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Atypical Fibroxanthoma in Young Omani Teenager a Rare Presentation Case Report and Literature Review

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Abstract Cutaneous Atypical fibroxanthoma (AFX) typically occurs on the head and neck of sun-damaged areas in older Caucasians & it's a diagnosis by exclusion of other malignant neoplasms with similar histopathology or morphology. In this case report and literature review, we report a much less common presentation as the first case to our knowledge of AFX on a teenage female with darker skin, this lesion on the dominant hand that needed re-excision in order to get a clear margin. A high index of suspicion of this less common type in a younger patient presenting with a cutaneous nodule is to be kept in mind to minimize the number of excisions and increase patient's satisfaction.

Keywords: *Atypical fibroxanthoma, Skin neoplasms, young patient*

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1. Introduction

Atypical fibroxanthoma (AFX) is a rare cutaneous disease that is associated with aging and sun-damaged area, typically occurring on the head and neck in white persons. A much less common appearance is in younger patients, where it is found in the extremities and trunk [1,2]. The differential diagnosis of AFX involves SCC, BCC, Merkel cell carcinoma, and amelanotic melanoma. The pathological finding atypical spindle-shaped cells with moderate amounts of cytoplasm and large atypical cells with abundant pale-staining vacuolated cytoplasm. A diagnosis of AFX is made mostly by exclusion. AFX is cured by complete surgical excision and it has a very low incidence of recurrence and is rare to metastasize.

2. Case Report

A 17-year-old healthy right-hand dominant female presented with swelling, on the dorsum of the right hand that had been noted by her for the preceding 3-4 weeks. The lesion had arisen spontaneously and enlarged rapidly. The patient denied any history of trauma and had no previous radiation treatment or other significant past medical history. No Family history of any skin cancer or any similar presentation. On examination, she has Fitzpatrick skin type V and had a mildly tender 1x1 cm well-defined oval-shaped subcutaneous swelling on the base of the second web space of the dorsum of the right hand (Figure 1). The range of motion of the fingers and hand strength were normal. MRI with contrast examination revealed a superficial subcutaneous well-defined mass on the dorsal aspect between 2nd and 3rd MCP joints with features likely to be peripheral nerve sheath tumor: schwannoma, ganglioneuroma, or, post-traumatic neuroma and advice histopathologic correlation (Figure 2, Figure 3). Excisional biopsy was done and the histopathological examination showed skin underlying dermal lesion. The lesion is centered at the dermis with a grenz zone of the uninvolved dermis. It is formed of the spindle to round and epithelioid tumor cells in haphazard and fascicular pattern. Multinucleated giant cells including tuton giant cells are noted (Figure 4, Figure 5). The lesion is entrapping collagen fibers and extending into subcutaneous tissue. Mitotic figures are seen. There is no necrosis. The tumor cells are noted at the margin.

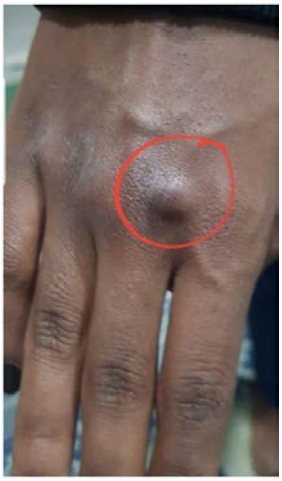

Figure 1. Subcutaneous nodule right hand. Patient consent was obtained

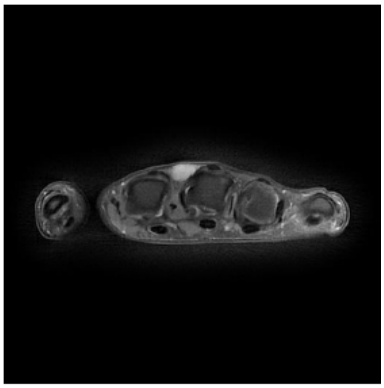

Figure 2. MRI with contrast: superficial subcutaneous well defined mass on the dorsal aspect between 2nd and 3rd MCP joints

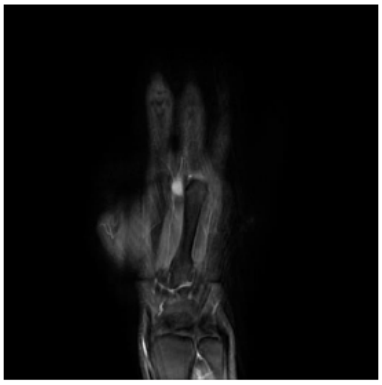

Figure 3. MRI with contrast: superficial subcutaneous well defined mass on the dorsal aspect between 2nd and 3rd MCP joints

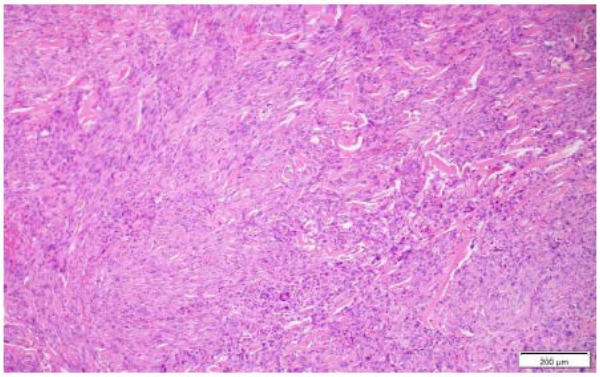

Figure 4. Hematoxylin and Eosin staining. (X100 magnification: cellular bundles and fascicles composed of atypical spindled cells with scattered multinucleated giant cells)

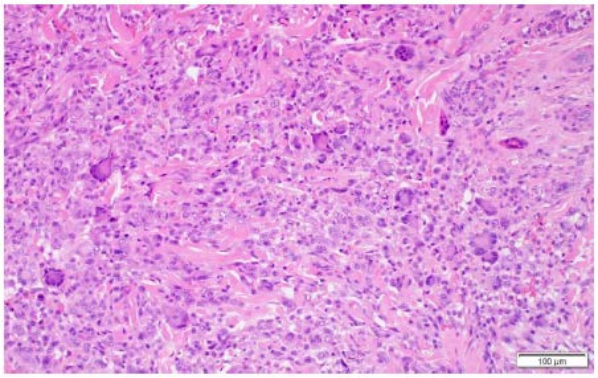

Figure 5. Hematoxylin and Eosin staining (X200 magnification: multinucleated giant cells, tuton giant cells)

The tumor cells are positive for CD10, CD99, Vimentin and focally positive for SMA. They are negative for CD68, S100, Desmin, Myogenin, MyoD1, AE1/3, BCL2, VWF, CD34, EMA and GFAP.

A diagnosis of Atypical Fibroxanthoma was made.

As the tumor cells are noted at the surgical margin and due to un-availably of Mohs's surgery in our center the patient underwent excision for 2nd time and the histology slides sent abroad for an expert opinion and a final report shows a cellular spindle cell neoplasm with admixed multinucleate giant cell and entrapment of hyaline collagen bundles, thus resembling cellular FH- except for the fact that there are multifocal scattered, mainly mononuclear cells with enlarged irregular atypical vesicular nuclei and more copious amphophilic cytoplasm. The tumor involves the excision margin and was advised to undertake further re-excision. Re-excision with a wider margin under frozen section was done and the final histopathology confirmed a clear margin. She was kept on regular follow-up and referred for hand physiotherapy. She was seen at 6 months follow-up post-surgery there was no clinical evidence for recurrence, the scar was well healed, and the patient had full range of motion of fingers. She is on regular 6 monthly follow-ups and the last follow up was two years post-surgery with a well-healed scar and normal hand function (Figure 6, Figure 7).

Figure 6. The scar and range of motion at two years post-surgery. Patient consent was obtained

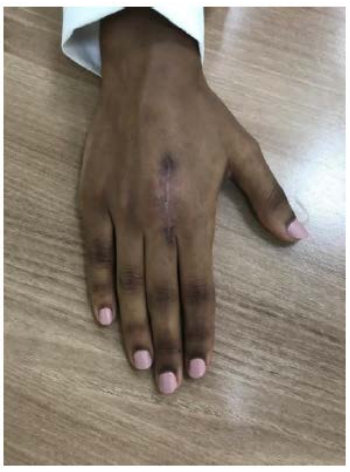

Figure 7. The scar and range of motion at two years post-surgery. Patient consent was obtained

3. Discussion

Atypical fibroxanthoma is uncommon, a low-grade sarcoma, rapid-growing, dermal-based mesenchymal neoplasm. It was first described by Helwig in 1963. [3] It is arising from myofibroblast or fibroblast-like cells that commonly affects the head and neck region of person usually older than 50-year-old and shows a male predominance. Males were more frequently affected than females, statistically significant only for patients with lesions on the head and neck [1]. The lesion usually presents as asymptomatic, painless, and appears as a rapidly enlarging reddish, dome-shaped nodule, often eroded ulcerated with the crusted surface.

Around 25% of the time however AFX can occur on normal-appearing skin of the trunk, arms, or legs of a person in their 30s or 40s [2,4,5]. AFX of unexposed areas of the body has similar histologic features and biological attitudes to atypical fibroxanthoma. In addition, it has been reported that the closely linked of ultraviolet damage in inducing AFX and involves with p53 mutation [4,6].

A case report of atypical fibroxanthoma in a young woman, propositioning that increase of AFX in young patients particularly in those without radiation damage history, may be due to increased tanned skin as a sign of beauty and has encouraged increased exposure, particularly in the younger population [4]. The case presentation was in New York, USA, and it was about a 21-year-old female who presented with a three-month history of an enlarging left nasal alar nodule and was diagnosed with AFX.

Moreover, case presentation in the Department of Pathology, In New Delhi, India, in 2016. The case was about a 24-year-old female who presented with a nodular lesion on her left leg for the last two months. The lesion was managed with surgical excision. It points out that AFX has been proposed to be a reactive or reparative process developing in response to chronic skin injury stimulated by solar irradiation. Moreover, some reports of the appearance of AFX in patients with internal malignancy indicators for the host defective immune response [7].

On the other hand, a study reported a 28-year-old female who had received irradiation as acne therapy 10 years previously to the diagnosis of facial AFX [8]. Another study reported of Li-Fraumeni syndrome in an 18-year-old female who had been diagnosed with AFX on her left arm [9]. A case reported a 23-year-old woman with SLE and Sjogren's syndrome who had an AFX on her right shin, and she had used an immunosuppressant [5]. These patients, however, had either a history of irradiation or genodermatosis or use immunosuppressants, are absent in our patient, which she was totally healthy with no history of radiation exposure or genetic disorder.

A case was reported in Texas a 70-year-old African American woman with blue eyes who had an AFX on her nose. The patient had no history of skin cancer, radiation exposure, or immunosuppression. It has been suggested that pale eyes may be associated with a greater risk of certain cutaneous malignant neoplasms. However, AFX is not a common effect in people with dark skin [10].

However, a case report of a very young patient diagnosed with AFX, it reported an 11-year-old boy who had no significant medical history, he presented with a lesion on the left neck for three months. Immunohistochemical study show positive for CD10, CD68, and Vimentin, but not S-100, cytokeratin, smooth muscle actin, or CD31. He was diagnosed with atypical fibroxanthoma [11]. Another case was reported of xeroderma pigmentosum (XP) type C in a 13-year-old girl who had been diagnosed with AFX on her left index finger [12].

The proper diagnosis of this neoplasm is vital to avoiding radical excision. Differential diagnoses are squamous cell carcinoma, basal cell carcinoma, malignant fibrous histiocytoma (MFH), and melanoma. These neoplasms have a similar clinical presentation to AFX, each of these skin neoplasms may have a component of or even a predominance of spindle cells and thus be confused histopathologically [4,13].

Our patient differs from the above reported cases in that she is very young, 17 years old, and has no history of radiation or skin tanning. Moreover, she is Fitzpatrick skin type V and populations with darker skin, though not entirely exempt, have a lower vulnerability to the harmful effects of UV light and thus to UV-induced neoplasia [10].

Atypical fibroxanthoma tumors usually have a benign clinical course. Surgical excision is the gold standard of treatment in all age groups who are diagnosed with AFX, either with Mohs' micrographic surgery (MMS) or wide local excision (WLE) with 1-2cm safety margins is recommended [14,15]. However, Mohs' micrographic surgery may result in a better outcome than wide local excision, in fewer recurrences and smaller defects [16]. Radiotherapy is not usually recommended as initial therapy, however, most often used during cases of reoccurrence [17].

We have been following the patient after excision Four times and no recurrence has been observed till, now and she is on regular six monthly follow-ups.

In conclusion; atypical fibroxanthoma (AFX) is to be kept as a possible differential diagnosis of a lump in the extremity of a young patient to minimize surgical excision and increases patient satisfaction. Further studies are needed to look into the etiology of the AFX in the younger population.

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