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Case report

Orofacial granulomatosis in children—A challenge for diagnosis and treatment

Kaija-Leena Kolho [a,*], Kaarina Heiskanen [a], Matti Verkasalo [a,b], Anne Pitkäranta [b]

[a] Hospital for Children and Adolescents, Helsinki University Central Hospital and University of Helsinki, Faculty of Medicine, Helsinki, Finland
[b] Department of Otorhinolaryngology, Helsinki University Central Hospital and University of Helsinki, Faculty of Medicine, Helsinki, Finland

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ABSTRACT

The data on orofacial granulomatosis, OFG, in children are sparse. We describe here 8 pediatric patients presenting with OFG, 2 of these cases associating with Crohn's disease. Therapeutic agents included systemic immunosuppressants such as glucocorticoids, methotrexate, anti-TNF-alpha agent, dapsone, antibiotics (metronidazole), and local treatment with topical tacrolimus or intralesional injections of triamcinolone hexacetonide. The treatment response ranged from good to poor results. The number of young patients suffering from OFG is not currently known and there are no gold standards for treatment. Thus, prospective follow-up studies on these patients are needed to gain more experience of the therapeutic responses.

1. Introduction

Orofacial granulomatosis, OFG, is a clinical entity encompassing chronic inflammation in the oral cavity or surrounding areas of the mouth with no specific etiopathology. The disease presentation may include deep linear ulcers, tags or cobblestone formation in the mucosa or lip swelling that may associate with angular cheilitis or chronic irritation in the eyes [1,2]. Melkersson–Rosenthals syndrome may mimic OFG especially if the classical triad of lingua plicata, facial paresis and swelling in the orofacial region are not simultaneously presented [3–5]. As the histological assessment may reveal granulomatous inflammation – although not necessary to settle the diagnosis of OFG – causative agents have been searched among mycobacteria or other infectious agents but with negative results [1]. The inflammation may resemble that seen in sarcoidosis or in Crohn's disease, the most important disease association [6,7].

It has been estimated that 40–50% of the young patients with OFG will eventually develop Crohn's disease, among adults the risk may be lower [8]. Of notice, Crohn's disease may manifest several years after the first symptoms of OFG. So far, there are no means to screen patients at risk for development of chronic inflammatory bowel disease. However, the presence of intestinal inflammation may be easily detected by measuring fecal calprotectin, a surrogate marker for mucosal inflammation in, e.g. Crohn's disease [9,10]. As the incidence of inflammatory bowel disease is increasing in many western countries [11–14], it is possible that the number of young patients with oral manifestations of the disease is increasing as well. However, there is no epidemiological data on the incidence of OFG, with most reports including few cases. As it is obvious that wider knowledge of the disease will help to recognize the patients, we report here our recent experience on OFG in children.

We reviewed all pediatric cases presenting with OFG during 2006–2010 at Helsinki University Central Hospital. Our hospital is a tertiary case hospital with a source population of 29% of the total child population in Finland. All except one patient were treated with systemic immunosuppressants or antibiotics, and four patients with intralesional triamcinolone hexactetone injections. The patients were followed-up for 35 months (median, range 7–53).

2. Results

All patients had symptoms related for OFG for months before the diagnosis was made. During the median follow-up of 34 months, all patients experienced recurring symptoms. The

* Corresponding author at: Hospital for Children and Adolescents, University of Helsinki, Box 281, FIN-00029 HUS, Finland. Tel.: +358 40 7615172.
E-mail addresses: kaija-leena.kolho@helsinki.fi, kaija-leena.kolho@hus.fi (K.-L. Kolho).

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Table 1
Clinical characteristics of the eight children presenting with orofacial granulomatosis.

Patient no.	Gender	Age at diagnosis (years)	Duration of symptoms prior to diagnosis	Main symptoms	Follow-up from diagnosis (months)	Associated conditions	Treatment	Treatment response
1	Male	10	2 years	Swelling of the lower lip	11	Atopy Allergic rhinitis	Dapsone Prednisolone Triamcinolonehexacetonide injections Metronidazole	No response, adverse effects Good Moderate Ongoing
2	Male	15	Three weeks	Swelling of the lower lip	14	Asthma bronchiale Atopy	Antibiotics Prednisolone Triamcinolone hexacetonide injections Methotrexate	No response No response No response Ongoing
3	Male	13	4 months	Swelling of the lower lip	36	Asthma bronchiale Atopy	Tacrolimus ointment Triamcinolone hexacetonide injections Prednisolone Metronidazole	Good for a year Good for 5 months Good Excellent
4	Male	15	1 year	Ulcer in the mouth	36	Atopy	Local chlorhexidine	Temporary
5	Female	13	4 months	Swelling of the lower lip	34	Asthma bronchiale Epilepsy Crohn's disease diagnosed 3 months later	Prednisolone Cetirizine Azathioprine TNF-alpha-antagonist Triamcinolone hexacetonide injections	Temporary No response No response No response Ongoing
6	Male	12	4 months	Swelling of the cheek, gingival inflammation, ulcers in the mouth	46	None	Antibiotics Tacrolimus ointment Chlorhexidine rinsing Prednisolone Triamcinolone hexacetonide rinsing	No response Temporary Temporary Temporary Temporary
7	Female	11	4 years	Swelling of the lower lip, ulcers in the angular region of the mouth, aphtous ulcers	53	Crohn's disease diagnosed 7 months later	Prednisolone Tacrolimus ointment	Temporary Temporary
8	Male	12	1 month	Swelling of the upper lip, gingival inflammation	7	Asthma bronchiale Atopy	Metronidazole	Ongoing