# Acanthoma of Murray William following pemphigus foliaceous, uncommon feature in a common disease

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A 53 year old male presented with multiple small blisters over face and trunk for 5 months duration. On examination, multiple superficial vesiculobullous lesions of size 1-4 centimeters were present. Lesions ruptured spontaneously and resolved with crusting and hyperpigmentation with raised irregular surface. Patient was clinically diagnosed as pemphigus foliaceous. Tzanck smear showed multiple acantholytic cells and the patient was treated with Dexamethasone Cyclophosphamide Pulse (DCP) therapy. After the completion, histopathological examination of the healed lesion revealed histological features of "Acanthoma of Murray Williams". Keywords: Acanthoma of Murray Williams; Pemphigus foliaceous.

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## **INTRODUCTION**

Eruptive acanthoma or 'Murray Williams wart' had been reported following resolution of inflammatory dermatosis.<sup>1</sup> In 1956, Williams reported the appearance of these acanthoma after healing of various eczemas.<sup>2</sup> The disease is primarily seen in older age group although cases had been reported in a 10 year old girl following sun burn reaction<sup>3,4</sup> and in a 35 year old male following contact allergic dermatitis to air borne allergens.<sup>5</sup> The disease tends to resolve in a few months.

# **CASE REPORT**

A 53 year old male, fisherman, presented with multiple fluid filled lesions all over the body for the past 5 months, which are insidious in onset, started as a small peanut sized lesion over the right side of abdomen and slowly progressed to involve the chest, back, scalp, face and arms in a span of 2-3 weeks. Lesions were associated with occasional itching. Lesions ruptured spontaneously and due to scratching and healed without scarring in a span of 1 week. The fluid from the lesions was clear, nonfoul smelling and not blood stained. The raw areas were associated with mild pain and burning sensation. There was history of increase in size of raw areas on trauma. There was no history of involvement of oral cavity. Patient was a chronic smoker and alcoholic for the past 30 years. Cutaneous examination showed multiple, erythematous to hyperpigmented erosions with crusting, size varying from 1-4 cm in diameter present over chest, abdomen, axilla, both upper limbs and back [Fig. 1]. No vesicle or bulla was present. Nikolsky's sign was positive, which was elicited at lower back. Pear sign, Bulla spread sign and Sheklakov sign could not be elicited. Multiple, ill-defined, post inflammatory hyperpigmented patches varying in size from 1-4 cm were seen over chest, abdomen and back [Fig. 2]. Scalp showed few erosions measuring 2-4 cm in diameter with mild crusting present over vertex and occipital region. There was no scarring alopecia. Diffuse greying of hair was observed. Similar skin lesions were present over right side of beard region. Oral cavity, nails and genitalia were normal. General examination, systemic examination and routine investigations were within normal limits.

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Figure 1

Figure 2



Figure 3

Figure 4A

Figure 4B



Legend

Figure 1: Multiple hyperpigmented plaques with erosions with crusting.

Figure 2: Multiple, ill-defined, post inflammatory hyperpigmented patches present over back.

Figure 3: Tzanck smear exhibiting acantholytic cells.

Figure 4A: [10x] Epidermis showing varying degree of pleomorphism and dysplasia.

Figure 4B: [10x] Epidermis showing bullae and dysplasic cells.

Figure 5: [40x] reduction in size of bullae and pigmentation in the basal layer.

Figure 6A: Multiple post-inflammatory hyperpigmented patches with few Acanthoma of Murray Williams over chest region.

**Figure 6B:** Closer view of Murray Williams Acanthoma – Hyperpigmented, hyperkeratotic, crusted plaques present over multiple postinflammatory hyperpigmented patches.

Figure 7: Shows mild hyperkeraosis, acanthosis, pigmentary incontinence.

On the basis of clinical features, patient was diagnosed to have pemphigus foliaceous. Tzanck smear showed acantholytic cells [Fig-3]. He was started on pulse therapy with dexamethasone and cyclophosphamide. After one month, the lesions healed with hyperpigmented patches and hypertrophic plaques with crusting [Figs. 3 and 4]. There was no erosion. Skin biopsy done from the plaque revealed hypertrophic epidermis, where squamous cells exhibited marked degree of dysplastic changes and invasion into the subepidermal zone up to the dermis. Invaded area showed dysplastic changes with mitotic figures and varying degrees of pleomorphism. Cells exhibited acantholytic changes. Inflammatory cell infiltrates were seen surrounding the tumor nodules. After next cycle of chemotherapy, the lesion showed reduction in the size of the bulla and increased pigmentation in the basal layer. A repeat biopsy done after 2 months exhibit resolution both clinically and pathologically [Figs. 6 and 7]. The features were highly suggestive of "Acanthoma of Murray William". Patient was treated with intravenous dexamethasone cyclophosphamide pulse therapy. Other additional treatments like nutritional supplements were given. Intralesional steroid injections and topical tacrolimus for acanthoma lesions were tried. Patient completed seven pulses till date and he was in complete remission.

## **DISCUSSION**

The episode of pemphigus foliaceous followed by eruption of typical hyperpigmented plaques having characteristic histopathology is in favour of Murray William's acanthoma. Perhaps the inflammatory dermatosis such as pemphigus foliaceous acts a stimulus to epidermal proliferation with resultant production of these acanthomas only at certain areas of skin<sup>5</sup>. Acanthoma is a generic name for a group of benign tumors of epidermal keratinocytes, with their unifying characteristics that include: a benign behavior, epidermal hyperplasia and lack of dysplasia. Solar keratosis or Bowen's disease would not be considered as members of this group<sup>6</sup>. Epidermolytic hyperkeratosis [EH] can appear in various clinical forms'. EH is a benign acquired tumour of the epidermis which usually appears on the back in middle-aged patients, which takes the form of numerous flat, discrete, grevish-brown papules of 2-6mm in diameter. In 1970, Shapiro and Baraf<sup>8</sup> described the first six patients with Isolated Epidermolytic Acanthoma (IEA) and a seventh with multiple lesions on the scrotum. Subsequently two further cases were described of patients with lesions on the abdomen<sup>9,10</sup>. The term Disseminated Epidermolytic Acanthoma [DEA] was first coined by Hirone and Fukushiro<sup>11</sup> in a patient with multiple lesions on the trunk, upper limbs and shoulders. Since then, six further cases have been described<sup>12-16</sup>. Electron microscopy, in DEA and in most cases of EH, demonstrates that the desmosomes are conserved and the mechanism of blister formation is cytolysis rather than acantholysis<sup>17</sup>. Disseminated Epidermolytic Acanthoma should also be distinguished from other benign acanthomas such as acantholytic acanthoma<sup>18</sup>. In this skin tumour, the characteristic histological abnormality is the presence of acantholysis resulting from a rupture of the desmosomes. None of these findings occurs with DEA<sup>19</sup>. The mechanisms involved in the development of DEA are not fully understood. It is known that under normal conditions, the basal cells of the epidermis synthesize keratins K5 and K14 and on some occasions K15 and K17. When the epidermis begins to be stratified and differentiated, the suprabasal keratinocytes begin to

synthesize K1 and K10<sup>20</sup>. In processes involving EH, patients have a mutation in the genes responsible for the synthesis of K1 and  $K10^{21}$  in the suprabasal cells<sup>22</sup>. It could be that these alterations in K1 and K10 have a hereditary basis in some disorders which involve EH, but in acquired forms such as IEA or DEA they may be induced by an exogenous factor, such as ultraviolet light or viruses. We do not know if DEA may represent some postzygotic/mosaic abnormality of K1 or K10 or whether trauma is involved in these skin lesions. Also tight clothes at the waistline and small-unnoticed traumas on the thorax have triggered DEA<sup>23</sup>. Acantholytic acanthoma presents as a solitary asymptomatic keratotic papule or nodule with occasional crusting ranging from 0.5 to 1.5 cm in size. A truncal predilection is observed with palms, soles, face and mucous membranes usually spared. Older patients are generally affected, age ranging from 32 to 87 years with median age of 60 years with Male: Female ratio of 2:1. Histologically, acantholytic acanthoma shows hyperkeratosis, papillomatosis and acanthosis. Acantholysis is prominent in all lesions, most often involving multiple level of the epidermis and closely resembles that seen in acantholytic dermatoses such as pemphigus, Hailey-Hailey disease, Grover's disease and Darier's disease. Acantholysis distinguishes acantholytic acanthoma from other types of acanthoma $^{24}$ . The following differential diagnoses are considered: viral wart, seborrhoeic keratosis, cutaneous fibroepithelioma, Bowenoid papulosis and epidermolytic acanthoma.

## **CONCLUSION**

In inflammatory dermatosis like pemphigus, lesions may heal with corticosteroids but to produce hyperkeratotic rough plaques, which has been described as "Acanthoma of Murray Williams" which is a rare entity and hence we report this case as it is an uncommon feature of a common disease.

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