CASE REPORT

Reactive Perforating Collagenosis; Dermatological Manifestation in a Patient with ESRD

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SUMMARY

Acquired perforating dermatosis (APD) is a heterogeneous group of skin disorders occurring in adults with underlying systemic disease, chronic kidney disease and diabetes mellitus, being the most common ones. Here we present a case of reactive perforating collagenosis in a 30 year old diabetic lady with End stage renal disease.

Key words: Acquired Perforating Dermatosis, Chronic Kidney Disease, End Stage Renal Disease

INTRODUCTION

Acquired perforating dermatosis comprises of acquired reactive perforating collagenosis (RPC), acquired elastosis perforans serpiginosa (EPS) acquired perforating folliculitis (PF) and kyrle disease. Among these, acquired reactive perforating collagenosis is the most common condition.

Clinically, acquired perforating dermatosis is manifested as multiple, firm, hyperkeratotic, umbilical papulonodules with central white crust varying in size from 2 to10 mm.

Histopathologically, they are characterized by trans-epithelial elimination of various substances like collagen, elastic fibers, fibrin and keratin. They usually appear on extensor surfaces of arms and legs. Their appearance on trunk and face has also been reported.

The pathogenesis of APD is still not clear but it has been postulated that there is overexpression of transforming growth factor beta and extracellular matrix protein which are responsible to stimulate the proliferation of keratinocytes[1,2]. There is a simultaneous trans-epidermal elimination of collagen and elastin and several mechanisms implicate to the development of these lesions which include diabetic microangiopathy, micro trauma caused by chronic pruritus, dysregulation of vitamin A and D, calcium deposits in skin and abnormalities of collagen fibers and/or elastin[3].

Management involves treatment of underlying cause along with antipruritic therapy in form of antihistamines, camphor or menthol based lotions, topical or intralesional corticosteroids, topical keratolytics like salicylic acid or topical retinoid and ultraviolet phototherapy with prevention of secondary bacterial infections.

The individual lesions can regress spontaneously and only general measures are needed to cure limited lesions like avoiding trauma or scratching, wearing gloves, applying menthol solution or behavioral changes. There is not a single gold standard treatment for the lesions with significant involvement but different treatment options have been tried with a variable success including topical retinoids, topical and intra-lesional steroids, emollients, laser ablation and cryotherapy. Other treatment measures are reserved for lesions with extensive involvement like antihistamines, oral retinoids, allopurinol, methotrexate and phototherapy.

The prognosis is better with considerable improvement in pruritus and skin lesions after the treatment of underlying systemic disease and usually no specific follow up is required.

CASE REPORT

A 30 year old lady who is type 1 diabetes for last 10 years and diagnosed with CKD stage V secondary to diabetic nephropathy one year ago with baseline creatinine of 4.0 mg/dl in December, 2018, presented to surgeon for construction of arterio-venous fistula (AVF) but at that time she was in fluid overload and severe metabolic acidosis. She was therefore referred to nephrology department for further management. Patient has never been dialyzed before and she was optimized by doing a dialysis session through temporary hemodialysis catheter

She had papulonodular lesions on her legs (Fig. 1), arm (Fig. 2), trunk and face for last 2 months that developed gradually and were associated with severe itching that was more at night and not relieved by lubricants.

Physical examination revealed multiple hyperpigmented, well demarcated, papulonodular lesions of varying size with central keratin plug. These lesions were non tender without discharge.

Fig. 1

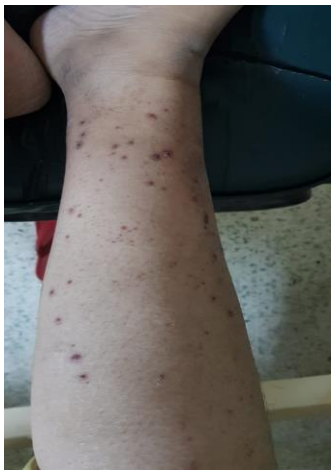
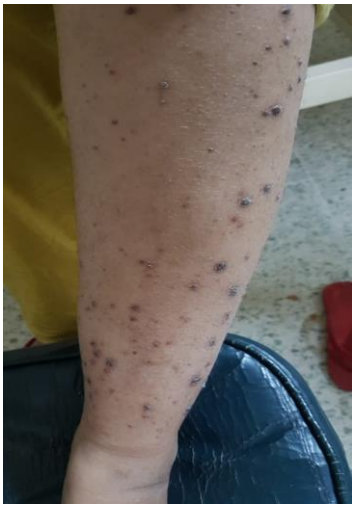


Fig. 2

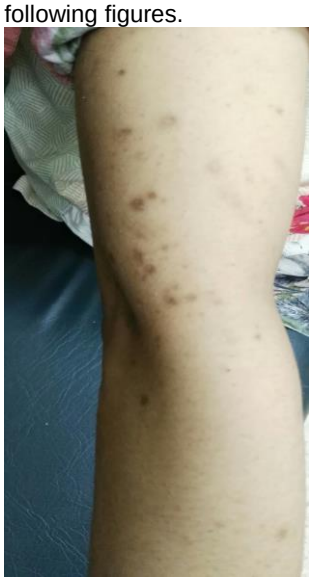


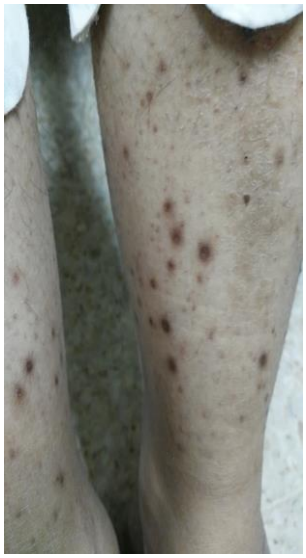
On admission, her labs showed Serum Creatinine of 13mg/dl, BUN of 90mg/dl, Albumin 2.7g/dl, Na+ 130 mmol/L, K+ 6.6 mmol/L, Ca++ of 5.6 mg/dl, Phosphate of 6.3 mg/dl, and Intact-PTH of 188 pg/ml and Uric acid of 5.1mg/dl.

We commenced renal replacement therapy in the form of hemodialysis. Total 4 sessions of hemodialysis were done and a dry weight of 62 kg was attained. She was given antihistamine and topical corticosteroids for her skin lesions. Skin biopsy was planned but patient refused it.

Patient was labeled as end stage renal disease (ESRD) and was discharged on thrice a week hemodialysis, topical treatment for skin lesions and 2 weekly follow up. On discharge her labs showed creatinine of 8.5 mg/dl, BUN of 42 mg/dl, albumin 2.3 g/dl, Na+ 137mmol/l, K+ 137mmol/l, and Ca++ 7.3 mgldl, PO4 of 4.6 mg/dl and uric acid of 4.5 mg/dl. The viral serology's for Hepatitis B, Hepatitis C and HIV were negative.

These lesions regressed after 8 weeks as shown in the following figures.





DISCUSSION

There has been a recent increase in numbers of patient with ESRD. The dermatological manifestations in dialysis patients are not uncommon these days but they are still under noticed as well as under documented. Various skin lesions in patients of renal replacement are either due to dialysis or as a result of immunosuppressant therapy.

Perforating dermatosis is a group of unusual skin disorders reflected by altered collagen and/or elastin. Reactive perforating collagenosis is a rare entity of perforating skin disorders and it was first explained by Mehregan et al in 1967[2,4].

It occurs in 2 forms, acquired and inherited, the former being more common. It has been found that 10% of dialysis patients suffer from this condition[3,5]. It is usually manifested after starting dialysis but sometimes it may occur even before the start of dialysis as is the case with our patient. Majority of studies has revealed its association with hemodialysis (HD) rather than peritoneal dialysis (PD) but still there is no study available comparing population on these two modalities of dialysis. More epidemiological studies are needed to evaluate the prevalence of APD in patients on PD and the risk factors associated with APD in the PD population[6,7].

The classical presentation of this condition is rash with umbilicated papules and central keratotic cap and the most common manifestation is pruritus but patient may also experience pain sometimes. Our patient's presentation is characterized by large plaques of 1-2 cm of diameter[6]. It closely resembles the giant variant of APD which was first described by Hoque et al. Only five patients with this presentation have been reported in the literature so far[8,9].

The diagnostic investigation of choice is skin biopsy and histopathology is required to show the findings of any of the perforating dermatosis. There is an epidermal invagination with the keratotic plug comprising of keratin collagen or elastin fibers along with neutrophils.

The mean age of presentation is 4th to 5th decade of life with both genders being similarly affected. The duration of illness varies from weeks to years whereas the disease duration before the time of diagnosis varies from three months to five years mainly because of unusual presentation and inability to make diagnoses in time. The most common site of these lesions is the extensor surface of lower extremities but it can also appear in the trunk, scalp or any scratch area of the patient[9].

APD is most commonly associated with CKD (73%) and DM (50%) according to a study but recent developments in diagnostic approaches have revealed its association with other systemic diseases also like chronic liver disease, thyroid disorders, scabies, atopic dermatitis, tuberculosis, HIV and malignancies including lymphoma and pancreatic cancer. The present statistics prove that APD has a more causal relationship to kidney diseases than to diabetes[3]. But it is still not clear that which type of dialysis modality is linked to the outcome of this disease. A case reported by Gonzalez Lara et al revealed a patient who was diagnosed with APD after peritoneal dialysis, with improvement after being switched to hemodialysis[6].

APD is sometimes associated with severe itching and can impair the quality of life of patients and may also lead

to significant depression, therefore nephrologists should be well aware of these eruptions in patients of diabetes and chronic kidney disease receiving hemodialysis.

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