Gianotti Crosti Syndrome: What does it mean for a Family and for a Paediatrician?

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Abstract

Viral exanthems are very common in children. They are a big concern for both the parents and the treating paediatricians and some of them turn out to be a diagnostic challenge for the unwary Paediatrician. Accurate diagnosis goes a long way in reassuring parents with a child with fever and rash, especially in epidemics of diseases. Gianotti-Crosti syndrome is a common, self-limiting dermatosis that occurs worldwide, but it is not often diagnosed as it is not thought of - many a time by the treating Paediatrician. It affects infants and children between the ages of 6 months and 12 years of age, with peak incidences occurring from ages 1 to 6 years.

We report our cases to highlight the frequent occurrence of the disease, which may give a relief to both the parents and the treating Paediatricians.

Keywords: Rash; Gianotti Crosti Syndrome; Parental anxiety; Vesicles; Varicella

Abbreviations: OPD: Outpatient Department; GCS: Gianotti Crosti Syndrome; EBV: Epstein Barr Virus

Introduction

Children with rash and eruptions form substantial part of pediatric OPD attendance. Most of them are benign viral exanthems without much clinical significance [1], however rashes in children are source of parental anxiety. Fever accompanied by rash is a common finding in pediatric patients. Although, in most cases the disease is trivial, in some cases it may be the first and/or the sole manifestation of a serious and life threatening condition in patients. The spectrum of differential diagnosis is broad and many different infectious and some noninfectious agents cause this syndrome [2]. Gianotti-Crosti syndrome, also known as infantile papular acrodermatitis or papular acrodermatitis of childhood, is a common, self-limiting dermatosis that occurs worldwide. It affects infants and children between the ages of 6 months and 12 years of age, with peak incidences occurring from ages 1 to 6 years [3]. In 1955, Gianotti and Crosti initially described the syndrome as a distinct infectious exanthem associated with the hepatitis B virus [4].

Case Reports

We came across three cases of the syndrome in our OPD in the last 6 months.

First case is a 4.5 year old boy – born to an Indian mother with a Caucasian husband – who had an URTI following which he developed rashes on the extremities mainly on the upper limbs and few on the lower limbs, with NO lesions noted on the trunk. Initially few Pediatricians had seen him and he had been diagnosed with various diseases including Chicken pox, allergic rashes, streptococcal infections etc. Finally he was seen by a Pediatric Dermatologist who clinically suspected it to be Gianotti Crosti Syndrome and biopsied the lesions. Finally the diagnosis of Gianotti Crosti syndrome was made based on histological findings of papillary dermis edema and exclusion of other entities. It came as a big relief for the family and also the treating pediatrician as they had been running from Pillar to post for 3 weeks by then.

Second case is a four year old boy who came with rash of 12 days duration with symptoms of upper respiratory tract infection. Rash was monomorphic red-brown to pink papules and vesicles distributed symmetrically on the extensor surface of the extremities but absent in face and buttocks. The rash was annoyingly itchy for the child. Parents were extremely worried about persisting rash for 3 weeks with the possibility of Varicella as told by G.Ps and any consequences arising thereof. They had consulted 4-5 doctors because no one could offer a proper counseling. A diagnosis of Gianotti Crosti Syndrome was made during OPD visit based on the clinical features and parents were counseled with evidence of literature regarding the benign nature of illness (Figure 1) – which was a sigh of relief for the parents as they had spent nearly 4000 INR on various medications by then. Currently the child is well with no stigmata and all the rashes have disappeared (Figure 2).

Third case is a two year old girl who came in OPD with rash primarily for 9 days. She was treated as “Varicella” from outside. The rash was pruritic and the vesicles discharged serous fluid. There was H/O fever two weeks back. The discharging vesicles left...
The disease is an eruption consisting of monomorphic almost exclusively female, with five documented cases affecting females with equal frequency unlike adults where it affects patients [5]. In the pediatric population, GCS affects males and adults, although incidence is much more common in pediatric with more than 90% younger than four years. It also occurs in that commonly affects children three months to 15 years of age, on the face, dorsal aspects of both upper and lower extremities and buttocks but spared trunk totally which is against the characteristic rash of Varicella. On parental request Varicella serology (ELISA) was done and reported negative in the light of vaccination status of the child. Clinically it behaved like Gianotti Crosti Syndrome and the rashes disappeared after four months, currently the child doing well.

Discussion

Gianotti Crosti syndrome (GCS) is a sporadic dermatosis that commonly affects children three months to 15 years of age, with more than 90% younger than four years. It also occurs in adults, although incidence is much more common in pediatric patients [5]. In the pediatric population, GCS affects males and females with equal frequency unlike adults where it affects almost exclusively female, with five documented cases affecting men [5,6]. The disease is an eruption consisting of monomorphic red-brown to pink papules and vesicles distributed symmetrically on the cheeks, extensor surface of the extremities, and buttocks; the trunk is strikingly spared, although a transient eruption can occur. Involvement of the trunk does not exclude the diagnosis. Rash may be pruritic, [3] hemorrhagic or edematous to the point of forming vesicles and is occasionally confused with Varicella. The mucous membrane is not involved [7] and the face may be the only area of involvement [8,9]. Hepatosplenomegaly and axillary or inguinal adenopathy are inconsistent findings although lymphadenopathy may be more common on physical examination than hepatosplenomegaly [9,10]. Symptoms related to the primary viral syndrome or underlying bacterial infection may include mild constitutional symptoms such as low-grade fever and malaise, pharyngitis and/or mucosal lesions, or symptoms of an upper respiratory tract infection [10].

The most likely explanation for the exanthem is a local type IV hypersensitivity reaction to the offending viral or bacterial antigen within the dermis. This is based on the immunohistochemical characterization of the cutaneous inflammatory infiltrate suggesting a reactive process other than an autoimmune phenomenon or direct infection of the skin [7]. Inciting factors include various viral and bacterial infections, as well as recent immunizations [11]. The rarity of GCS in adults suggests lifelong immunity to a common viral triggering agent [9]. GCS is more common among children with atopic dermatitis again suggesting an immune mechanism. Occurrence of GCS following Immunization has been frequently described in case reports [11,12,13].

In general, no laboratory studies are needed in patients with Gianotti-Crosti syndrome. It is a diagnosis that can be made on clinical grounds with doubtful cases requiring a skin biopsy. Jaundice or hepatomegaly should prompt a search for the hepatitis B virus and elevated levels of liver enzymes. However, elevated levels of liver enzymes are most likely related to Epstein-Barr virus infection. Certain histology types like abundance of cytotoxic T cells in the inflammatory infiltrate favor EBV infection [14]. Close differential diagnosis includes Hand foot and mouth disease, Molluscum contagiosum, Pityriasis rosea and Scabies [15].

Gianotti-Crosti syndrome is a benign self-limited condition and requires no specific treatment. Topical steroids are generally not effective, although anecdotal responses have been reported. Systemic treatment with antihistamines has been moderately helpful in relieving pruritus [9].

Prognosis is excellent but lesions might take 4-12 weeks to clear up. Long-term complications are usually not known to be associated with GCS but it’s not uncommon to have high degree of parental anxiety for lengthened rash. In rare cases, chronic liver disease can occur after the initial phase of infection with hepatitis B virus [10]. Correct diagnosis and counseling helps in allaying the parental anxiety and drug overuse with this benign condition.

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References


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