Acquired reactive perforating collagenosis in association with pulmonary papillary adenocarcinoma

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INTRODUCTION

Many dermatoses occasionally exhibit the phenomenon of transepithelial elimination (TEE), in which material from the dermis is extruded through the epidermis to the exterior with little or no disruption of the surrounding structures 1.

Acquired reactive perforating collagenosis (ARPC) is an uncommon dermatosis characterized by TEE of altered collagen 2. ARPC has been seen in association with multiple disorders including diabetes mellitus, renal failure, hyperparathyroidism, liver disease, neurodermatitis, IgA nephropathy, and periampullary carcinoma with jaundice 3. An incidence rate of 11% has been reported with a particular association with long-standing diabetes 1. The keratotic lesions develop on the trunk and limbs, and are usually pruritic dome-shaped papules with central crusts, which are not related to trauma 1.

The exact pathogenesis is unknown but there are some theories; fibronectin has a role in epithelial cell signaling, locomotion, and differentiation. It binds to type IV collagen and keratinocyte, and may affect epithelial proliferation and perforation 2. Others include abnormal Vit-A or Vit-D metabolism, enzyme release from neutrophils, microangiopathy in diabetes, and abnormal expression of metalloproteinases 2.

Treatment is difficult, and some include topical retinoids, topical corticosteroids, emollients, systemic retinoids, antihistamines, systemic corticosteroids, antibiotics, methotrexate, allopurinol if there is uremia, phototherapy, cryotherapy, and laser ablation 2.
CASE REPORT

A sixty-year-old Iranian man presented with a two-month history of multiple pruritic erythematous/violaceous umbilicated dome-shaped papules and nodules with central scaling and excoriation on the extensor surface of both legs (Figure 1) in association with weight loss, sweating, and chills and fever. He suffered from non-productive cough and mild dyspnea since ten days ago. In past medical history, he had diabetes mellitus since two years ago, and used glibenclamide 2.5 mg daily. Also, he was a heavy smoker since thirty years ago.

On physical examination, there was no abnormal cervical mass, no cervical lymphadenopathy, and no palpable organomegaly. On chest physical examination, breath sounds were increased in the right hemithorax, and increased tactile fremitus was observed at this site. There were multiple erythematous/violaceous umbilicated papules and nodules on the skin with central scaling and excoriation effects. Vital signs during admission were in the normal range.

Laboratory tests showed mild anemia (Hb=12.1 g/dl), with increased ESR (ESR=130) and positive CRP (CRP=3+). Thyroid, liver, and kidney function tests, and the serum calcium level were normal, and skin tuberculin test was reported negative.

Biopsy of the skin lesion revealed hyperkeratosis with a vertical cup-shaped invagination of the epidermis, containing a mixture of basophilic materials and degenerated collagen bundles (Figure 2). In the base of the invagination, the

Figure 1. Multiple pruritic  erythematous/violaceous umbilicated dome-shaped papules and nodules with central scaling and excoriation on the extensor surface of both legs

Figure 2. Epidermal necrosis replaced by fibrinoleukocytic exudates. A: collagen fibers extruded into the plug (H&E*40 ). B: Blue staining collagen bundles extruded into the keratinous plug (masson-trichrome*40 )
lining keratinocytes were eroded in some foci and the collagen bundles were vertically oriented.

Chest X-ray showed a solitary mass measured 5*5 cm in diameter in the middle lobe of the right lung with hilar lymphadenopathy in the same side (Figure 3), which was confirmed with chest CT scan. This mass had compressed the right middle bronchus, but there was no atelectasis in the lung. Pulmonary consultants suggested trans-thoracic biopsy with the guide of CT scan. Biopsy of the lung mass revealed malignant neoplasm, composed of sheets of round to oval cells with high nuclear cytoplasm ratio, pleomorphic and hyperchromatic round nuclei, and rare mitotic figures occasionally forming papillary structures, compatible with papillary adenocarcinoma. The patient was referred to the oncology service with the diagnosis of acquired reactive perforating collagenosis in association with pulmonary papillary adenocarcinoma.

DISCUSSION

Reactive perforating collagenosis is one of the perforating dermatoses; the acquired form is characterized by TEE of altered collagen bundles. It is an uncommon dermatosis with peak incidence in adolescence.

ARPC presents as pruritic erythematous/violaceous umbilicated dome-shaped papules and nodules with central scaling, with characteristic histopathology of TEE of collagen bundles. In some case reports and studies, this entity has been reported in association with different diseases but the most common associations are diabetes mellitus and chronic renal failure. Kreuter et al. reported a fifty-eight-year-old woman with typical lesions and a histopathologic feature in association with diabetes mellitus and chronic renal disease, receiving hemodialysis. Similarly, Mahanupab et al. reported a fifty-seven-year-old woman with dry gangrene of the right toe, reported to be ARPC lesions, in association with diabetes mellitus and chronic renal failure, who received hemodialysis. In a case series in 1994, Faver et al. showed ARPC in association with hypothyroidism, hyperparathyroidism, liver diseases, and neurodermatitis. There has been only one case report of ARPC in association with papillary adenocarcinoma of the thyroid. In the present study, there was a history of diabetes mellitus without any complications. The kidney and thyroid function tests were reported normal, and there was no sign of parathyroid dysfunction; therefore, the skin lesion was only associated with pulmonary papillary adenocarcinoma.

The differential diagnoses are other dermatoses with papules or nodules with a keratotic plug including prurigo nodularis, folliculitis, arthropod bites, perforating exogenous foreign materials or endogenous substances, and multiple keratoacanthomas; however, histopathologic study of TEE of collagen bundles suggests the diagnosis of RPC, and the onset of skin disease over 18 years of age shows the acquired perforating form. Therefore, ARPC remains a rare dermatosis.
but a well recognized one 8.

According to the literature, there is no report of ARPC in association with pulmonary papillary adenocarcinoma, and our study is the first to report it. Further studies are required to confirm our findings regarding the association between ARPC and pulmonary papillary adenocarcinoma.

REFERENCES


