Post-steroid panniculitis: A rare case report

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ABSTRACT

Post-steroid panniculitis (PSP) is a rare clinical entity which presents after rapid withdrawal of high doses of systemic corticosteroids. As few as 20 cases have been reported in literature till now. Here, we report a case of post-steroid panniculitis occurring in a 9-year-old boy after rapid tapering and discontinuation of corticosteroids administered for the treatment of nephrotic syndrome. He presented with multiple erythematous painful indurated nodules over the face, arms, forearms, thighs, and legs. Histopathologic examination of the nodule revealed lobular panniculitis with lymphocytes, neutrophils, and multiple multinucleated giant cells. It also showed multiple needle-shaped clefts. Based on history, clinical features, and histopathologic findings, a diagnosis of post-steroid panniculitis was made and the patient was restarted on systemic corticosteroids. The lesions resolved in 4 weeks. We report this case to highlight the importance of gradual tapering of corticosteroids.

Key words: Needle-shaped clefts, post-steroid panniculitis, systemic corticosteroids

INTRODUCTION

Post-steroid panniculitis is a rare complication of systemic corticosteroid therapy, which develops within days or weeks following rapid tapering or cessation of the drug. Till date, 20 cases have been described in medical literature. Most of the reported cases have been in children. Clinically it presents with erythematous nodules varying in size from 0.5 to 4 cm, and tend to localize in those areas where there is the greatest accumulation of fat from steroid therapy, typically on the cheeks. Clinically it has to be differentiated from lupus profundus, and Weber–Christian disease, which also present with similar lesions but heal with atrophy and depressed scars. Histopathology of post-steroid panniculitis shows lobular panniculitis with needle-shaped clefts and has to be differentiated from sclerema neonatorum and subcutaneous fat necrosis of the newborn (SCFN). Most cases are self-limited, with very few cases requiring reintroduction of high doses of corticosteroids.

CASE REPORT

A 9-year-old child, referred from nephrology department, presented with multiple painful swellings over the cheeks, arms, forearms, thighs, and legs of 3 days duration. Child was a known case of nephrotic syndrome for the past 2 years. Child had two previous episodes of nephrotic syndrome. For the first episode, the patient was put on 30 mg prednisolone once a day for 6 weeks, then tapered to 20 mg once daily alternate day for 6 weeks, and then stopped. For the second episode, the child was treated with 30 mg prednisolone once a day for 2 weeks, then tapered to 20 mg once daily alternate day for 4 weeks, and then stopped. For the present episode, he was put on 30 mg of prednisolone for 5 days, then tapered to 20 mg for 5 days, followed by 10 mg for 5 days, and then stopped as he had developed features of steroid-induced Cushing’s disease. The present episode started 9 months after the previous episode and the child was started on steroids within a week after aggravation. After 3 days of stopping the drug, the patient developed painful erythematous nodules over the cheeks, arms, forearms, thighs, and legs. Child was afebrile. Cutaneous examination revealed multiple erythematous tender nodules varying in size from 1 to 5 cm, distributed over the cheeks, arms, forearms, thighs, and legs bilaterally [Figures 1–3]. The lesions were tender and indurated.

Investigations revealed normocytic normochromic anemia. Erythrocyte sedimentation rate was 32 mm/h. Antistreptolysin O titer and Mantoux test were negative. Biopsy revealed lobular panniculitis with lymphocytes, neutrophils, and multiple
multinucleated giant cells. It also showed multiple needle-shaped clefts [Figures 4 and 5].

A diagnosis of post-steroid panniculitis was made based on history, clinical findings, and histopathologic examination. The patient was restarted on 30 mg of prednisolone with resolution of the lesions in 4 weeks without any sequelae. Patient was followed up for 3 months without any evidence of relapse.

DISCUSSION

An inflammatory disorder that is primarily localized in the subcutaneous fat is termed as panniculitis. This group of disorders may be challenging both for dermatologist and pathologist.[2] Clinically, they present as erythematous or skin-colored subcutaneous nodules or indurated plaques that may be tender.

The etiology of panniculitis varies from physical insults to infections to malignancies. Histopathologic features in all these conditions may show considerable overlap making the diagnosis difficult. In addition to biochemical and other tests, expert interpretation of the histopathology is needed for correct classification. In spite of this, the exact etiology may not be apparent in many cases.[3]

Smith and Good first described post-steroid panniculitis in 1956.[4] Post-steroid panniculitis has been reported in children who were receiving high doses of systemic corticosteroids that were tapered quickly or suddenly withdrawn. In the adult population, only one case has been reported.[5] Patients who
have developed this variant of panniculitis have received high doses of systemic corticosteroids for a variety of conditions, including rheumatic fever, leukemia, nephrotic syndrome, secretory diarrhea, brain stem glioma, and cerebral edema.[6] Clinically, cutaneous lesions of post-steroid panniculitis consist of erythematous subcutaneous nodules and plaques that appear 1–10 days after stopping high doses of systemic corticosteroids. As the lesions develop mostly in those areas in which steroids induce the greatest accumulation of fat, subcutaneous nodules appear mostly on the cheeks, posterior neck, and upper trunk. In contrast to this, our patient showed nodules over cheeks, arms, forearms, thighs, and legs. Clinically it has to be differentiated from cold-induced panniculitis, popcorn panniculitis, erythema nodosum, lupus profundus, and Weber–Christian disease.[7]

Histopathologically, post-steroid panniculitis shows features identical to those of subcutaneous fat necrosis of newborn. They consist of a mostly lobular panniculitis with an inflammatory infiltrate of foamy histiocytes and lymphocytes involving fat lobules. Often narrow strands of needle-shaped clefts are evident within the cytoplasm of some histiocytes.[6]

Needle-shaped clefts within adipocytes and giant cells can be seen histologically in only two other conditions: sclerema neonatorum and SCFN.[7] SCFN is an uncommon, benign disorder found in full-term or post-mature neonates. It usually presents in neonates who have experienced perinatal difficulty such as asphyxia, peripheral hypoxemia, hypothermia, meconium aspiration, or trauma, usually detected between 2nd and 21st day of life. Clinically presents with nodules symmetrically distributed over buttocks, thighs, shoulders, back, cheeks, and arms. General condition of the child is not impaired, and within a few months the nodules disappear.[8]

Sclerema neonatorum is a rare disorder occurring during the 1st week of life in premature and small for dates babies with underlying severe illness, particularly severe infections, congenital heart disease, and other major developmental defects. It clinically presents with woody induration of the skin on buttocks, thighs, or calves, and extends rapidly symmetrically to involve almost the whole surface except palms, soles, and genitalia. Prognosis is poor with up to 75% mort’[1]

Sclerema neonatorum and SCFN occur almost exclusively in infants, whereas PSP can also present in older children, as well as in adults. Additionally, PSP occurs following the tapering or cessation of systemic steroid therapy, whereas the other disorders lack this medication history.

The precise mechanism by which the panniculitis arises is not known. It is hypothesized that the withdrawal of systemic corticosteroid leads to abnormal lipid metabolism, resulting in the elevation of saturated to unsaturated fatty acid ratio.[3] As most of the cases resolve spontaneously, no treatment is needed. Rarely reinstitution of high doses of systemic steroid therapy followed by gradual tapering is advised in severe cases to avoid ulceration and scarring.[5]

We are reporting this case for its rarity and to highlight the significance of gradual tapering of systemic steroids when given in high doses in children.

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