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An alternative technique using a gutta percha points and blue methylene to excision of congenital fistula of lower lip in patient with Van der Woude syndrome

Carlos Henrique Bettoni Cruz Castro, Matheus Furtado De Carvalho, Debora Cury Veloso, Marcio De Moraes

SUMMARY

Van der Woude syndrome is a rare disorder of craniofacial development, characterized by the triad (congenital lip fistula, cleft lip and/or cleft palate). Discomfort caused by spontaneous or induced drainage of saliva/mucus when pressure is applied or during a meal as well as poor aesthetics match is one of the main complaints of patients with congenital lip fistula. This paper aims to describe an alternative technique for excision of the congenital fistula of lower lip in patient with Van der Woude syndrome, using gutta percha points and methylene blue for better identify the the fullest extent of the fistula. This approach aims to facilitate a dissection of fistula tissues providing better preservation of the tissues around the fistula, faster healing and better aesthetics lip of the patient.

Key words: fistula, lip, surgery, oral.

INTRODUCTION

The presence of congenital fistula of lower lip was firstly observed by DeMarquay in 1845 (1). In 1954, Anne Van der Woude described an association between congenital lip fistula, cleft palate (CP) and cleft lip (CL), identifying a genetic and an hereditary component in some studied patients that characterized a new syndrome (2).

The Van der Woude syndrome (VWS) consists of a rare disorder of craniofacial development. There is an autosomal dominant inheritance with high penetrance and variable specificity (3). Even being a most common manifestation in cleft lip and cleft palate patients, the lip fistula is still misdiagnosed due to a great clinical variability (4).

Congenital lip fistula is a very rare malformation and represent the most common clinical finding in VWS having an interruption in the development of lateral groove of lower lip which is on the fourth week in the intra uterine life. Consequently, there is a gradual obliteration of it until finishes the growth, resulting in lip fistula (5).

It may be related to minor salivary glands in its internal portion or even invaginating to orbicular muscles of lip that communicates to ductal system of major salivary glands. Discomfort caused by spontaneous or induced drainage of saliva/mucus when pressure is applied or during a meal as well as poor aesthetics match is one of the main complaints of patients with congenital lip fistula (6). It is asymptomatic and there is not tendency to obstruction or infection that may be inflamed in a special situations such as individuals with suction of lip, lacerations and others (7).

Several techniques for excising congenital lip fistula were found in the literature (8). However, all techniques are considered invasive regarding tissue preservation around the fistula way. This paper aims to describe an alternative technique using a gutta percha point and blue methylene in order to identify better fistula pathway to be excised in a congenital lip fistula in patients with VWS.

CASE REPORT

A 19 years old woman, with CL and CP sought the Center of Oral and MaxilloFacial Surgery of

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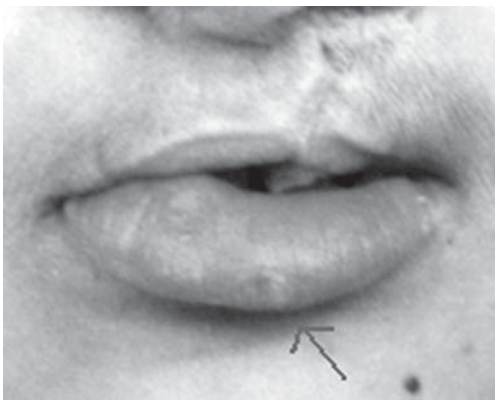

Fig. 1. Clinical aspect of lower lip. Observe an enlargement in the right part of lower lip as well as a depression which may indicated to be a congenital lip fistula on the left side

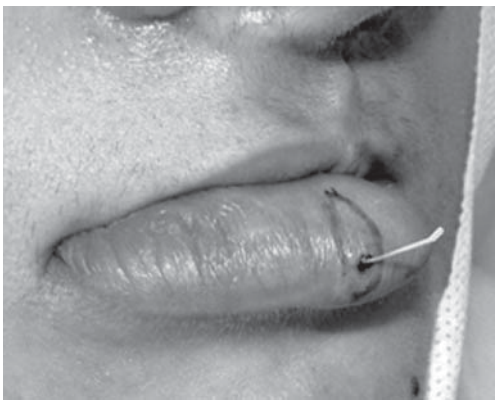

Fig. 2. An accessory gutta percha point FM (28 mm-Dentsply), soaked in blue methylene solution, is placed into the lip pit down to the end of fistula pathway

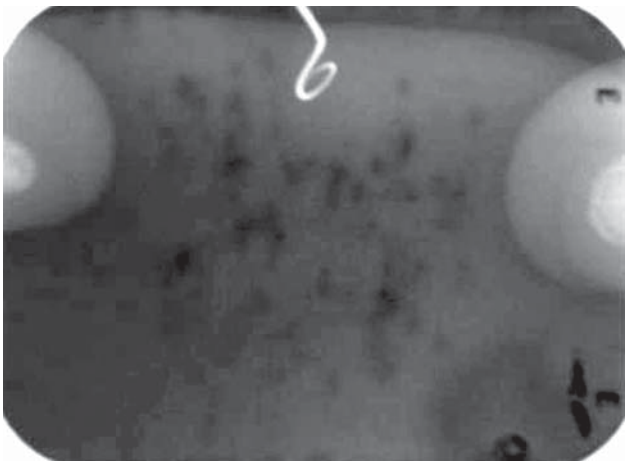

Fig. 3. Periapical x-ray was taken to visualize a gutta percha points in the fistula pathway

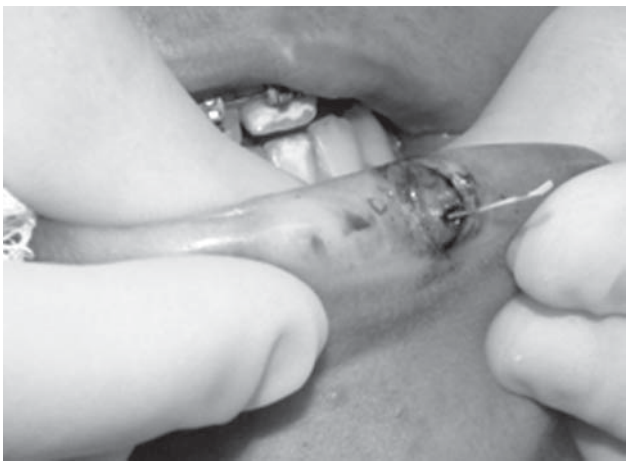

Fig. 4. Elliptical excision was done close to the fistula opening and parallel to fibers of lower lip

Pontificia University Catolica of Minas Gerais (FO-PUCMG), referred to surgical exposure of upper right canine from the Orthodontic Department of the same university.

On the extra oral examination, it was noted that there was an enlargement in the right part of lower lip of this patient as well as a depression which may indicated to be a congenital lip fistula on the left side (Fig. 1)

The patient reported that she has undergone to a surgical procedure to remove a lip fistula on her right side of lower lip seven years ago. In addition, she said about her dissatisfaction regards to aesthetics of her lower lip due to size discrepancy between both half of lower lip. Also, there was discomfort caused by draining of secretion from lip fistula on the left side.

The discrepancy between both sides was caused by fibrosis resulting from a surgical technique used to remove a fistula on the right side of the lip. Thus, it was recommended to the patient to undergo a surgical removal a fistula on the left side of the lip by using a technique with gutta percha points soaked in blue methylene in order to improve the detection of fistula's pathway and preservation of soft tissues.

Description of surgical technique

1. An accessory gutta percha point FM (28 mm – Dentsply), soaked in blue methylene solution, is placed into the lip pit down to the end of fistula pathway (Fig. 2);
2. X-ray was taken to visualize the fistula pathway (Fig. 3);
3. Elliptical excision was done close to the fistula opening and parallel to fibers of lower lip (Fig. 4);
4. Dissection of entire extension of fistula's pathway (Fig. 5);
5. Exeresis of the fistula as well as minor salivary glands around the fistula pathway (Fig. 6);
6. Suture (Fig. 7).

The surgical piece was sent to Laboratory of Histopathology and Anatomical Analysis of School of Dentistry – PUCMG. On microscopic examina-

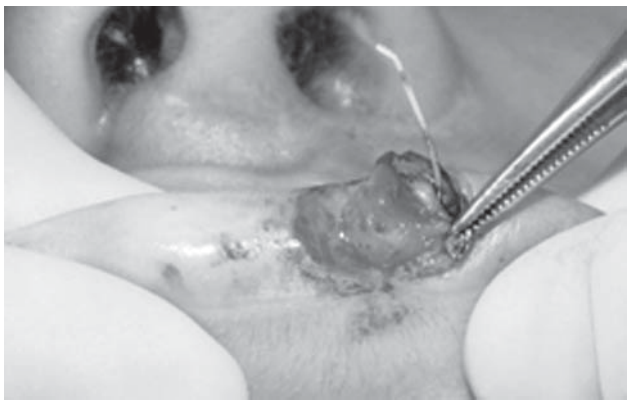

Fig. 5. Dissection of entire extension of fistula's pathway

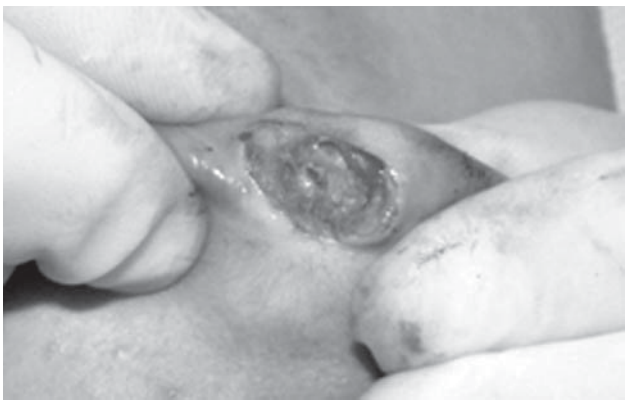

Fig. 6. Exeresis of the fistula as well as minor salivary glands around the fistula pathway

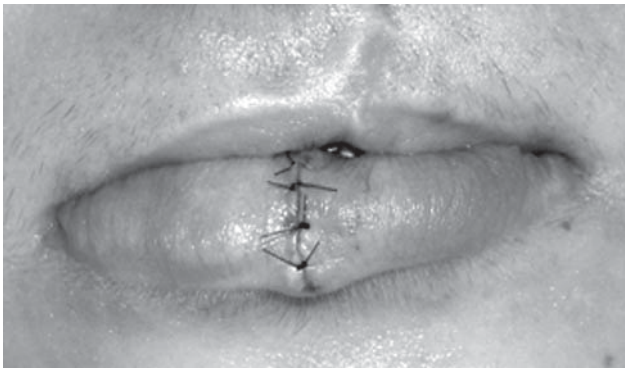

Fig. 7. Suture

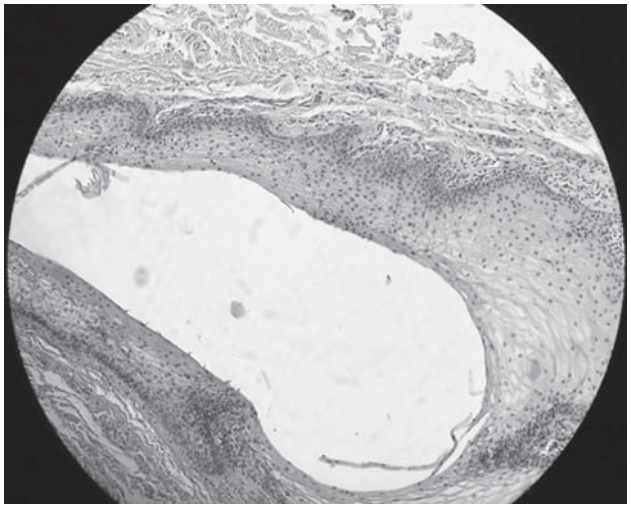

Fig. 8. Histopathological aspect

tion, it was observed that there is a stratified epithelium without keratin formation on whole extension of the walls of fistula ending in blind floor. Also, cells which resemble the cells of vermilion lip on the basal layer and cells of squamous layer were enlarged with small nuclei and less marked cytoplasm. Muscular fibers in submucous lining, acini of mucous glands and small ducts were seen adjacent to fistula walls. These findings confirmed the initial diagnosis of congenital lip fistula (Fig. 8).

At this time, the patient is on the fifteen days follow up period without cosmetic and functional complaints (Fig. 9)

DISCUSSION AND CONCLUSIONS

Lip fistulas are located bilaterally and symmetrically in each side of midline of lower lip that originates a small defect on the lip vermillion or in the limit of lip vermillion. Varying from a small fold to a relatively deep invagination measuring 0,5 to 2,5 cm of deepness ending in blind floor (7) .

The prevalence of patients with congenital fistula of lower lip is 3:100.000 and incidence of VWS in general population is 1,6:100.000 (9). In patients with CL and CP, there is an occurrence of congenital fistula in lower lip varies from 0,5% to 20% (7). There is not influence relating to sex (3).

Since the original article written by Anne Van der Woude, it has been suspected that there is an inherited character and a genetic alteration in this syndrome. Based on that, some genetic studies were done in order to determine a genetic mutation in VWS. The mutation which is responsible to this syndrome is present in the gene which encodes Interferon Regulatory Factor 6 (IRF6) which has been mapped to chromosome locus 1q32-q41 (9-14).

Diagnosis of congenital fistulas on the lower lip in patients with CL and CP or even in their family is very important to establish a prognosis and genetic advising. Then, the presence of congenital lip fistula increases the chance of having a child with CL or/and CP that is a severe deformity having a complex recovering and limited in the majority of cases (8). A carrier of mutation in the gene IRF6, there is a relative risk of transmission a CL or/and CP is about 26,4% and 23,55% for passing a lower lip fistula (15).

The diagnosis of VWS (isolated or familial) is conclusive when a patient has, at least, one of following clinical features: isolated lip fistula, lip

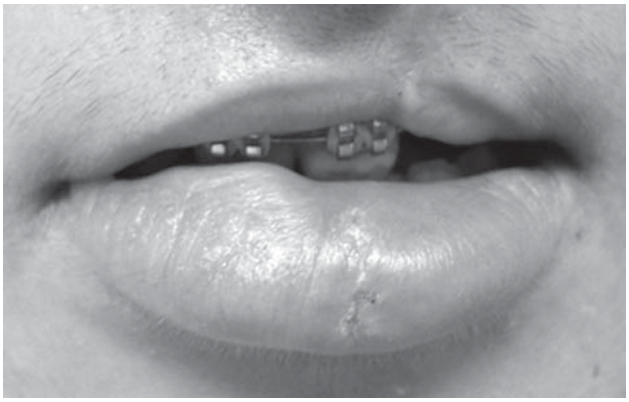

Fig. 9. Clinical aspect of lower lip. The patient is on the fifteen days follow up period without cosmetic and functional complaints

fistula and cleft lip (CL) and/or cleft palate (CP), isolated and familial lip fistula having first degree with CL and/or CP, CP and CL with a close family member with a lip fistula (16).

Patients with VWS may also have dental agenesis, defect in midline of cerebrum and hydrocephaly (9,12,17). As a main differential diagnosis is the poptliteal pterygium syndrome (PPS), allele entity of VWS (13).

Congenital lip fistula should be surgically removed when there is a recurrent infection, discomfort caused by draining secretion and/or aesthetics complaint (8, 18, 19).

Some surgical techniques have been shown in the literature. One of these is an elliptical incision close to fistula opening and parallel to a margin of lip vermilion followed by fistula removal (20); removal of fistula using two triangle shape incisions that base includes both fistulas (21); surgical excision in longitudinal direction of lip including part of adjacent salivary glands (22); removal of a unique block linking both fistula openings (23); marking a fistula pathway with blue methylene followed by its filling with a low fusion paraffin, making it easier to dissect the fistula pathway. When fistulas are small, it is harder to explore an accurate pathway (24).

On the other hand, all these techniques are considered invasive regarding preservation of tissues which surrounding a fistula pathway. Losing tissue in the lip midline may lead to irregular contour of lower lip as well as developing the mucus retention cyst or mucocele from these remnants. Complications may be caused by a surgical technique used to remove congenital fistula (7).

An alternative technique using gutta percha point and blue methylene to exeresis a congenital lip fistula in patients with VWS provides to excise the entire fistula pathway that maintenance tissues around the fistula resulting in better healing process and improvement in lip aesthetics to a patient.

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