Journal of Pakistan Association of Dermatologists. 2021;**31(4)**:720-724.

Case Report

A generalized, pruritic, papulo-vesicular eruption in an infant: An uncommon presentation of Gianotti Crosti Syndrome

Jaspriya Sandhu, Shriya Garg, Sunil Kumar Gupta, Bhavna Garg*

Department of Dermatology, Venereology & Leprosy, DMCH, Ludhiana, Punjab, India.
* Department of Pathology, DMCH, Ludhiana, Punjab, India.

Abstract A 1-year- old boy was brought by his parents with a 2-week-history of skin colored to erythematous crusted, papulo-vesicles distributed symmetrically over face, extremities and prominent truncal involvement. Lesions were intensely pruritic, disrupting the child's sleep; an episode of upper respiratory tract infection (URTI) preceded the eruption. A skin biopsy was done keeping the differential of Gianotti Crosti Syndrome (GCS), histiocytosis and pityriasis lichenoides et varioliformis acuta (PLEVA); epidermis showed acanthosis, hyperkeratosis and follicular plugging, with scant perivascular lymphocytic infiltrate in the dermis. Emollients, topical steroids and antihistamines led to marked improvement at 1-week follow-up; on clinico-pathological correlation, a diagnosis of an Atypical GCS was made. To the best of our knowledge, such an atypical presentation of GCS has not been previously reported.

Key words
Gianotti Crosti Syndrome, acrodermatitides, pediatrics, exanthem, atypical Gianotti-Crosti syndrome.

Introduction

Gianotti Crosti Syndrome (GCS) also known as infantile papular acrodermatitis or papular acrodermatitis of childhood, is a self-limiting non- pruritic papular eruption following a viral infection/vaccination.[1] Commonly seen in the age group of 1-6 years, may be seen in adolescents and adults.[2]

The classical lesion morphology is monomorphic, flat topped, pink-brown papules located symmetrically on extensor surfaces of limbs, buttocks, and cheeks lasting for at least 10 days.[1] Over pressure points (elbows and knees), lesions may coalesce to form plaques.[2]

Presence of truncal lesions is generally considered a negative finding in GCS, as per the diagnostic criteria proposed by Chuh *et al*.[1] Truncal lesions, if present, are usually mild and transient. Lesions may be associated with mild to moderate pruritus.[2] Various atypical presentations of GCS have been reported in the literature.[3,4] Histopathological findings are generally non-specific.[2] We present here, a case of atypical GCS associated with intense pruritus and extensive truncal involvement.

Case report

A 1-year-old boy was brought by his parents to the dermatology outpatient department with chief complaints of itchy, reddish, raised lesions on body for 2 weeks. The lesions first appeared on the upper limbs and later progressed to

Address for correspondence
Dr Jaspriya Sandhu, Assistant Professor
Dayanand Medical College & Hospital,
Ludhiana, Punjab 141001, India.
Ph: 9582710091
Email: sandhu.jaspriya@gmail.com

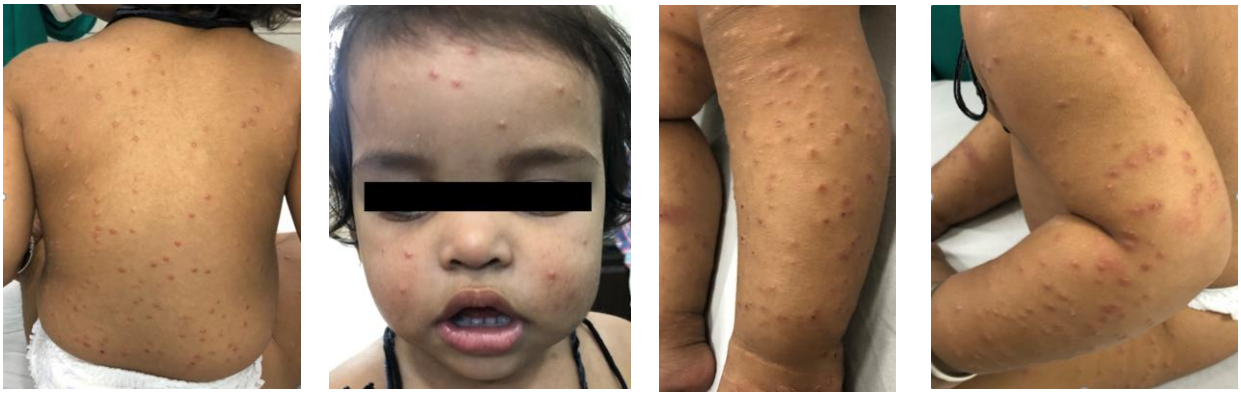

Figure 1 Skin colored to erythematous crusted, papulo-vesicular eruption on back, face, lower limbs,upper limb.

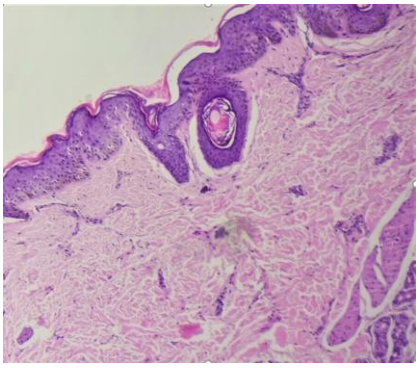

Figure 2 (H&E, 100x) Epidermis shows hyperkeratosis, acanthosis, follicular plugging; and scant perivascular lymphocytic infiltrate is seen in the dermis.

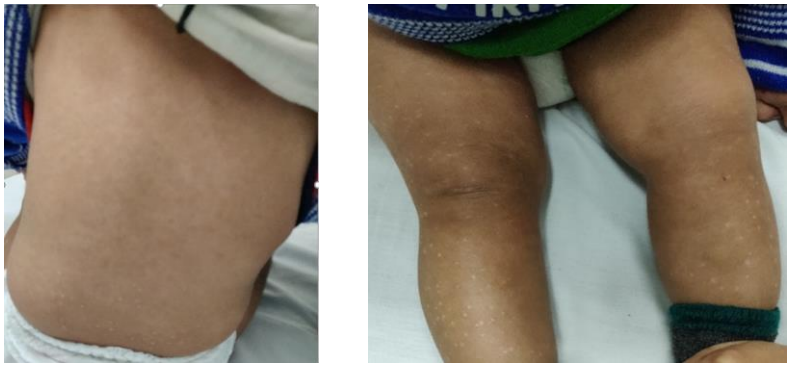

Figure 3 Resolution of lesions at 1 week follow up with mild hypopigmentation on back & lower limbs.

involve the lower limbs, face & torso. An associated history of intense pruritus was reported, which disrupted the child's sleep. The child had an episode of fever associated with upper respiratory tract infection (URTI) 10 days prior to the onset of lesions. There was no history of recent immunization. No personal/ family history of atopy could be elicited.

The child was active, alert and playful. Skin colored to erythematous crusted, papulo-vesicular lesions were present symmetrically over the trunk, buttocks, bilateral upper and lower limbs as well as face. Excoriation marks at few sites were seen **(Figure 1).** Mucosae were unremarkable; palms and soles were spared. No lymphadenopathy or organomegaly was seen. Rest of the systemic examination revealed no abnormality.

A complete haemogram and liver function test (LFT) were sent and were within normal limits. Viral markers were non- reactive. A skin biopsy was done, keeping a differential of Gianotti Crosti Syndrome (GCS), histiocytosis and pityriasis lichenoides et varioliformis acuta (PLEVA). On histopathological examination; acanthosis, hyperkeratosis and follicular plugging of the epidermis was seen. The dermis showed scanty perivascular lymphocytic infiltrate **(Figure 2)**. The child was prescribed topical steroids, emollients and anti-histamines; marked improvement was seen after one week. The lesions had resolved completely with few sites showing post- inflammatory hypopigmentation **(Figure 3)**. On clinico-pathological correlation, a final diagnosis of atypical GCS was made.

Table 1 Diagnostic Criteria for Gianotti Crosti Syndrome as proposed by Chuh *et al.* [1].

A patient is diagnosed as having Gianotti-Crosti syndrome (GCS) or papular acrodermatitis if:

1. On at least one occasion or clinical encounter, he/she exhibits all the positive clinical features, and
2. On all occasions or clinical encounters related to the rash, he/she does not exhibit any of the negative clinical features, and
3. None of the differential diagnoses is considered to be more likely than GCS on clinical judgment, and
4. If lesional biopsy is performed, the findings are consistent with GCS.

The positive clinical features are:

1. Monomorphous, flat-topped, pink-brown papules or papulovesicles 1–10 mm in diameter on at least three of the following four sites: cheeks, buttocks, extensor surfaces of the forearms, extensor surfaces of the legs.
2. Being symmetrical.
3. Lasting for at least 10 days.

The negative clinical features are:

1. Extensive truncal lesions.
2. Scaly lesions.

Discussion

An eruptive, acral possibly infectious exanthem was first described by Ferdinando Gianotti in 1955.[5] Two years later, Ferdinando Gianotti and Agostino Crosti reported a possible viral etiology of this acral, papular exanthem.[6] Since then, GCS has been studied and reported by various authors and a diagnostic criteria for GCS was proposed by Chuh *et al.* **(Table 1).**[1]

The exact aetio-pathogenesis is uncertain; however, a possible delayed hypersensitivity to a pathogen or vaccine has been proposed.[2]About one- third of patients may have a prodrome of cough, low grade fever, sore throat, malaise about a week prior to the onset.[2] In this case, the child had a URTI episode 10 days prior to the episode.

Anecdotal reports of unusual or atypical clinical presentations of GCS, found on literature review, have been summarized in **Table 2.**[3,4,7,8]

Unusual clinical features in this case were extensive papulo-vesicular truncal lesions and intense pruritus. The lesions resolved with mild post- inflammatory hypopigmentation; which is known to occur in darker skin types.[2,7]

To the best of our knowledge, such an atypical presentation of GCS has not been previously reported from India.

Table 2 Review of literature: Atypical Gianotti Crosti Syndrome seen in pediatric age group (<18 years)

S.No.	*Author, Journal, Year (Country)*	*Age*	*Sex*	*Clinical Presentation*	*Histopathological examination*	*Remarks*
1	Linke *et al.* [8], Acta Derm Venereol, 2011 (Germany)	7 yrs.	F	Papules and plaques on the limbs; tense bullae present on hands & feet only. *Truncal lesions absent* *Pruritus present*	*Epidermis:*Focal acanthosis, spongiosis and pseudo-Pautrier abscesses. *Dermis:* A moderate superficial perivascular lymphocytic infiltrate with few eosinophils.	Oral steroids (Methylprednisolone 32mg/ day- gradually tapered) and antihistamines were prescribed with complete resolution after 1 week.
2	Lam [4], J Am Acad Dermatol, 2011 (Canada)	2 yrs.	F	Erythematous, monomorphic, edematous,non- scaly, papulo-vesicles coalescing to form plaquesover left cheek & ear; scattered, discrete papules present on the right cheek *Truncal lesions absent* *Pruritus absent*	Not done	Lesions appeared after H1N1 vaccination and thereafter resolved spontaneously. Recurrence was reported with booster dose.
3	Sarma *et al.* [7], Indian J Dermatol, 2013 (India)	1.5 mo.	M	Extensive haemorrhagic vesicles along with papules over hands, feet, extensor surfaces of lower & upper limbs, face. Hepato-splenomegaly present. *Truncal lesions absent* *Pruritus absent*	Not done	EBV serology was positive Lesions resolved with mild hypopigmentation
4	Marcassi *et al.* [3], An Bras Dermatol, 2018 (Brazil)	22 mo.	M	Grouped vesicles localized to the face & extensor aspect of limbs. *Truncal lesions present* *Pruritus absent*	Not done	EBV serology was positive. Resolved spontaneously within 20 days
5	*Present Case (India)*	1 yr.	M	Crusted, papulo- vesicular lesions present symmetrically over B/L upper & lower limbs, face, buttocks *Truncal lesions present* *Severe pruritus present*	*Epidermis:* Acanthosis, hyperkeratosis and follicular plugging. *Dermis:* Scant perivascular lymphocytic infiltrate	Preceding history of URTI was present. Emollients, topical steroids and antihistamines were prescribed. Lesions resolved after 1 week with mild hypopigmentation.

References

1. Chuh A, Lee A, Zawar V. The diagnostic criteria of Gianotti-Crosti syndrome: are they applicable to children in India? *Pediatr Dermatol*.2004;**21(5)**:542-7
2. Leung AKC, Sergi CM, Lam JM, Leong KF. Gianotti-Crosti syndrome (papular acrodermatitis of childhood) in the era of a viral recrudescence and vaccine opposition. *World J Pediatr*.2019;**15(6)**:521-7.
3. Marcassi AP, Piazza CAD, Seize MBMP, Cestari SDCP. Atypical Gianotti-Crosti syndrome. *An Bras Dermatol*.2018;**93(2)**:265-7.
4. Lam JM. Atypical Gianotti-Crosti syndrome following administration of the AS03-adjuvanted H1N1 vaccine. *J Am Acad Dermatol*.2011;**65(4)**:e127-8.
5. Gianotti F. Report on a special case of toxic infection characterized by a desquamative erythemato-infiltrative eruption with lenticular foci and a selective localization at the extremities. *Soc Ital Dermatol Sifilogr Sezioni Interprov Soc Ital Dermatol Sifilogr*. 1955;**96(6)**:678-97
6. Crosti A, Gianotti F. Eruptive dermatosis of probable viral origin situated on the acra. *Dermatologica*.1957;**115**:671–7.
7. Sarma N, Sarkar UK, Das MK, Das S. Hemorrhagic Gianotti-Crosti syndrome in a one and half month old infant: an extremely unusual presentation. *Indian J Dermatol*.2013;**58**:65–7.
8. Linke M, Géraud C, Schneider SW, Goerdt S, Utikal J. Acute disseminated erythematous papulovesicular skin lesions in a 7-year-old child: a quiz. Diagnosis: vesicular Gianotti-Crosti syndrome. *Acta Derm Venereol*.2011; **91**:491-4.