Bejeweled: Chronic Bullous Disease of Childhood in a 2-Year Old Treated with Colchicine: A Case Report*

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ABSTRACT

Linear IgA bullous dermatosis, also known as chronic bullous disease of childhood when present in the pediatric age group, is a rare blistering disease more predominantly seen in females less than five years old. This case describes a 2-year old girl who presented with scattered, tense vesicles and bullae on an erythematous base forming the classic "cluster of jewels" appearance. This clinical picture is often mistaken as bullous impetigo, commonly seen in children, delaying diagnosis and prompt treatment. Histopathologic examination showed subepidermal blistering with a predominantly neutrophilic inflammatory infiltrate. The direct immunofluorescence studies revealed a linear band of IgA deposition in the basement membrane zone consistent with the diagnosis of CBDC. The patient was started on colchicine and oral prednisone at 1 mg/kg/day and complete resolution was achieved within two weeks of therapy.

Keywords

Linear IgA bullous dermatoses, Chronic bullous disease of childhood, colchicine

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INTRODUCTION

Chronic bullous disease of childhood (CBDC) is a rare autoimmune blistering disease predominantly seen in children less than five years old with a slight female predominance.[1] This entity presents as multiple scattered tense vesicles and bullae on an erythematous base. These vesicles occur in clusters exhibiting a "cluster of jewels" or a "string of pearls" appearance. Fresh lesions are sometimes seen around older ones presenting as a "collarette of blisters". Histopathologically, it presents as subepidermal blistering confirmed through direct immunofluorescence, seen as a homogenous linear band of IgA1 deposition at the basement membrane zone.[2] Antibodies are said to target more than 1 antigen. IgA is bound to a 97 k-Da antigen present in the lamina lucida. This antigen is identical to the 180 k-Da BP antigen essential in anchoring basal keratinocytes to the epidermal basement membrane.[1] This disease will occur with spontaneous resolution of lesions within two years however, treatment is aimed at controlling flares and inducing remission. The standard of care involves the use of dapsone at 25-100 mg/day however, due to difficult access for this drug, colchicine was used and was noted to control the flare.

Case Report

A 2-year old female presented with a 1-month history of a solitary tense blister in the left lower leg that eroded upon scratching. There was subsequent spread to the lower extremities, trunk, and neck leaving erosions due to incessant scratching. The patient was given hydrocortisone cream applied twice a day for a week and oral antihistamines which provided no relief. Pulverized amoxicillin was later applied with an increase in the number of lesions. The blisters eventually spread to the face which prompted consult. The primary service diagnosed this as a case of bullous impetigo and referred to the dermatology service. Cutaneous examination revealed multiple well-defined, salmon pink-red annular plaques bordered by tense vesicles and bullae arranged in a cluster of jewels appearance. Some plaques appear with central erythematous erosions topped with hemorrhagic crusting. [Figure 1 A-D]

Histopathology revealed a subepidermal blister with a moderately dense superficial perivascular and interstitial infiltrate composed of neutrophils mostly, and occasional lymphocytes. [Figure 2 A-B] Direct immunofluorescence showed a homogenous linear band of IgA at the dermo-epidermal junction consistent with linear IgA disease. [Figure 3]

As dapsone is not locally available, the patient was started on oral prednisone at 1 mg/kg/day after breakfast. There was slight resolution of lesions, but new lesions erupted. [Figure 4 A-F]. Colchicine was started at 500 mg twice a day with resolution of lesions in two weeks. [Figure 5 A-J]

Discussion

This case of a 2-year old girl with a solitary blister on the left leg that progressed to tense vesicles and bullae within the border of erythematous annular plaques in a cluster of jewels appearance was treated with an oral corticosteroid to immediately suppress the inflammation coupled with a second line drug and noticeable improvement was seen. Most cases of CBDC are treated with dapsone at an initial dose of 25 mg to 100 mg per day. This can be given together with oral corticosteroids. The average treatment duration is 20 months.[5] In the Philippines, dapsone is only available as part of the multidrug treatment blister packs for the treatment of leprosy hence colchicine was given. Colchicine inhibits microtubule assembly through the formation of tubulin that interferes with mitosis and cellular migration. It also inhibits cellular adherence, motility, and chemotaxis. One of its off-label uses includes the treatment of CBDC at a dose of 0.5 mg/day. [6] This drug has been proven safe and well tolerated by children with a safety profile comparable to its use in adults. [7]. The most common side effects reported are gastrointestinal in nature. Diarrhea, abdominal pain, nausea and vomiting may be seen in patients taking high doses (about 4.6 mg in 6 hours). This patient was given a dose within the prescribed therapeutic dose per age and did not experience any of these side effects. A baseline complete blood count was taken and then monitored for agranulocytosis, anemia, and thrombocytopenia. The blood urea nitrogen, serum creatinine, aspartate amino transferase and alanine aminotransferase were also taken since dosing requirements are adjusted in cases of hepatic and renal impairment.[8]

According to Banodkar et al., CBDC, histopathologically, presents with a neutrophilic predominance and the action of colchicine against neutrophils was a clear indication for using this medication for this case. [7,8] Colchicine dosed at 0.5 mg twice a day with concomitant decrease in oral prednisone was said to achieve total remission with no relapse noted on maintenance. In the case reported by Zeharia et al., colchicine at the said dose was started twice daily and remission was achieved in two weeks without any side effects noted in the child.[9] Another case of CBDC was reported by Salud et al., wherein oral prednisone at 0.5 mg/kg/day tapered over a month resulted in resolution of the bullae within 2 weeks.[10]. Prednisone at 1 mg/kg/day may be also started initially as one waits for the histopathologic report and colchicine at 0.5mg twice daily be added once certain with the diagnosis. [11]

Conclusion

Chronic bullous disease of childhood or childhood linear IgA disease is a rare acquired autoimmune blistering disease in children less than 5 years old with a classic appearance of tense vesicles and bullae arranged seemingly like a cluster of jewels. Due to the neutrophilic nature of the disease, dapsone remains the first choice in treatment. Although mostly used in cases of G6PD deficiency, colchicine exhibits excellent action against neutrophils and is a viable option. This drug is readily available, accessible, and affordable. In the Philippines, literature is scarce in in terms of the use of colchicine as first line agent for linear IgA disease. We present this case to demonstrate the efficacy and safety of colchicine in chronic bullouse disease of childhood and to advocate its use as a first line agent.

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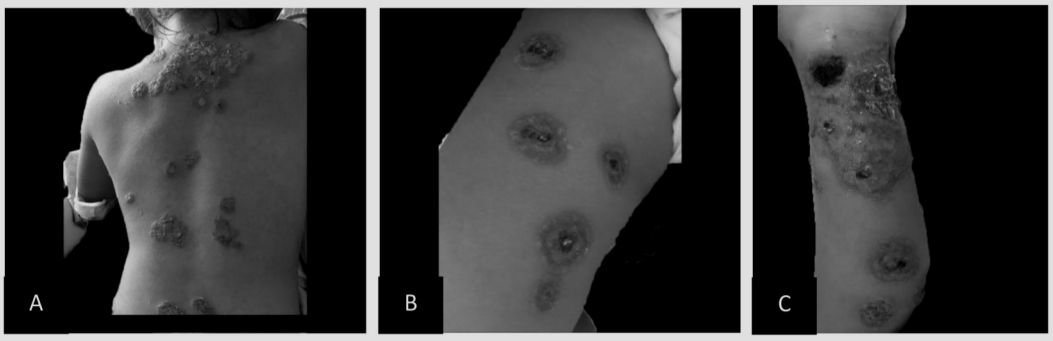
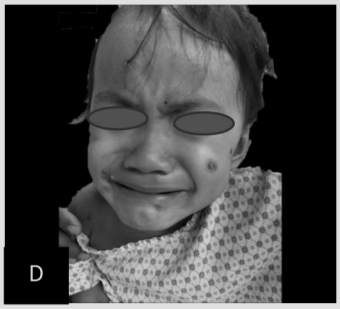
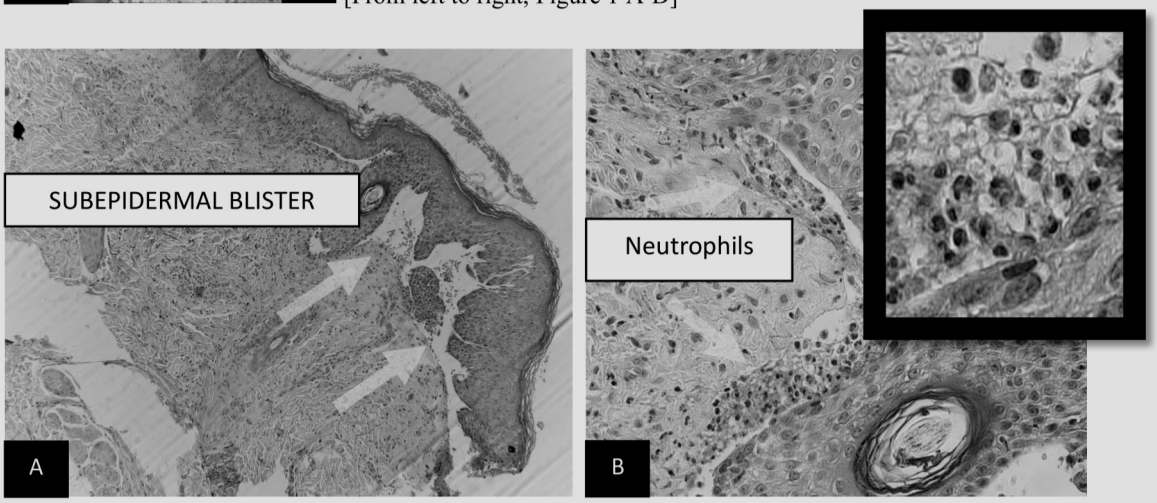
APPENDIX

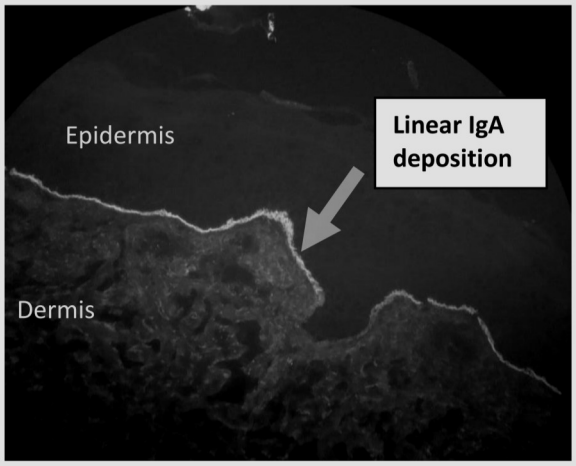




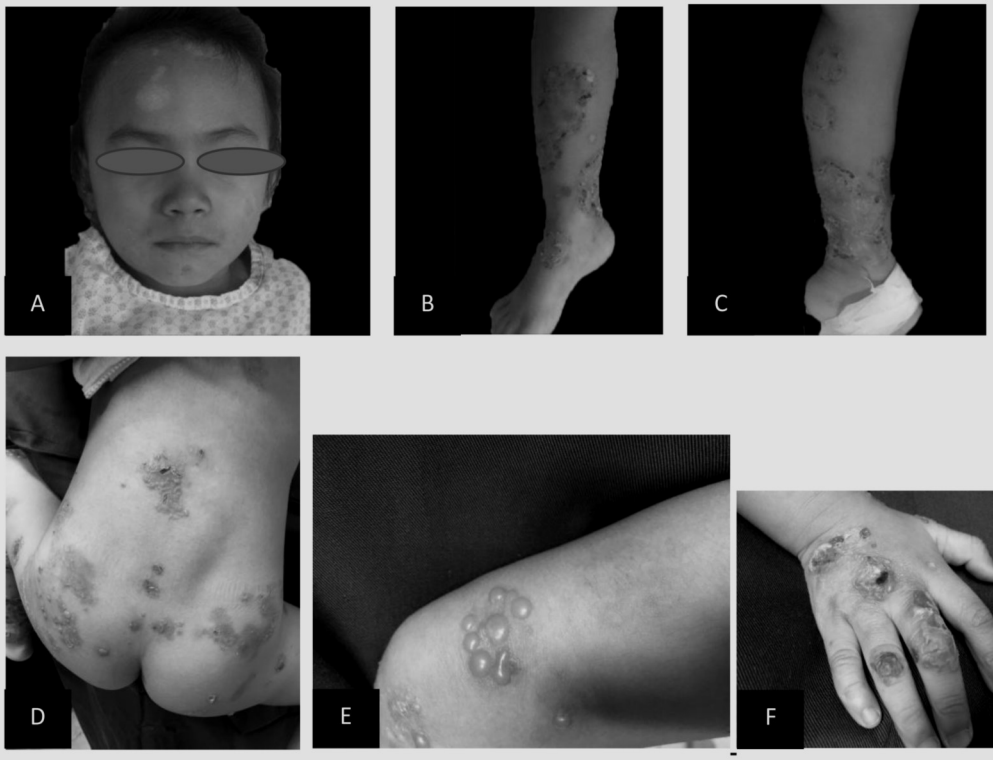
[From left to right, Figure 1 A-D]



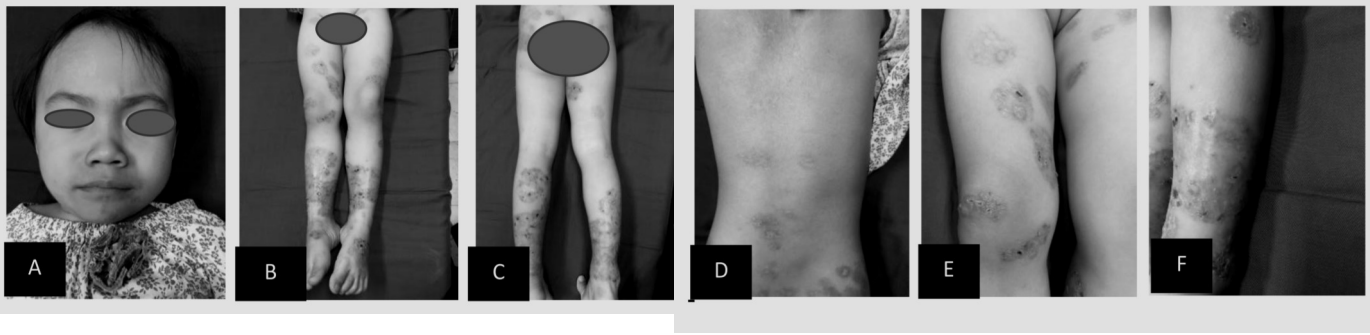
[Figure 2 A-B]

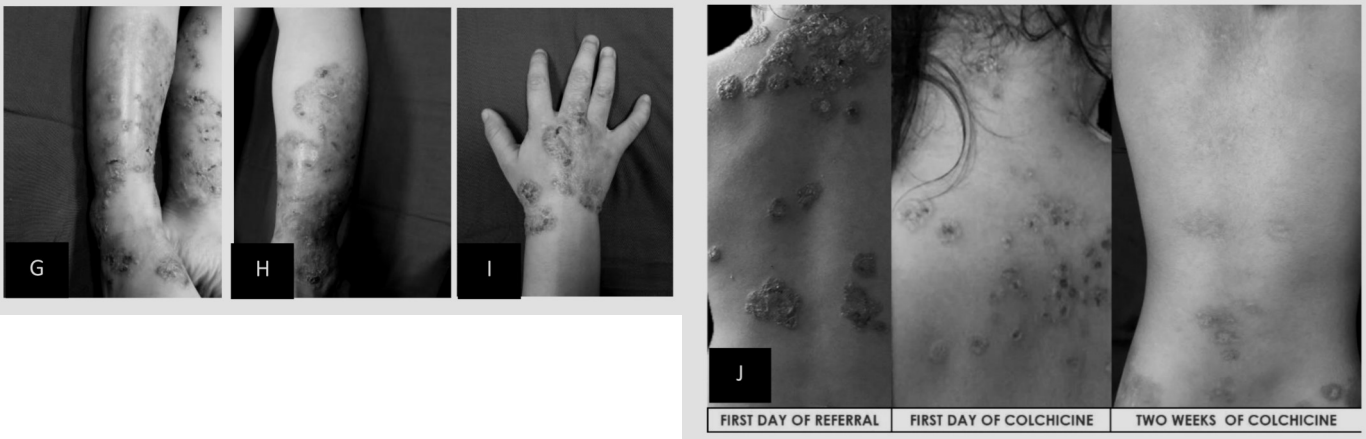


[Figure 3]



[Figure 4 A-F]





[Figure 5 A-J]