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Morphea-A Case Report

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Abstract:
Introduction: *Skin can be the site of manifestations of many systemic diseases. Morphea which is also includes thickening & hardening of skin & sub cutaneous tissues may have a varying range of manifestations. Surgery is rarely indicated.*
Case Report: *A 30 year old female came with complaints of dark brown sclerotic plaque over the upper half of the right thigh with restriction of movements at the right hip joint. X-Ray of the pelvis was suggestive of mottled calcifications & biopsy proved the diagnosis of morphea with calcinosis circumscripta. Excision of the lesion with split skin grafting was done.*
Conclusion: *Morphea is a rare disease & surgical intervention results in successful outcome when indicated*
Keywords: *Morphea, calcinosis, excision*

I. Introduction

Skin, the largest organ of the body, is often neglected by the general surgeon. Besides being site of primary tumours, it can also be site of manifestation of many systemic diseases. Morphea also known as localized scleroderma, is thickening and hardening of the skin and subcutaneous tissue due to excessive collagen deposition. It may have range of manifestations varying from a very small plaque involving only the skin, to disease causing functional and cosmetic deformities. Calcinosis within the involved area is rare and surgical intervention to overcome consequent functional loss even rarer.

II. Case Report

A 30 year old female patient presented with complaints of dark brown plaque over upper half of anterolateral aspect of right thigh of eleven months duration. It had gradually progressed and resulted in painful restriction of movements of right hip joint, especially while squatting particularly during act of defecation. Dermatological examination revealed a solitary, 25cm X 12cm size dark brown irregular sclerotic plaque over anterolateral aspect of upper half of right thigh [Figure 1]. Skin was shiny, alopecic and could not be pinched. There were no signs of inflammation or induration of edge. All movements of right hip joint were restricted at extremes. RA factor was negative and ANA values were within normal limits. X-ray of pelvis including both hips showed mottled calcifications along anterolateral aspect of upper thigh and hip with loss of adjacent soft tissue compared with normal side [Figure 2]. Incisional biopsy was reported as morphea with calcinosis circumscripta [Figure 3 & 4] MRI of right hip region showed a well defined altered signal intensity hypointense lesion in lateral aspect of right thigh which was hyperintense on T2W1 and PD and hypointense T1W1. It was involving predominantly skin and subcutaneous tissue showing marked atrophy, whereas muscles and osseous structures appeared normal. Excision of the lesion with split skin grafting was done [Figure 5, 6, 7, 8]

III. Discussion

Morphea is an uncommon condition affecting 0.2 to 0.04 in 100,000 people and has a female to male ratio of 3:1[1,2]. Morphea is classified into five subtypes: plaque morphea, generalized morphea, linear scleroderma, bullous morphea and deep morphea. Most common variety is plaque type. Our case was of plaque variety. Morphea can present as an innocuous looking very small plaque only involving skin to widespread disease causing functional and cosmetic deformities[3]. Our case belonged to the second category and sought consultation so as to be relieved of the difficulty experienced while squatting for daily chores especially at defecation. Morphea is usually asymptomatic. The first signs of the disease are reddish patches of skin that thicken into firm, oval-shaped areas [4]. In most cases, sclerosis involutes spontaneously over time (typically 3 to 6 years); with residual discoloration and in rare cases, muscle damage [5]. Though the aetiology is obscure, it is postulated that it results from a T-cell mediated fibroblast activation and endothelial injury. Case reports of morphea co-existing with other systemic autoimmune diseases such as primary biliary cirrhosis, vitiligo, and

systemic lupus erythematosus lend support to morphea being an autoimmune disease [6]. Our case did not show any such association. All forms of morphea occur due to the common mechanism of overproduction of collagen by fibroblasts in the affected tissues. Major use of histopathology is in planning treatment as it indicates disease activity and depth of involvement. Biopsy is taken from inflammatory/indurated edge or from sclerotic centre. Laboratory has a limited role in diagnosis of morphea. Increased ESR, eosinophilia, altered CRP levels, hypergammaglobinaemia, autoantibodies like antinuclear antibodies and positive rheumatoid factor may be detected and also indicate disease activity.In our case, x-ray of pelvis with both hips AP view was done which showed mottled calcifications along anterolateral aspect of upper thigh and hip with loss of adjacent soft tissue compared with normal contralateral side. Ultrasonography as a tool for diagnosing, differentiating and monitoring disease activity has been reported in various studies [7]. It can measure skin thickness, which correlates with disease activity. MRI is useful specially to evaluate depth of involvement and disease activity [8]. In our case it was involving predominantly skin and subcutaneous tissue. Diagnosis of morphea is usually made by clinical examination. Immunohistochemistry studies may show areas of intercollagenous staining for connective tissue antigens in thereticular layer of dermis [9]. No antibodies were detected in the skin lesion by immunofluoroscence in our case. Plaque type of morphea usually regresses spontaneously over a period of 3 to 6 years, leaving only a hyperpigmented area or some amount of functional disability if crossing joint line. Evidenced based recommendation for treatment of morphea indicate that limited plaque morphea in active phase should be treated with topical agents, other subtypes need systemic therapy [10]. Our case was complicated because of significant dermal calcinosis. To enable patient to perform daily chores of her life without discomfort, a single procedure consisting of excision of the skin lesion (plaque) and resurfacing it with split skin graft was performed. The patient during the follow up period of over an year, has maintained functional improvement without recurrence or appearance of any new lesion.

IV. Figures And Tables

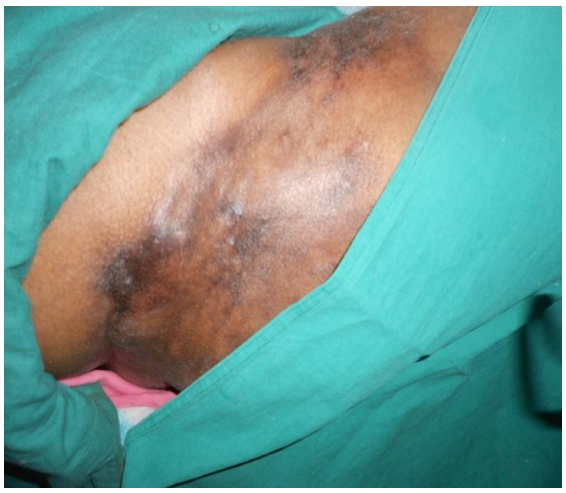

Figure 1: Plaque on anterolateral aspect of right thigh.

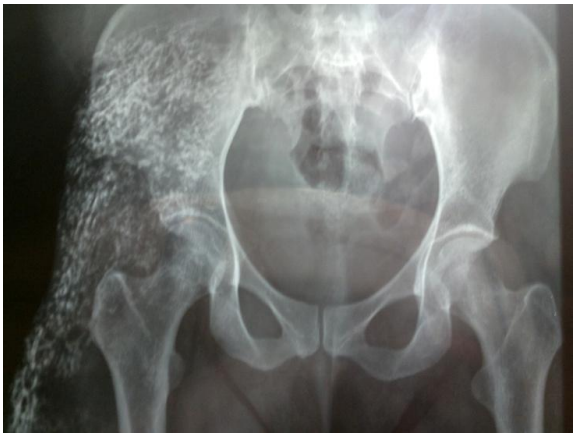

Figure 2: X-ray showing calcification.

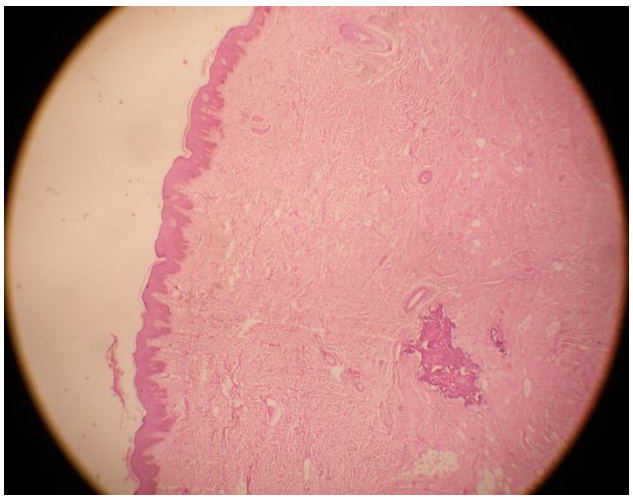

Figure 3: Histopathological slide (H & E staining) showing atrophic eccrine glands in collagenous stroma.

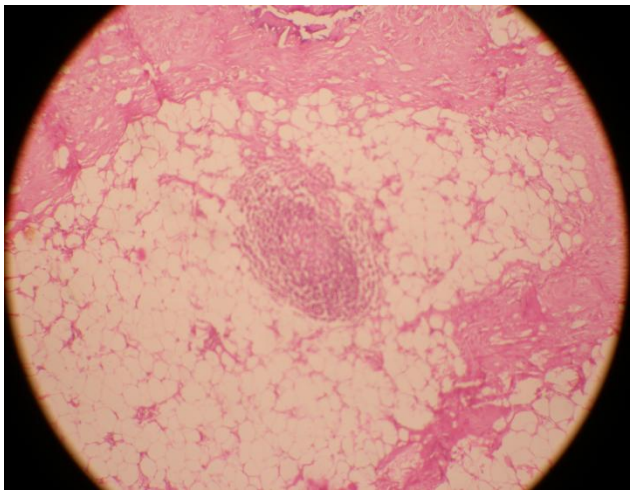

Figure 4: Histopathological slide (H & E staining) showing excess collagen and calcium deposition in subcutaneous adipose tissue septae.

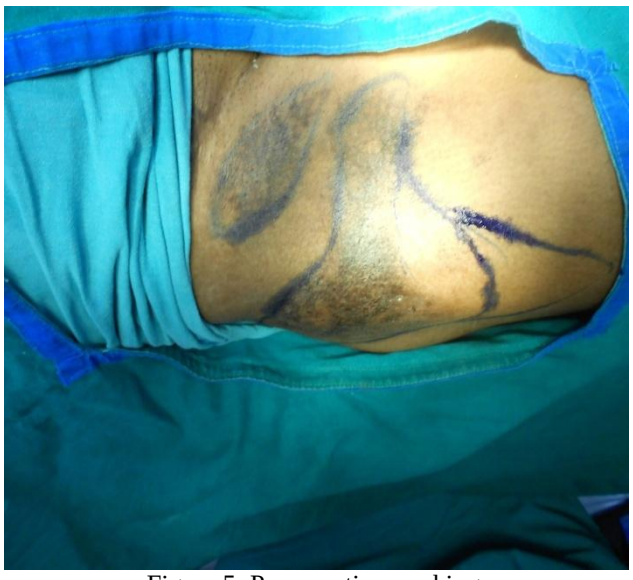

Figure 5: Preoperative marking.

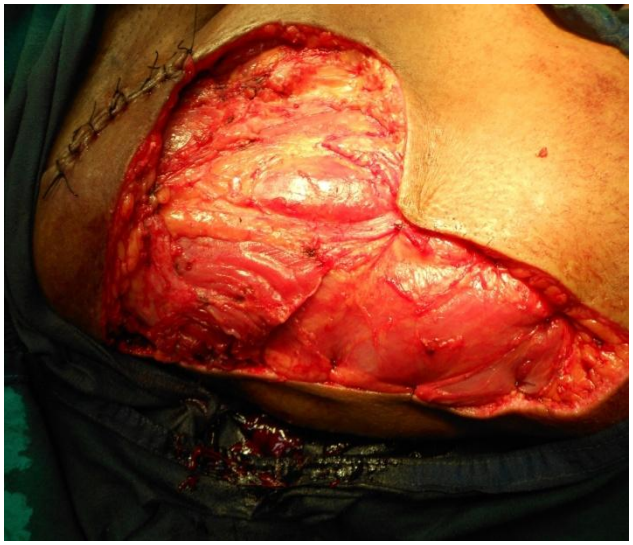

Figure 6: Intraoperative photograph after excision of lesion.

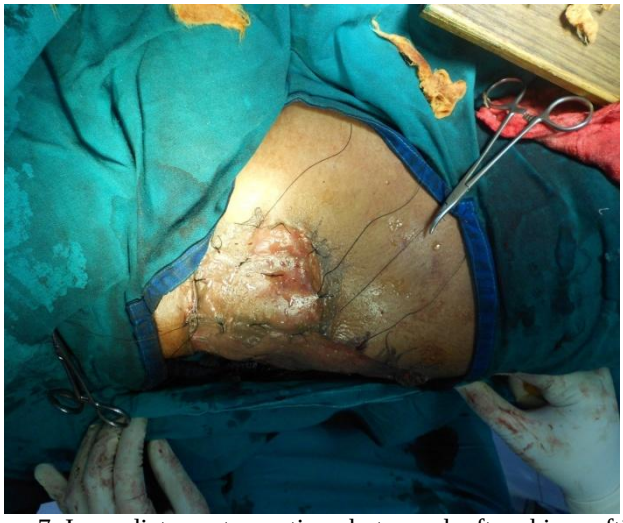

Figure 7: Immediate postoperative photograph after skin grafting.

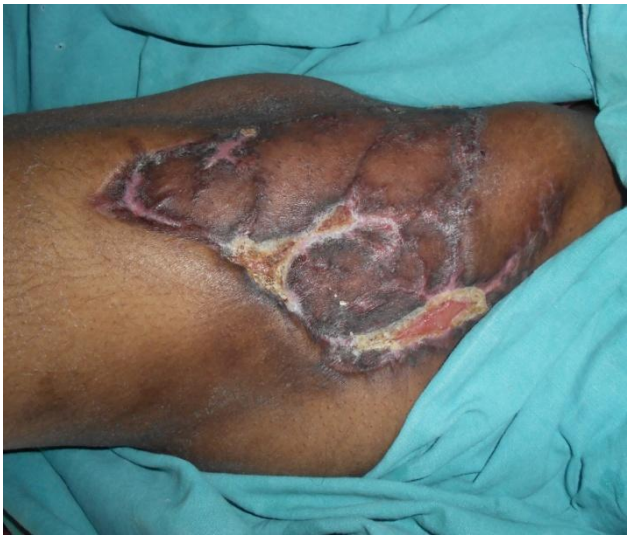

Figure 8: Postoperative photograph after healing.

V. Conclusion

Morphea Is A Rare Disease And Calcinosis In A Large Plaque Even Rarer. Surgical Intervention Has Resulted In Successful Outcome.

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