Deep suppurative glandular cheilitis: A pediatric case report

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INTRODUCTION

Glandular cheilitis (GC) is an extremely rare condition affecting minor salivary glands of the lips, mostly described in adult men, whose pathogenesis and ideal management remain elusive. Three types of GC are recognized as GC simplex, superficial suppurative, and deep suppurative, with only few case reports in literature of the latter form.[1,2]

Here, we report a case of deep suppurative GC occurred in a female pediatric patient, refractory to conservative measures and successfully treated with vermilionectomy without recurrences at 3-year follow-up.

Abbreviations used:

GC: glandular cheilitis
GH: growth hormone

CASE REPORT

A 13-year-old female presented to our evaluation for swelling of the lower lip associated with recurrent painful fissurations and erosions, started one month before (Fig 1, *A*).

She was affected by growth hormone (GH) deficiency, for which she was undergoing substitutive therapy with synthetic GH and she had atopic asthma with sensitization to grasses.

On physical examination, the patient showed multiple erosions, ulcerations, and deep pustules with edema and macrocheilia causing eversion of the lower lip. The upper lip also exhibited a slight edema and desquamative areas (Fig 1, *B*). She reported pain and difficulties in lip movements affecting daily activities.

Mucosal swabs for infective agents as well as blood samples for autoimmune profile and allergic evaluation with prick and patch tests were negative. Moreover, a gastrointestinal evaluation excluded celiac or inflammatory bowel diseases. In addition, neuropsychiatric evaluation excluded self-inflicted nature of the lesions. Synthetic GH suspension did not lead to any improvements.

Treatment with topical emollients, antiseptic, and steroidal medications was found to be ineffective.

After 3 months of local therapy, the lesions were still present (Fig 1, *C*). A mucosal biopsy for histologic examination and direct immunofluorescence was performed and revealed non-specific findings, only excluding a granulomatous disease. Over the next few months, the patient worsened and presented with painful swelling, multiple profound ulcerations, and purulent discharge of the lower lip (Fig 1, *D*). A subsequent biopsy showed histopathologic findings of lymphoplasmacellular glandular infiltrate with ductal ectasia and inflammation, slight mucin deposition, and recognition of an ectopic minor salivary gland, leading to the diagnosis of deep suppurative GC (Fig 2). Magnetic resonance imaging confirmed the presence of dilated minor salivary glands.

Systemic antibiotic (first clarithromycin 250 mg twice daily for 6 days every month for 3 months and subsequently, doxycycline 100 mg once daily for 15 days every month for 3 months) and topical application of gentamicin and betamethasone cream and hyaluronic acid were prescribed without

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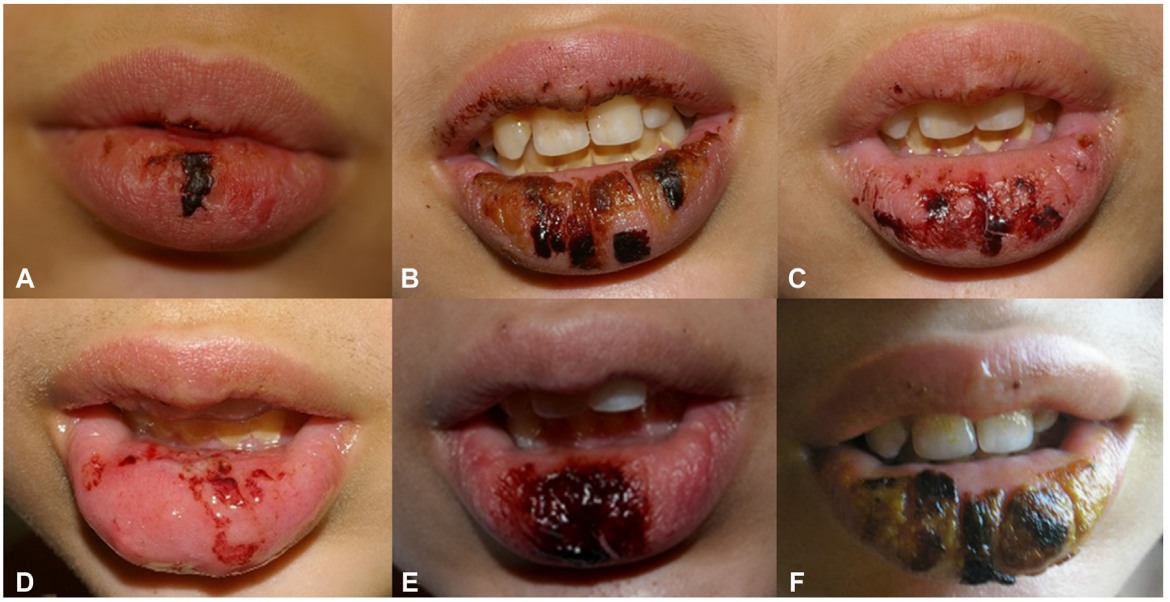

Fig 1. **A,** Initial presentation of the disease with fissurations and erosions associated to swelling of the lower lip. **B,** Clinical presentation after 2 months of disease: multiple erosions, ulcerations, and pustules with edema and macrocheilia causing eversion of the lower lip. The upper lip also exhibited a slight edema and desquamative areas. **C,** Clinical presentation after 3 months of topical therapy. **D,** Clinical presentation after 6 months from initial evaluation. **E,** Clinical presentation after 16 months from initial evaluation. **F,** Clinical presentation before the procedure of vermilionectomy: important swelling with multiple profound ulcerations of the lower lip associated with purulent discharge and pain.

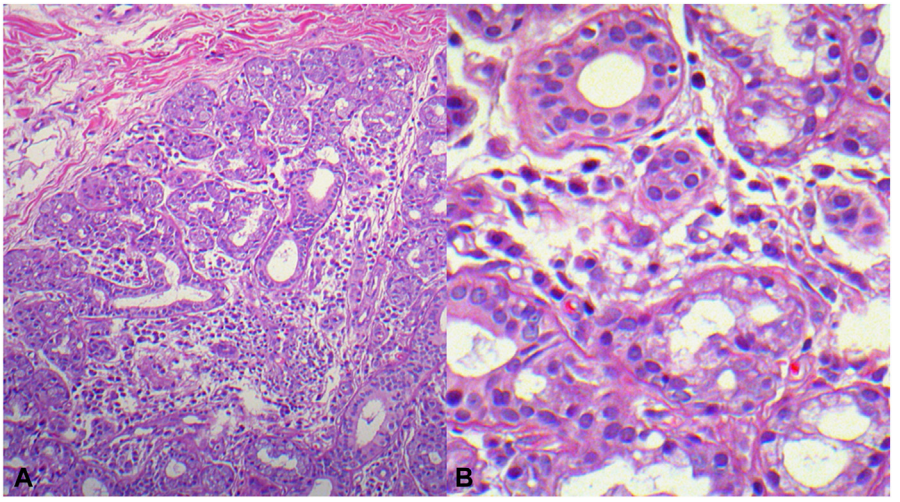

Fig 2. **A** Histologic sample showing lymphoplasmacellular glandular infiltrate with sialectasia and inflammation and slight mucin deposition. (**A** and **B,** Hematoxylin-eosin stain; original magnifications: **A,** 10×; **B,** 40×.)

improvement. Intralesional steroid injections with triamcinolone acetonide (40 mg once a month for 2 months) were attempted, leading to scarce results (Fig 1, *E* and *F*).

In accordance with the patient's parents, a vermilionectomy of the lower lip was performed, which led to complete remission of the lesions and pain (Fig 3). After 3 years of follow-up, our patient remained disease-free with no further therapy.

DISCUSSION

GC is a rare inflammatory disorder affecting accessory salivary glands of the lips, first described by Volkman[3] in 1870. GC is characterized by chronic

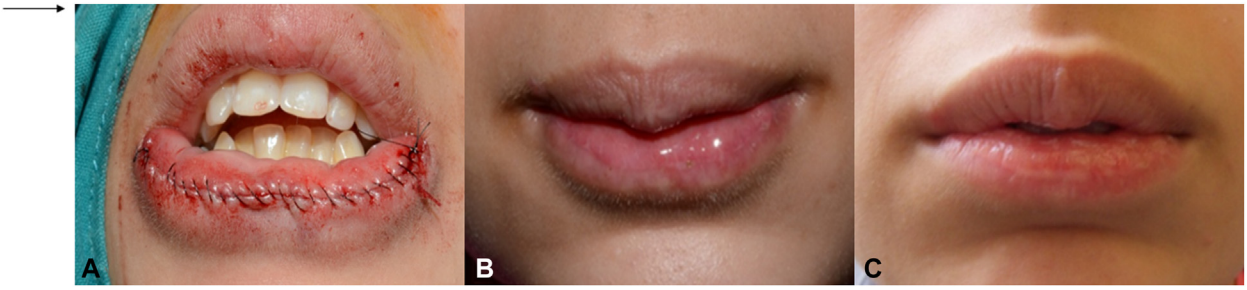

Fig 3. **A,** Clinical photograph during procedure of vermilionectomy of the lower lip. **B,** Clinical results showing complete remission after 3 months from vermilionectomy. **C,** Clinical results showing complete remission without recurrences after 1 year from vermilionectomy.

ductal hyperplasia and ectasia, frequently associated with actinic cheilitis.[4] Three subtypes of GC are recognized in literature: simplex, superficial suppurative, and deep suppurative.[1,2] The most frequent form is the superficial one, whereas the profound or deep form, recognized in our patient, seems to be the rarest.

GC tends to affect more often men (1.8:1 male-to-female ratio) in their fifth decade. Pediatric reports in female patients, such as the one presented, are extremely uncommon.[1,2] This condition is observed more often in white-skinned individuals and it usually involves the lower lip.[1,5]

The exact pathogenesis is still unclear and multiple risk factors have been proposed, such as emotional disturbance, chemical exposure, immune dysfunction, traumatism, viscous salivation, xerostomia, cosmetic filler injections, poor oral hygiene, immunosuppression, bacterial infections, actinic damage, and smoking, with the latter 2 being the most reported in literature as possible causative factors.[1,2,5] However, the rare pediatric cases tend to highlight the role of genetic predisposition, which determines saliva quality and aquaporin expression in affected salivary glands.[6]

Clinically, GC is characterized by swelling, erythema, crusts, and dilated ductal openings with symptoms of discomfort and pain: in particular, the simplex form tends to present with superficial erosions and crusts, whereas deeper variants can lead to eversion of the lip, profound ulcers, suppuration, and scarring.[1]

Diagnostic criteria have been proposed by Reiter et al,[4] including both clinical and histopathologic aspects, suggesting the importance of biopsy in the diagnosis. Histologically, GC shows ductal mucin deposition and ectasia, mucous /oncocytic metaplasia, sialectasia, and chronic inflammation.[2,4,6]

The most common differential diagnoses are actinic cheilitis, irritant and allergic contact cheilitis, atopic cheilitis, lichen planus, sarcoidosis, granulomatous cheilitis, actinic prurigo, multiple mucoceles, chronic sialadenitis of the minor salivary glands, as well as infective forms caused by human herpes virus infection, primary or secondary syphilis, and HIV.[1,2,7]

Moreover, the association between GC and the development of squamocellular carcinoma is not fully clarified. Although cases of squamocellular carcinoma of the vermilion border have been described in the context of deep suppurative form, GC is not considered a premalignant condition per se.[2]

Currently, there are different possibilities of management described in literature, without a definitive consensus and with short follow-up period. In milder forms, sun screen, topical and intralesional steroids, topical calcineurin inhibitors, and systemic antibiotic therapy together with avoidance of risk factors have been proposed for treatment. In the deep suppurative subtype, after the failure of conservative measures, surgical therapy with labial stripping or lower lip vermilionectomy and dissection of the minor salivary glands is often the therapy of choice.[1,2]

Our patient was extremely symptomatic and refractory to conservative treatment: vermilionectomy of the lower lip, even if performed at a young age, led to complete remission, which was maintained without any other recurrence or further therapy after 3 years of follow-up, suggesting good results of this surgical procedure in deep suppurative GC.

In conclusion, GC is a rare and difficult-to-diagnose entity, especially in its deep suppurative form, which can be present also in pediatric patients. Vermilionectomy could be a valid treatment option also in this population. However, more studies are necessary to understand its prevalence, pathogenesis, and the ideal management.

Conflicts of interest

None disclosed.

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