Idiopathic facial aseptic granuloma

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DESCRIPTION

A 12-month-old girl was referred to the dermatology department due to multiple erythematous-violaceous papular and nodular facial lesions, elastic, with smooth surface and variable dimensions, that progressed over 3 months. Both upper and lower eyelids had non-exudative erythematous plaques, some with a yellowish hue (figure 1).

The girl had no systemic symptoms or palpable regional lymphadenopathy.

Initially, she was presented with recurrent bilateral eyelid chalazia and was treated with erythromycin (40 mg/kg/day) after being observed by ophthalmology. One month later, she developed similar lesions on her cheeks.

There was no history of the previous infection, systemic disease, recent onset of new drugs, trauma or insect bite. Ultrasound study of the nodule on the right cheek showed a well-demarcated solid, hypoechoic dermal lesion, without calcium deposits. Repeated assessment of the skin, using the skin pressurisation method, revealed no Demodex parasites. Laboratory workup did not reveal any significant abnormalities.

Cultures for bacteria, fungi and mycobacteria were negative.

Histology of the right cheek nodule revealed a granulomatous infiltrate with granulation tissue and hair shafts, consistent with idiopathic facial aseptic granuloma (IFAG) (figure 2).

She was initially medicated with oral erythromycin and methylprednisolone 0.5 mg/kg/day was added for 10 weeks, with gradual reduction of its dosage. Treatment with topical 1% metronidazole was also added during this time, with good clinical response and complete healing of the nodule on the right cheek.

IFAG is a paediatric condition, which was first described in 1999 by a group of French dermatologists, and named ‘pyodermite froide du visage’, due to its similarity with an abscess with very few inflammatory signs. IFAG is a rare, chronic and benign condition that occurs in children, between 8 months and 13 years, and usually heals without scarring. It is characterised by painless inflammatory facial nodules, usually presenting as a single lesion, localised in a particular triangle-shaped cheek area, delimited by the external limit of the orbit, labial angle and ear lobe.

The pathogenesis is still unclear; some authors have postulated that these lesions may be related to a granulomatous process appearing around an embryological residue or as a manifestation to include in the spectrum of granulomatous rosacea in childhood. There are no predisposing factors, no family history and no evidence for a microbial cause; parents sometimes recall a previous trauma or insect bite at the site of the nodule.

Diagnosis is clinical but skin biopsy may be very useful; IFAG and granulomatous rosacea have very similar histopathological findings with perifolliculitis, granulomas, folliculitis and lymphocytes surrounding epithelioid histiocytes. Ultrasound studies revealed hypoechoic oval structures in the dermis with posterior hyperchogenicity, without calcification.

Differential diagnosis include benign tumours, such as pilomatrixomas, epidermoid or dermoid cysts, chalazia; bacterial, mycobacterial, fungal or parasitic infections; dermato­myscopic signs; traumatic granuloma; dermatozytic papillomatosis; pilomatrixoma; pilo­folliculitis and lymphoid hyperplasia; neoplasias.

The lesion is surrounding variably-sized cavitory spaces and is accompanied by a richly vascularised granulation tissue (H&E × 100).

Figure 1 Multiple erythematous-violaceous papular and nodular facial lesions, elastic, with smooth surface and variable dimensions; both upper and lower eyelids had non-exudative erythematous plaques, some with a yellowish hue.

Figure 2 Skin biopsy of the right cheek nodule revealed a dense granulomatous inflammatory infiltrate, composed of histiocytes, multinucleated giant cells, small collections of neutrophils and plasma cells. The infiltrate is surrounding variably-sized cavitory spaces and is accompanied by a richly vascularised granulation tissue (H&E × 100).
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parasitic infections; pyogenic granulomas; Spitz nevi; xanthogranulomas, and vascular malformations or haemangiomas.1–3

IFAG has a prolonged course and may heal spontaneously.2 Usually oral or topical antibiotics are ineffective, although, some cases have shown a good response to oral macrolides or topical metronidazole.2

Learning points

► Idiopathic facial aseptic granuloma (IFAG) is a challenging diagnosis, mainly due to its rarity, and it is probably underdiagnosed.
► The diagnosis is clinical but skin biopsy may be very useful; we must include IFAG in the differential diagnosis of facial nodules acquired in childhood.
► This case aims to emphasise the association of IFAG with granulomatous rosacea in childhood, but mostly to raise awareness of this condition and avoid unnecessary surgical interventions or other treatments.

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Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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