Gianotti-Crosti syndrome as the only manifestation of primary Epstein-Barr virus infection: a case report

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Gianotti-Crosti syndrome (GCS) presents as a symmetric erythematous papular and papulovesicular eruption of the face, extremities, and buttocks, and usually occurs in young children. GCS has been classically associated with hepatitis B virus, as well as rarely with Epstein-Barr virus (EBV) and cytomegalovirus. We report a case of GCS related with EBV infection without any systemic symptoms in a two-year-old male patient who presented with symmetric red papular eruptions on the cheeks, buttocks and both upper and lower extremities.

Key words: Gianotti–Crosti syndrome, Epstein-Barr virus infection, rash.

Gianotti-Crosti syndrome (GCS), first described by Gianotti in 1955, is a self-limiting symmetric papular eruption often associated with an underlying viral infection[1,2]. GCS occurs several weeks after resolution of the viral infection responsible for producing the rash. Papules are symmetrically localized to the acral extremities, face and buttocks. Mucous membranes, trunk, palms and soles are spared, but the trunk can occasionally show a few lesions, so that its involvement does not exclude the diagnosis of GCS[3,4]. Lesions typically persist for three to six weeks. Resolution occurs spontaneously and does not produce scarring. It is thought that GCS results from a pattern of immune dysregulation[4]. Recent reports indicate that a variety of infectious agents are associated with GCS. Though hepatitis B virus is known to be the most common etiologic factor, Epstein-Barr virus (EBV) infection does not seem to be rare.

In this report, a two-year-old male patient who was diagnosed with GCS caused by acute EBV infection without any systemic symptoms was presented with red papular eruptions symmetrically on the cheeks, buttocks, and both upper and lower extremities without clinical symptoms. We would thus like to emphasize via this case that asymptomatic EBV infection might present as GCS.

Case Report

A two-year-old male was referred for evaluation of a rash that had been present for two weeks. Past history revealed a short-lived upper respiratory tract infection several weeks before the onset of the rash. The rash had begun on the cheeks and spread to involve both upper and lower extremities and buttocks. Red papular eruptions measuring about 2-4 mm in diameter appeared symmetrically and the trunk was spared (Figs. 1 and 2). The patient did

Fig. 1. Symmetrically distributed erythematous papules measuring approximately 2-4 mm in diameter are seen on both cheeks.

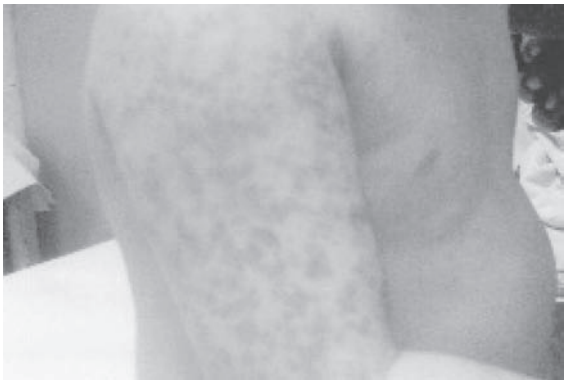

Fig. 2. Erythematous papules measuring approximately 2-4 mm in diameter are seen on the extremities.

not have fever, nasal discharge or coughing. Major complaint was mild pruritus. On physical examination, throat and auscultation of the heart were normal. There was no hepatosplenomegaly or lymphadenopathy.

The complete blood cell count and liver function tests were in normal range. Serology was negative for hepatitis B surface antigen, hepatitis A and C virus antibody, cytomegalovirus (CMV) IgM antibody and parvovirus B-19 IgM antibody. The EBV serology results were as follows: EBV viral capsid antigen IgM positive (titer of 5.6, normal: 0-1.1), EBV VCA IgG positive (titer of >200), EBV early antigen IgG positive (titer of >200), and EBV-associated nuclear antigen (EBNA) IgG negative, consistent with an acute onset EBV infection. The rash disappeared over a period of four weeks without any medical intervention.

Discussion

Gianotti-Crosti syndrome is often associated with underlying viral and bacterial infection such as hepatitis A and B viruses[6-8], EBV[2,5], CMV[9], human immunodeficiency virus (HIV)[10] and some immunizations such as polio, diphtheria, influenza, pertussis and measles[11,12]. Although the hepatitis B virus was previously thought to be the main cause of GCS, the incidence of associated infections may depend in part on the different geographic distributions of infectious organisms[5]. For example, in Japan and Mediterranean countries, GCS is commonly associated with hepatitis B virus infection, whereas EBV is the most common etiologic factor documented in the United States[6,7]. We confirmed that our two-year-old male patient with red papular eruptions symmetrically on the cheeks, buttocks, and both upper and lower extremities associated with GCS was caused by primary EBV infection without any systemic symptoms such as hepatomegaly or lymphadenopathy.

Primary infection with EBV during childhood is usually indistinguishable from other childhood infections. Major clinical manifestations, which are rarely apparent in children under four years of age, are fatigue, pharyngitis and generalized lymphadenopathy[2,3,5]. However, as seen in our patient, GCS may also be related with acute EBV infection without any systemic symptoms. The diagnosis could be confirmed by serologic studies for IgM and IgG in the absence of positive serology for other viral infections and no history of recent immunization[5].

Localized facial rash alone is an alternative clinical picture of GCS-associated EBV infection. Yoshida et al.[13] reported five patients who had symmetrical rashes on their cheeks as associated GCS caused by primary EBV infection. However, those patients had systemic symptoms such as hepatomegaly, increased liver function tests, and cervical lymphadenopathy. On the contrary, there was no finding suggesting EBV infection in our patient.

In view of the routine vaccination programs in Turkey, other rare causes of GCS should be considered as the primary causative agent. Since primary infection with EBV seen in early childhood may present with no systemic symptoms, the rash may be the only unique manifestation. Thus, in such patients, EBV and other rare infections should be considered in the differential diagnosis of GCS.

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