Gianotti-Crosti syndrome due to Epstein-Barr virus infection with possible role of palivizumab.

Key words: Gianotti-Crosti syndrome, EBV infection, palivizumab.

A 16 months old girl with a congenital atrial septal defect was admitted to our emergency department (ED) with a history of a “rash” in the last 7 days. There was no history of fever or preceding illness. However, she had undergone palivizumab passive immunization 2 weeks before the development of the first symptoms. Initially, the “rash” was characterized by symmetrical pink papules on the extensor surface of both legs. Five days later, the eruption affected the upper limbs with multiple, pruritic, red-violaceous papules coalescing into plaques on both lower extremities and elbows. Physical examination at ED showed 2 mm in size, confluent, symmetrical and monomorphic red-violaceous papules on the extensor surface of both legs, knees and elbows, sparing the trunk, palms and soles.

The liver enzymes were within normal limits as well as antibodies for cytomegalovirus (CMV), Herpes, Parvovirus and polymerase chain reaction (PCR) for toxoplasmosis and Mycoplasma pneumoniae. The Epstein-Barr virus (EBV) IgM antibodies (viral capsid antigen - VCA - IgM) were positive, while VCA IgG and EBV nuclear antigen (EBNA) were negative.

Three weeks later, the EBV antibody tests were repeated to confirm the infection: the VCA IgM was positive, VCA IgG and EBNA were negative. The eruption started to resolve 6 weeks after hospital admission; 8 weeks later the skin was back to normal without scars or depigmentation.

Discussion

Gianotti-Crosti syndrome (GCS) was first described by Ferdinando Gianotti in 1955 and in the following year by Crosti and Gianotti (6). GCS was originally called papular acrodermatitis of childhood (8) because characterized by the abrupt onset of 1 to 10 mm in diameter, pink-brown, flat-topped papules, more densely located on the face, buttocks, feet, and extensor aspects of forearms and legs (1). Initially, GCS was supposed to be associated only with hepatitis B virus (HBV) infection in children. Later, Gianotti and Crosti described a different viral form, non-HBV, the infantile acrolocated papulovesicular disease (5). In 1992 Caputo et Al. (2) evaluated a series of 308 children with GCS: 239 (77.6%) had non-HBV viral infections and only 69 (22.4%) had a confirmed HBV infection. The Authors were not able to differentiate clinically the two forms thus recommending the use of the term Gianotti-Crosti syndrome as previously suggested (13).

The distribution of lesions in GCS is characteristic. Usually the trunk, knees, elbows, palms and soles are spared. However, their involvement does not exclude the diagnosis (4). The resolution of the lesions usually starts after 6-8 weeks (3), without leaving scars, and the prognosis is excellent, with rare complications (1).

Because of its self-limited nature GCS is underdiagnosed, making the incidence of the GCS unknown (1). It primarily occurs in children between 3 months and 15 years of age, with a peak between 1- to 6-years old (1). However, adult cases with a worse prognosis were reported.

The exact pathogenesis is still unknown. However, a delayed hypersensitivity reaction to viral infections (9) was hypothesized. Familiar or individual history of atopy may also play an important role (11).

The list of conditions associated with CGS is large. It includes several infectious agents, above all viruses – EBV (14), CMV (14), hepatitis A and B virus (12), Coxsackie virus (14) and many
more -, but also bacteria and immunizations with live, killed, and recombinant vaccines (7).

Some of these children had a clinical or subclinical viral infection at the time of vaccination (10), suggesting that the risk of developing GCS is greater in these cases (10). In our case, the child had a previous immunization with palivizumab and a subclinical viral infection with EBV, supporting the previously reported evidence of the role played by concomitant factors (10).

References


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