin cancers (e.g., basal cell carcinoma, squamous cell carcinoma). Consequently, a definitive diagnosis can only be established through histopathological examination. The treatment of choice is complete surgical excision, which should involve wide local excision with a 1 cm margin. In cosmetically significant regions, where tissue preservation is crucial, Mohs micrographic surgery should be considered [6]. Due to the high incidence of other malignant skin tumors in patients with AFX, regular monitoring is recommended [7]. The prognosis for patients with AFX is generally excellent, with a median 20-year survival rate of 97.8% [8]. Factors contributing to the development of AFX include genetic factors, skin trauma, radiotherapy, diabetes, viruses (particularly HIV), as well as organ transplantation and associated immunosuppression [9, 10].

AIM OF THE ARTICLE

This article presents AFX as an exceedingly rare diagnosis. The case report aims to underscore the importance of oncologic vigilance and the necessity for regular, long-term patient monitoring.

MATERIAL AND METHODS

A retrospective review of patient management and observation was undertaken based on the analysis of medical records, interviews, physical examinations, and patient observations during his hospital stay.

CASE REPORT

PATIENT INFORMATION

The patient, aged 73, of Caucasian race, with Fitzpatrick skin type III, presents with a medical history of arterial hypertension, hypercholesterolemia, and nicotine addiction (approximately 30 cigarettes per day for 50 years). Past medical history includes a myocardial infarction years ago, which necessitated percutaneous coronary intervention, as well as an appendectomy and inguinal hernia repair surgery. The patient does not report a family history of cancer, particularly skin cancer, and denies any allergies or sensitivities. He works as a carpenter and occasionally engages in hunting. He reports spending most of his life outdoors, often exposed to significant amounts of sunlight without adequate protection.

COURSE OF DISEASE

The first concerning symptoms appeared around June 2022. The patient noticed a small, flesh-colored thickening on the margin of the left earlobe. This thickening rapidly increased in size, accompanied by ear pain rated by the patient as 6 on the Visual Analog Scale (VAS). The pain was stabbing in nature, palpably tender, and woke the patient at night. Over time, the lesion began intermittently oozing blood. The patient decided to consult their primary care physician, who subsequently referred them to the otolaryngology clinic. Six months later, in January 2023, the patient presented to the otolaryngology clinic. The tumor measured 1.5 cm in diameter, was relatively soft, non-mobile, with visible ulceration in the upper part, intermittently oozing blood, and accompanied by pain. The physical examination of the head and neck revealed no abnormalities, and no enlarged lymph nodes were detected. The patient was referred to the hospital's otolaryngology department for surgical treatment and further diagnostic evaluation. Physical examination and laboratory tests did not reveal any abnormalities. The following day, under general anesthesia, a radical resection of the left external ear tumor with clear tissue margins, including the auricular cartilage, was performed. To reconstruct the tissue defect, skin grafts were harvested from the left clavicular region. The excised tissue was sent for histopathological examination. After 3 days, the patient was discharged home in good general condition, with appropriate recommendations.

Following the histopathological examination, the diagnosis revealed AFX. The immunophenotype of the lesion cells showed CKPAN-, S-100, SOX10-, CD10+, CD 68+, with a Ki67 proliferative index of approximately 50% of cells. The macroscopic description noted that "the tumor appears not to invade the cartilage of the earlobe".

Six months later, during a follow-up visit, the ear was noted to have healed properly with no signs of recurrence and no palpable lymph nodes. However, there was alarming thickening in the left supraclavicular area, prompting a referral for ultrasound examination of the lymph nodes. The ultrasound, however, did not reveal any abnormalities.

In January 2024 patient showed up at the otolaryngology clinic with a new skin lesion – forward to previous excision site on the margin of the left earlobe, a new lesion approximately 6 mm in size had developed, accompanied by serous discharge upon compression and ulceration in the central part. Periodic bleeding was noticeable, and the lesion itself was relatively soft and non-mobile. There was mild tenderness upon palpation and when lying on the ear. Suspecting a second focus of AFX, the patient was referred to the otolaryngology department for excision of the lesion and histopathological diagnosis.

The procedure was performed under local anesthesia, and the excised tissue was sent for histopathological examination. The procedure was uneventful, and the patient was discharged home in good general condition. The histopathological examination did not confirm recurrence of AFX.

During a follow-up visit in June 2024, the patient palpated another small lesion on the posterior aspect of the left ear, distant from the initial excision site. Given the absence of accompanying symptoms, a watchful waiting approach with appropriate oncologic vigilance was adopted.

The patient remains under the continuous care of the otolaryngology clinic.

DISCUSSION

The definitive diagnosis of AFX requires a combination of clinical assessment, histopathological examination, and immunohistochemical analysis [2]. Currently, the only diagnostic option available is exclusionary diagnosis through histopathological examination. Regarding therapeutic methods, there are 2 surgical solutions: complete surgical excision of the lesion with an appropriate safety margin of 1 cm. In cosmetic-sensitive areas, Mohs surgery should be considered [6]. According to Koch et al. the prognosis for patients with AFX is notably favorable, with a reported 20-year median survival rate of 97.8% [8]. However, patients should be informed about the increased risk of developing other malignant skin tumors [7] and the importance of avoiding risk factors such as viral infections, especially HIV, skin trauma, and prolonged inadequate sun protection [9, 10].

Study limitations: The study was confined to a single patient. In the future, it would be beneficial to expand the research to include additional case studies, allowing for comparisons of diverse symptoms, lesion locations, improvement of diagnostic and therapeutic pathways, as well as investigating the impact of sun protection use on the incidence of AFX.

CONCLUSION

Diagnosis of AFX primarily relies on exclusionary criteria, as there are no specific tumor markers for AFX. With appropriate treatment, AFX carries an excellent prognosis. Surgical excision of the tumor remains the treatment of choice. Following AFX excision, regular long-term patient monitoring is recommended due to the heightened risk of developing other malignant skin tumors [10]. It is crucial to initiate an informative discussion with the patient regarding the meticulous care of sun-exposed skin, emphasizing the use of SPF 50+ sunscreen and the necessity for regular reapplication throughout the day.

PATIENT PERSPECTIVE

From the patient's perspective, the most important aspect was achieving relief from suffering. Currently, the patient diligently cares for sun-exposed skin by using SPF 50+ sunscreen and remembering to reapply it. He have been educated about the course of the disease, its potential recurrence, and are aware of prodromal symptoms, maintaining vigilance. However, he is knowledgeable about the benign nature of the condition and its very high survival rate. The patient remains optimistic and resilient, stating that he have learned to live with it.

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