PEDIATRIC DERMATOLOGY PHOTOQUIZ

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Acute Onset of Generalized Pruritic Rash in a Toddler

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Case Report

A 2-year-old boy presented to the Emergency Room with a 4-day history of a rash and a 1-day history of fever. The eruption began on his back and rapidly spread to become generalized over 24 hours. He was sleeping poorly due to severe pruritus, and his appetite was diminished. No response occurred to treatment with oral antihistamines or prednisolone. His previous medical history included asthma, controlled with intermittent use of a salbutamol inhaler, and dermatitis from contact with sleeper snaps in infancy. His mother had recently taken an oral antibiotic for presumed streptococcal pharyngitis.

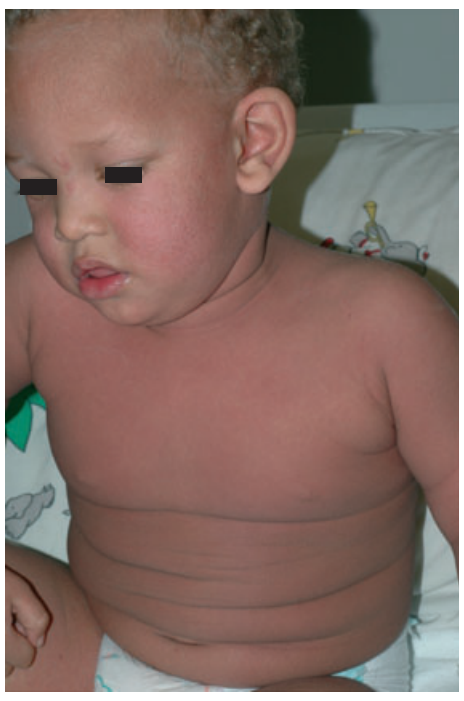

Figure 1.

On examination, his temperature was 38.3°C. He was irritable and scratching. His skin was erythrodermic with coalescing 2 to 5 mm edematous red papules involving the trunk, extremities, face, and ears (Fig. 1). The perioral skin, palms, and soles were spared. The oral mucous membranes and conjunctivae were unremarkable. Physical examination otherwise revealed only mild cervical lymphadenopathy.

A complete blood count was normal except for the presence of atypical lymphocytes (2.13×10^9/L; normal range < 0.01) and an elevated eosinophil count (1.13×10^9/L; normal range < 0.50). C-reactive protein and erythrocyte sedimentation rate were normal. Throat culture was negative. A diagnostic test was performed.

What is the diagnosis?

Hyperkeratotic Papules in a Child with Down Syndrome

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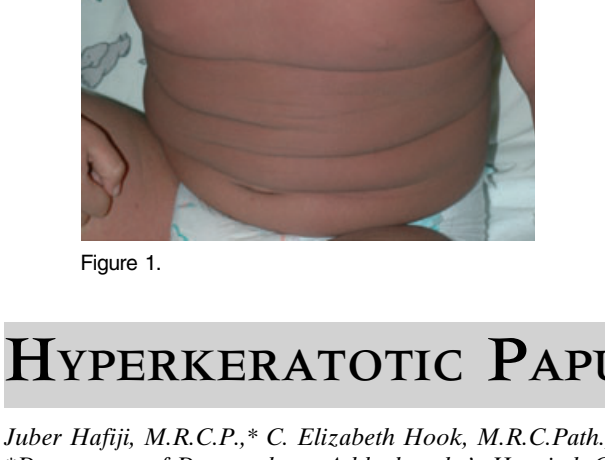
Case Report

A 15-year-old boy with Down syndrome (DS) presented with a 12-month history of multiple, asymptomatic papules on the extensor aspect of the limbs and buttocks. The mother and patient denied any scratching. A 1-month trial of Tetracycline 250 mg BD had no effect, and a combination of betamethasone valerate and fusidic acid (Fucibet®, Leo Pharm, Princes Risborough, Buckinghamshire, UK) was unhelpful. No relevant family history was known. Examination revealed multiple 5 to 10 mm erythematous papules and nodules with central keratin plugs on the upper and lower limbs and buttocks, some occurring in a linear fashion (Figs. 1 and 2). Folliculitis was evident on the buttocks and to a minor degree on the thighs. Laboratory investigations were all normal or negative including full blood count, renal and liver function, bone profile, glucose, inflammatory markers, thyroid function, antinuclear antibody (ANA), complement, and immunoglobulins. Skin swabs revealed no growth. Skin biopsies from the arm and thigh showed similar features (Figs. 3 and 4).

What is the diagnosis?



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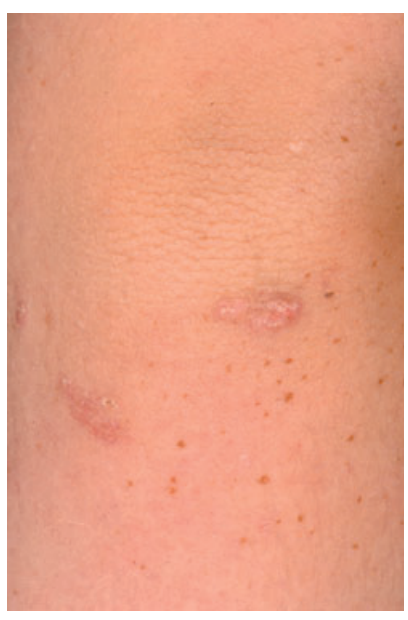

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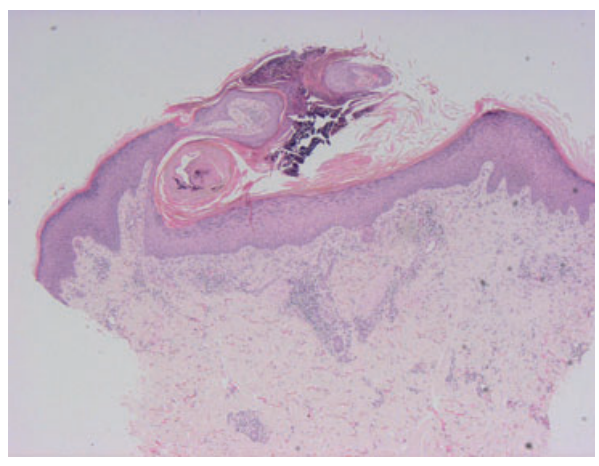

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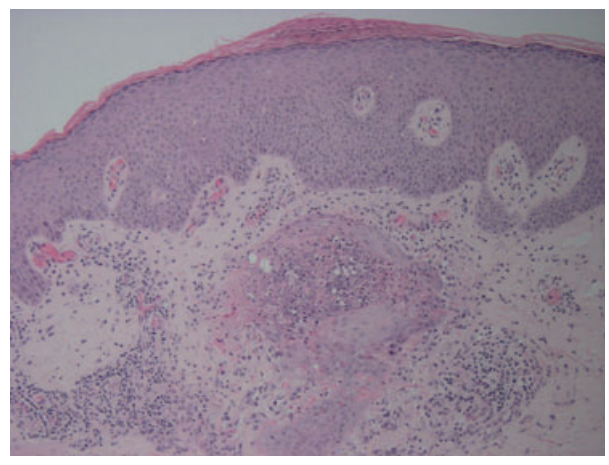

Figure 4.

Multiple Translucent Papules on the Nose of a 5-Year-Old Boy

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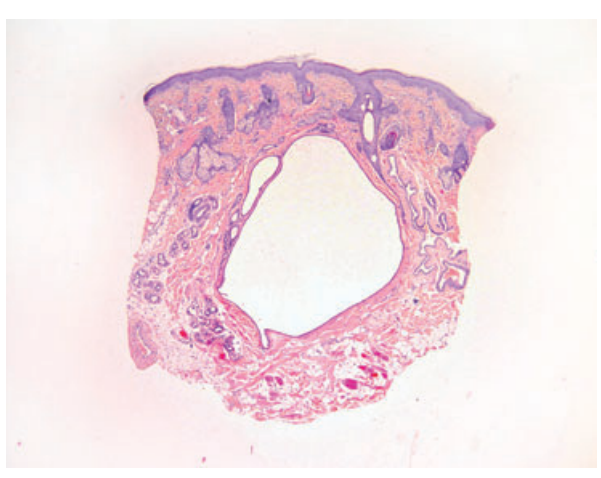
Case Presentation

A 5-year-old boy presented with approximately 15 skin-colored to bluish, translucent papules of 1–3 mm diameter scattered over the dorsum of his nose (Fig. 1). The parents had first noticed them 2 years previously and reported a marked increase in number and size after physical exercise as well as seasonal variation, with exacerbation during summer months (Fig. 2). The boy was said to sweat easily, especially in the craniofacial area. He was small, with height and weight just above the 3rd percentile, but was otherwise in good health and on no medication. Laboratory parameters including thyroid hormones and autoantibodies revealed no pathological findings. A facial Minor's iodine-starch test showed pronounced sweating on the forehead, cheeks, and chin, after physical exercise, while the nose was largely spared. Correspondingly, gravimetric assessment of sweat production showed it to be remarkably high on the forehead while being within normal range on the palms. After informed consent from the parents, one of the papules was removed by punch biopsy and histologically examined (Figs. 3 and 4).

What is the diagnosis?



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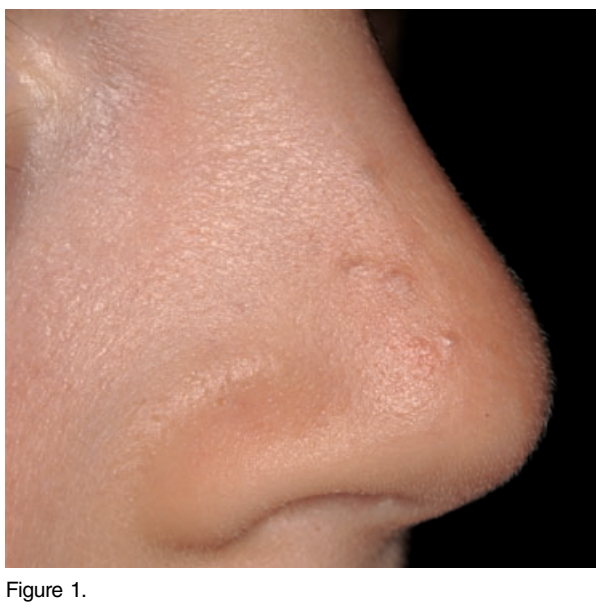

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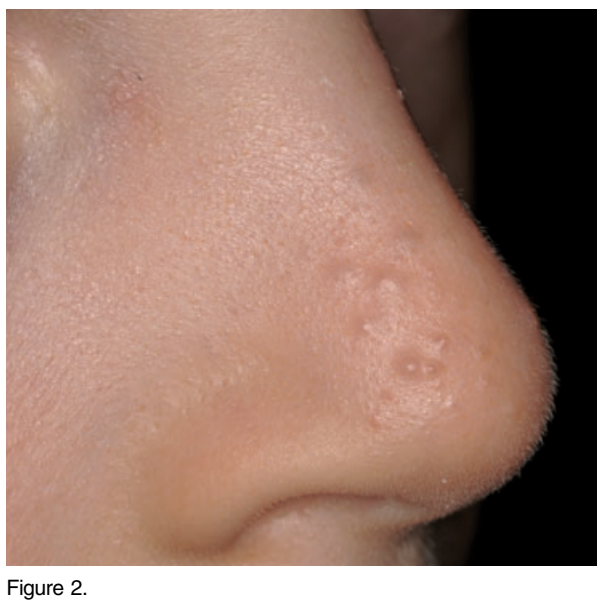

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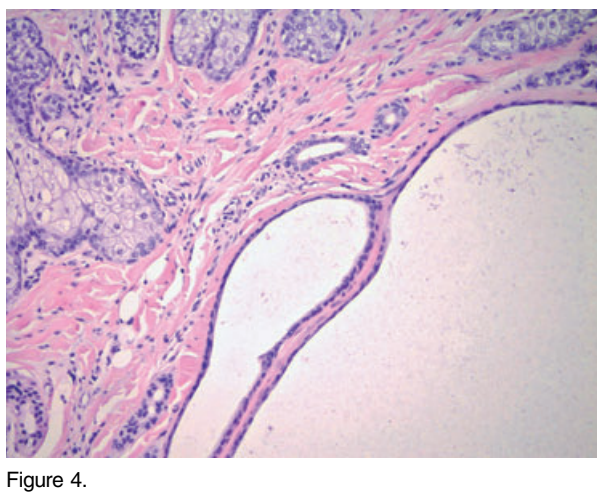

Figure 4.

Acute Onset of Generalized Pruritic Rash in a Toddler

Diagnosis: Systemic allergic (contact) dermatitis to nickel from ingestion of metal coins

Radiographic Findings and Clinical Course

The generalized acute dermatitis without localizing features and history of dermatitis from contact with sleeper snaps prompted a question about ingestion of metal. His mother recalled a recent episode when he was found putting coins in his mouth. An abdominal radiograph revealed coins in the region of the gastric antrum (Fig. 2). An initial endoscopy identified two coins entrapped by a surrounding thick network of fibrin threads, which also obscured most of the gastric lumen. An attempt to remove the coins failed. He was administered intravenous methylprednisolone 2 mg/kg/day in divided doses. On the following day, a repeat endoscopy revealed normal appearing gastric mucosa and a Canadian quarter (25 cents) and a nickel (five cents) were easily retrieved. He received a further 3-day course of oral prednisolone 20 mg daily, and the skin eruption resolved completely. One year later, he developed acute generalized dermatitis again, and a nickel was removed from his stomach. Patch testing was considered unnecessary.

Discussion

Nickel is a common cause of allergic contact dermatitis in children. Positive patch tests to nickel have been found in infants as young as 6 months of age (1). Sensitization in infants and young children may result, as in our patient, from contact with metal in clothing snaps and other fasteners worn next to the skin (2). Jewelry and early ear piercing may also play a role (3).

Systemic (contact) allergic dermatitis occurs when nickel sensitive individuals develop dermatitis following systemic exposure to the metal (3). Ingestion of 4 mg of nickel is expected to result in widespread dermatitis in most nickel-allergic adults, and low nickel diets are sometimes recommended for nickel-sensitive patients with refractory dermatitis (3). Other potential sources of systemic exposure include corroded orthopedic devices, endovascular stents and canulae, and orthodontic appliances.

Acute generalized dermatitis from swallowing coins was reported previously in three children, aged 8, 6, and 8, respectively (4–6). In each case, the dermatitis developed within 24 hours of ingesting the coin. Two reports described inflammation of the gastric mucosa; in one child, the coin was embedded in edematous gastric mucosa and proved impossible to dislodge by endoscopy, necessitating surgery (4). A similar situation in our patient was resolved by administration of intravenous systemic steroids, which reduced the mucosal inflammation, and the coins were successfully removed. As in our case, two children had associated fever, and one had a high circulating eosinophil count (4,5). All three children had a prior clinical history of nickel dermatitis. Patch testing to nickel was performed in two cases, and this was positive (4,5). Two children tested had high serum nickel levels (5,6).

It is noteworthy that the coin in each case was a Canadian quarter or nickel. From 1968 to 1999, Canadian quarters and dimes were minted from 99.9% nickel and nickels from 25 to 99.9% nickel (7). The extracted coins were heavily tarnished. It is likely that release of nickel from corrosion by gastric acid resulted in the rapid onset of local mucosal and systemic inflammation. Early coin removal by endoscopy is recommended because of the systemic reaction and because mucosal inflammation may prevent spontaneous passage of the coin through the gastrointestinal tract (6).

Unlike the three previous cases, our patient was too young to tell his parents or physicians he had swallowed a coin. Systemic allergic (contact) dermatitis from ingestion of a metal object is worth considering in the differential diagnosis when a nickel-sensitive toddler presents with an unexplained acute generalized dermatitis.

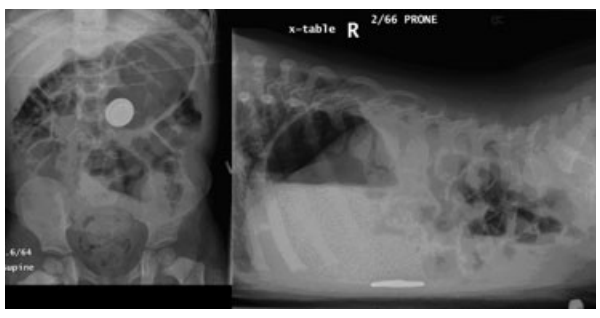

Figure 2.

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Hyperkeratotic Papules in a Child with Down Syndrome

Diagnosis: Acquired reactive perforating collagenosis in Down syndrome

Microscopic Findings

Histology revealed focal parakeratosis and necrotic debris within the dermis including collagen fragments, associated with an infolding of the epidermis. The collection of debris communicated with and was extruding through the epidermis. No evidence of folliculitis existed.

Discussion

Reactive perforating collagenosis (RPC), first described in 1967 (1), is an uncommon dermatosis of multifactorial aetiology that clinically presents as spontaneously involuting, skin-colored, or erythematous papules with a scab-like central plug. Two forms have been described: an inherited autosomal recessive form and a sporadic acquired form. The former occurs in childhood, and the latter occurs in adults and is associated with systemic diseases, in particular, diabetes mellitus (DM) and renal failure (2). Other reported associations include lymphoma, acquired immune deficiency syndrome (AIDS), hypothyroidism, hyperparathyroidism, neurodermatitis, liver dysfunction, atopic eczema, malignancy, scabies, and lung fibrosis (3).

The pathogenesis of acquired RPC (ARPC) is poorly understood. Lesions are often linear, and can be reproduced by scratching the skin (Koebner-phenomenon), and, therefore, ARPC may represent a cutaneous response to superficial trauma. The association of ARPC and DM has led to the suggestion that scratching causes microtrauma and necrosis of the dermal structures, possibly because of diabetic microangiopathy (4). Transforming growth factor–β (TGF-β), a superfamily of multifunctional peptide growth factors, has also been implicated (5). Transforming growth factor–β3, the most abundant isoform, is involved in the initiation and progression of apoptosis in the normal epidermis, suggesting a specific role of TGF-β3 in epidermal homeostasis: it therefore may be important for tissue remodelling in ARPC.

Individuals with DS are more susceptible to cutaneous infections including furuncles, abscesses, impetigo, and folliculitis; the latter having been reported to occur most commonly (6). Immunological disturbances are common in DS, with defects in antibody and cell-mediated immunity and phagocytic function, resulting in reduced in vitro killing of staphylococcus aureus (7). Pruritus was not a feature in this case, and we postulate that the background mild folliculitis in our patient could be a trigger for developing ARPC, although this was not confirmed on histology.

Various treatments have been tried for ARPC. These include topical and systemic steroids, topical and oral retinoids, antibiotics, phototherapy, methotrexate, and allopurinol (3,8). Allopurinol is thought to be effective via its antioxidant effects and tetracycline antibiotics via their potent inhibitory function on leucocytes and matrix metalloproteinases.

Down syndrome is associated with a variety of dermatoses. Advances in medical care have led to an increased life expectancy, and, with the increasing number of individuals with DS, dermatologists are more likely to encounter the wide spectrum of skin diseases in these patients. To our knowledge, this is only the second reported association of DS and ARPC in the literature (9). Acquired reactive perforating collagenosis is probably an underdiagnosed condition, and this case illustrates the importance of recognizing rare dermatoses in individuals with DS.

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Multiple Translucent Papules on the Nose of a 5-Year-Old Boy

Diagnosis: Multiple eccrine hidrocystomas

Microscopic Findings and Clinical Course

Histological examination of a papule showed a unilocular cyst in the middermis. The cyst wall was composed of two layers of cuboidal or flattened epithelial cells. Neither decapitation secretion nor periodic-acid Schiff (PAS)-positive, diastase-resistant granules were detectable. Immunohistochemical staining for human milk fat globulin 1 (HMFG-1) and smooth muscle antigen (SMA) were negative, fulfilling the criteria for an eccrine hidrocystoma.

Nightly treatment with 15% aluminum chloride hexahydrate aqueous solution for 4 months led to partial regression of the cysts without adverse effects. However, improvement due to seasonal variation cannot be ruled out.

Discussion

Eccrine hidrocystomas are benign cystic lesions thought to result from blockage of the sweat ducts and consequent dilatation of their secretory portion. Two typical clinical presentations are differentiated: multiple translucent, dome-shaped papules of 1 to 6 mm size on the periorbital and malar areas of middle-aged to elderly women, exposed to heat and a humid environment (Robinson type) (1), and larger, solitary lesions mostly on the eyelids of adults without sex preference (Smith and Chernosky type) (2). Less frequently involved sites include other parts of the head and neck, chest, axillae, palms, and popliteal fossae. Swelling after physical exercise and seasonal variation with exacerbation in summer are often noted.

To our knowledge, multiple eccrine hidrocystomas have not previously been reported in preschool children. In view of their assumed pathogenesis, facial hyperhidrosis may have contributed to its manifestation in our case. Reduced sweating on the nose itself may be explained by the blockage of the ducts of visible, and possibly a larger number of subclinical, lesions.

Clinical and histological differential diagnoses of multiple nasal papules in young children include epidermal inclusion cysts, mucoid cysts, lymphatic malformation, milia, closed comedones, pseudoacne of the nasal crease, and apocrine hidrocystoma. The latter usually presents as a larger, solitary, brownish or bluish cyst with its size not influenced by environmental factors (2). Histologically, apocrine hidrocystoma is characterized by a lining of myoepithelial cells and secretory cells with decapitation, and immunohistochemical staining for HMFG-1 is positive. Apocrine cystadenoma can be distinguished also by expression of SMA (3,4). In our case, staining for both these antigens was negative. All other differential diagnoses could easily be ruled out by histological examination.

As eccrine hidrocystomas often occur on the face, they can be cosmetically troubling. Many patients wish for treatment despite the benign character of the lesions. The effect of lesional drainage by needle puncture is of short duration with recurrence usually after 6 weeks. Oral or topical anticholinergic agents have been shown to be effective but may cause side effects such as nausea and blurred vision (5). Perilesional injections of botulinum toxin may be temporarily helpful but are not feasible in young children (6). Surgical excision is the treatment of choice for individual cysts but is impractical with multiple lesions. Treatment with flashlamp-pumped pulsed dye lasers has shown variable success, whereas destructive methods such as CO_2 laser vaporization or electrodesiccation carry a risk of scarring (7).

In conclusion, multiple eccrine hidrocystomas should be included in the list of differential diagnoses of cystic lesions on the nose of children. A biopsy is helpful in uncertain cases.

Acknowledgment

We thank Dr. Diana Anders for carrying out the sweat testing.

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