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Gianotti-Crosti Syndrome: Atypical Presentation.

Kumar Arvind*, and Kumar Amrendra.

Department of Paediatrics, All India Institute of Medical Sciences, New Delhi, India

ABSTRACT

Gianotti-Crosti syndrome (GCS) is a viral-associated eruption that most commonly occurs in children aged 3 months to 15 years. It consists of monomorphic red-brown to pink papules and vesicles distributed symmetrically on the cheeks, extensor surface of the extremities and buttocks and spares trunk, palms and soles. The eruption usually spontaneously resolves over the course of 10 to 60 days. We report the rare case of GCS in a child without any prodromal symptoms, vaccination history or viral infection and no systemic involvement. An otherwise healthy 10-year-old child presented with a pruritic eruption of 10 days' duration on the dorsal aspect of her hands and feet, elbows, and knees and over buttocks and trunk. No lymphadenopathy was observed. A biopsy revealed histopathologic findings consistent with a diagnosis of GCS. The patient's aspartate aminotransferase (AST) and alanine aminotransferase (ALT) levels were normal. Over the course of the next 2 weeks, the patient's skin findings completely resolved with normalization of liver function tests. The clinical and histologic correlation was consistent with GCS in a child. Clinicians should keep GCS in their differential diagnosis when examining a child with trunkal lesion as well..

Key words: *Gianotti-Crosti syndrome (GCS)*, Papular acrodermatitis of childhood (PAC).

**Corresponding author*

INTRODUCTION

Papular acrodermatitis of childhood (PAC) or *Gianotti-Crosti syndrome (GCS)* was first described separately by Gianotti and Crosti in 1955[1]. *Gianotti-Crosti syndrome* is a self-limited disease that presents as an acute onset multiple, monomorphic pink to red-brown papules or papulovesicles, which may be slightly pruritic and can become confluent that measure between 2 to 510 mm in diameter, and persist for 3 to 5 weeks. Typically, the papules are localized on the face and limbs, with sparing of the flexural surfaces; they are nonpruritic, well circumscribed, nonrelapsing, and may Köbnerize. Occasionally, the papules may appear to be lichenoid or purpuric. They do not affect the mucous membranes[2].

Systemic findings may include malaise, low-grade fever, or diarrhea. In 25% to 35% of patients, lymphadenopathy can be found, usually favoring the cervical, axillary, or inguinal regions[3]. Hepatic involvement is uncommon; prevalence figures are not available. Presumably there is significant variation between industrialized and less-developed lands. The most usual finding is hepatomegaly. If hepatitis is present, it is usually anicteric. Splenomegaly is even more uncommon. Almost all patients are between 3 months and 15 years of age, with a peak between 1 and 6 years of age[4].

It has been linked to immunization or various viral infections, such as Epstein-Barr virus, cytomegalovirus (CMV), hepatitis B virus, coxsackievirus, human immunodeficiency virus (HIV), and parainfluenza. Numerous studies confirm that EBV is now the most common cause of GCS[5]. We are describing a case of *Gianotti-Crosti syndrome* in a child without recent history of vaccination or any prodromal symptoms suggestive of viral or bacterial infection.

Case Report

A previously healthy 10-year-old boy presented with 10 days history of a cutaneous eruption that initially appeared on his legs and subsequently progressed to affect his buttocks, arms and trunk.

Physical examination demonstrated an afebrile child with pinkish papules and papulovesicular eruption ranging from 2mm to 10 mm on the legs, buttocks, arms and trunk, mostly discrete but few are coalescent as well (figure 1-4). There was no history of prodromal illness, sore throat or recent vaccination. There was no lymphadenopathy or hepatosplenomegaly. Laboratory investigations revealed mild leukocytosis (white cell count, 9,600/mm³) with a normal differential. Liver function test and other routine tests were within normal limit. Based on the morphology of the lesions and sudden eruption in a child differential diagnosis of *Gianotti-Crosti syndrome* and papular urticaria was made and biopsy was sent in the lab.

Histopathologic examination of a skin biopsy specimen from the right buttock revealed a perivascular and somewhat interstitial lymphocytic infiltrate in the superficial and mid-dermis with intraepidermal exocytosis of lymphocytes, microabscesses of Langerhan's cells, spongiosis

and papillary dermal edema which was consistent with the diagnosis of *Gianotti-Crosti syndrome (GCS)*. He was treated with 2.5% hydrocortisone cream, and the eruption resolved.



Figure: 1



Figure: 2



Figure: 3



Figure: 4

DISCUSSION

We have observed that exceptionally long and short courses of *GCS* do exist. The rash duration in standard textbook descriptions is 14–56 days. But in this case we have observed a 10 days duration rash. The shortest duration of rash reported is 5 days [6]. In this case we have also observed few trunkal lesions as well. Trunkal lesion is considered as negative clinical feature for *GCS* but few trunkal lesions should not exclude the diagnosis of *GCS* [7].

Although lymphadenopathy is significantly correlated with a diagnosis of *GCS*, we found no lymphadenopathy in our case. We thus believe that lymphadenopathy should not be a mandatory positive clinical feature and should not be included in the diagnostic criteria [8].

Hepatic involvement is uncommon; prevalence figures are not available. It is caused not only by HBV, but also by EBV or CMV. The most usual finding is hepatomegaly. If hepatitis is

present, it is usually anicteric [9]. Splenomegaly is even more uncommon. In a recent Indian study, the reliability of the diagnostic criteria for CGS were assessed in 23 affected children and 74 control subjects; there was not a positive correlation between hepatosplenomegaly and CGS[8].

The peripheral blood may show modest lymphocytosis or lymphopenia. Occasionally the number of monocytes is increased, probably in patients with EBV[4].⁴

The histologic picture of GCS may be dramatic but is not diagnostic. Both vesicular and nonvesicular patterns can be seen. The vesicular variant shows striking epidermal changes with mild acanthosis accompanied by diffuse spongiosis and vesicles. The dominant cells in the vesicles are Langerhans cells. The papillary dermis features an intense lymphocytic perivascular infiltrate can be seen in many inflammatory dermatoses[10].

The course is benign. The lesions heal without scarring over 10 to 60 days; with or without treatment[11].

The diagnosis of GCS is usually straightforward. One should then be alert to the rare possibility of systemic signs and symptoms. If there is hepatosplenomegaly or malaise, viral hepatitis should be excluded, especially in countries where immunization programs are not standard. Therapy is rarely needed. Most important is to reassure the parents that GCS is a benign, harmless disease in more than 99% of cases.

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