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SCALP AND FACE EDEMA IN SCHÖNLEIN-HENOCH SYNDROME*

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Schönlein-Henoch syndrome, or anaphylactoid purpura, is a generalized vascular disorder resulting from acute aseptic vasculitis involving arterioles and capillaries. It is generally considered to be related to hypersensitivity, although a definite allergen is rarely identified[1].

The clinical picture of this syndrome is dominated by a hemorrhagic, maculopapular rash which appears in recurrent crops and which characteristically involves the buttocks and extensor surfaces of the legs and arms. Other features of the syndrome are recurrent, colicky abdominal pain, mild to moderately severe arthralgia, and renal involvement manifesting as hematuria. The clinical course is variable, but the majority of children recover completely. In some cases, classical symptomatology is not present. Therefore, an early diagnosis is rather difficult[1].

Although painful edema of the scalp in Schönlein-Henoch syndrome is mentioned in current general pediatric textbooks[1,2], we would like to describe this lesser known symptom, which was observed in our two cases both prior to and after the onset of the rash.

Case Reports

Case 1

A three-year-old boy was admitted to the Hacettepe Children's Hospital with a four-day history of abdominal pain, vomiting and a rash. There were no complaints about the joints. On examination he was found to have a hemorrhagic-urticarial rash over the lower limbs and buttocks. Blood pressure was normal. There was slight abdominal tenderness. Urinalysis revealed no abnormalities. Stools were negative for occult blood. The erythrocyte sedimentation rate was 55 mm/hour. The results

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of the following investigations were normal or negative: complete blood count, tests for LE cells, antinuclear and anti-DNA antibodies, rheumatoid factor, immune complexes, serum C3 level, antistreptolysin O titer, C-reactive protein and throat culture.

On the seventh day the patient developed periorbital edema, first around the left eye, then spreading rapidly to involve both eyes, the face, lips and forehead, and finally the entire scalp was swollen and very tender (Figure 1). He remained afebrile. There was also edema on the neck. He was hospitalized owing to the danger of airway obstruction. After treatment with diphenhydramine hydrochloride (5 mg/kg/day), edema and tenderness of the face and neck soon subsided, but swelling of the scalp persisted for two days.

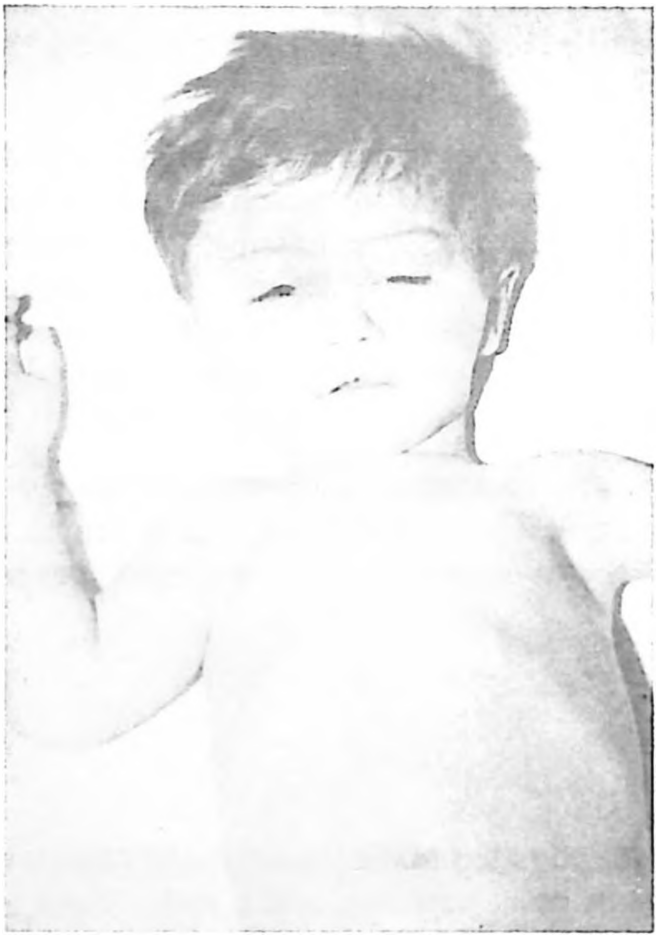

Figure 1.

Scalp and face edema (Case 1).

Case 2

An eight-year-old girl was brought to the emergency unit with a four-day history of abdominal pain and facial swelling. On the day of admission she had no rash on her

body. It was learned that the edema had first appeared on the left side of the face, scalp and forehead. On the next day the swelling had spread to the other half of the head and was very tender. Moderate abdominal tenderness was present. The rest of the examination was non-contributory. On the fourth day she developed an extensive urticarial rash on the backs of the legs, thighs and buttocks. Both feet were edematous and there was painful swelling of both ankle joints. At this point, Schönlein-Henoch syndrome was diagnosed. Urinalysis was normal. Stools were negative for occult blood. The blood pressure remained normal throughout the follow-up period. During the edematous phase of the disease diphenhydramine hydrochloride (5 mg/kg/day) treatment seemed to be helpful, and the swelling and tenderness disappeared in two days.

Discussion

Schönlein-Henoch syndrome may occur without the characteristic purpuric rash or with manifestations limited initially to internal organs and with a rash appearing sometime after the onset of other symptoms. Edema is present at some time during the course of the illness. Most commonly it is present on the legs and ankles and coincides in time with the development of the skin rash but is not necessarily of the same duration[4]. Localized swelling of the dorsum of the hands and feet, and occasionally the scalp, ears and periorbital region may be noted[5]. Most children with scalp edema also have edematous areas on the extremities, again emphasizing the greater tendency for development of edema in the young patient. In most patients there can be other possible causes for the swelling, such as hypoalbuminemia, renal disease, cardiac failure, venous thrombosis or arthritis, but the only possible explanation for the edema in Schönlein-Henoch syndrome seems to be that it is due to cutaneous vasculitis[4]. The diagnosis is usually not difficult if the classical rash is present. However, it is important to realize that other symptoms of the syndrome can be present long before and after the occurrence of typical manifestations. Therefore, swelling of the scalp and face prior to the appearance of the rash, as observed in our second case, is an important sign. The swelling in the first case may be considered as an interesting feature of the disease since it appeared seven days after the occurrence of the rash. The diagnosis of such conditions can be very difficult unless the pediatrician has a high index of suspicion. Patients with Schönlein-Henoch syndrome showing facial and/or scalp edema can be misdiagnosed, as pointed out in a previous publication[6], since this unusual symptom may make the diagnosis difficult. In some cases, the scalp may become swollen enough to produce a grotesque deformity. Periorbital and scrotal swellings tend to be of short duration, but can be quite painful[7]. A careful history should be taken every time one is confronted with this symptom, and there should be an examination for the rash and other manifestations of the Schönlein-Henoch syndrome.

Edema of the face and scalp is said to occur mainly in the very young, or those under 3 years of age[3,5]. Since one of our patients was 8 years old, this case can be considered as unusual. To the best of our knowledge, there is only one other case[8] recorded in the literature. The real incidence of scalp and face edema in Schönlein-Henoch syndrome is not known, but in one report it was found in only 25% of 131 children[6]. However, in another study one in four were reported to have had facial or scalp edema[5]. Other reports contained only case presentations with emphasis on this symptom[3,8].

The treatment of scalp and face edema has been documented in the literature[5]. Indications for corticosteroid therapy in Schönlein-Henoch syndrome include severe and uncomfortable localized soft tissue swellings, particularly those about the scalp and eyes, painful joint involvement, abdominal pain and melena. More impressive improvement following corticosteroids was noted in a group of patients who were treated for soft tissue swelling or joint symptomatology, but one patient developed a subarachnoid hemorrhage during corticosteroid therapy for painful scalp edema[7]. An antihistaminic agent, diphenhydramine hydrochloride, seemed to relieve the edema of both our patients, and this is in accordance with the theory that the etiology of Schönlein-Henoch syndrome is related to hypersensitivity.

Summary

Scalp and face edema are described in two patients with Schönlein-Henoch syndrome. In one, abdominal pain and swelling of the face and scalp were noted for one week prior to the onset of the rash and joint involvement, and in the other swelling was seen seven days after the rash. We believe that the possibility of anaphylactoid purpura should be considered in the differential diagnosis of children with abdominal pain and localized edema on the head prior to the onset of rash. Antihistaminic agents seemed to be useful in the relief of edema.

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