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CASE REPORT

Sialolithiasis in the Duct of Submandibular Gland: A Case Report in Patient with Epidermolysis Bullosa

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ABSTRACT

Aim: To describe the options of treatment to remove a sialolith associated with the submandibular gland duct in a patient with epidermolysis bullosa (EB).

Background: Treatment of patients with EB is very complex and involves a multidisciplinary team. This condition is characterized by a spectrum of blistering and mechanical fragility of the skin. One main feature of this disease is the esophageal constriction and possible constriction to the submandibular duct. This alteration may induce the formation of calculi in this duct, which is called sialolith. Once the sialolith obliterates the trajectory of the duct this will lead to a sialolithiasis. The calculi have to be removed.

Case report: Seventeen years old female patient with dystrophic EB developed a sialolith at the submandibular duct. She has a limited mouth opening and her tongue was collapsed with mouth floor. The first choice of treatment was the lithotripsy, once this procedure is less invasive and a surgical remove could worse the collapsed tongue. She was with acute pain and with a great augmentation in the submandibular area. Once the patient was debilitated and has difficult to swallow she invariably needed to be hospitalized in order to receive intravenous medication. During the hospitalization the sialolith could be seen through the opening of the duct and the calculi was removed with local anesthesia.

Conclusion: The treatment of sialolithiasis usually does not present major challenges, nevertheless if the sialolithiasis is associated with EB, the treatment became an extremely challenge. In this particular case the option of treatment was the less invasive.

Clinical relevance: This case report has an enormous clinical relevance once there is no protocol to treat patients with EB and buccal diseases.

Keywords: Bullosa dystrophica, Epidermolysis, Sialolithiasis, Sialolith, Lithotripsy.

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INTRODUCTION

Epidermolysis bullosa (EB) represents a spectrum of conditions characterized by blistering and mechanical fragility of the skin. This group of disease has a genetic heterogeneity and even more marked variation in clinical phenotypes. A recent classification recognizes four major EB groupings and more than 30 EB subtypes. These groups are: Intraepidermal EB (simplex), junctional EB, dermolytic EB (dystrophic) and mixed EB (Kindler syndrome). The molecular changes are well known for 13 subtypes of EB.[1] Each type of EB will lead to a different morbidity involving the soft and hard tissues of the maxillofacial complex. During the normal daylight activities the patients could suffer mild to moderate trauma even from wear and tear that can cause large bullae and ulcers of the skin and mucous membrane. In some cases bullae develop spontaneously.[2]

Recessive dystrophic epidermolysis bullosa (RDEB) is characterized by lamina dura separation, due to blistering below the lamina densa of the basement membrane zone. This particular type is associated with the absence of type VII collagen fibers.[3] As with all forms of EB, specially in this one there are widespread bullae involving the skin and mucosa that heal with atrophic scarring. There also defects resulting in retardation of bone growth.[4] In RDEB, the teeth are not directly affected although severe anterior crowding is common. The tongue generally is collapsed with the mouth floor and the mouth opening is restricted due the commissures scars. The satisfactory dental treatment of patients with this presents a great challenge to the general dentist and others specialties involved in the case.

One main feature of the disease is the esophageal constriction,[5] making a relationship with maxillofacial

complex the submandibular gland duct equally could be constricted. The constriction in association with the path of the duct increases the possibilities to development of a sialolithiasis. Sialoliths are calcified structures that develop within the salivary ductal system. They are believed to arise from deposition of calcium salts around a nidus of debris within the duct lumen. This debris may include inspissated mucous, bacteria, ductal epithelial cells or foreign bodies. The cause of sialoliths is unclear, but their formation can be promoted by chronic sialadenitis and partial obstruction. Their development is not related to any systemic derangement in calcium and phosphorous metabolism. The most frequent location that sialoliths appears is at the submandibular duct.[6]

The submandibular gland has both mucous and serous cells that empty into ductules, which in turn empty into the submandibular duct. The duct exits anteriorly from the sublingual aspect of the gland, coursing deep to the lingual nerve and medial to the sublingual gland. It eventually forms submandibular duct between the hyoglossus and mylohyoid muscles on the genioglossus muscle. Submandibular duct, the main excretory duct of the submandibular gland, is approximately 4 to 5 cm long, running superior to the hypoglossal nerve while inferior to the lingual nerve. It empties lateral to the lingual frenulum through a papilla in the floor of the mouth behind the lower incisor tooth. The openings for the sublingual gland, or the sublingual caruncles, are located near the midline of the sublingual fold in the ventral tongue.[7]

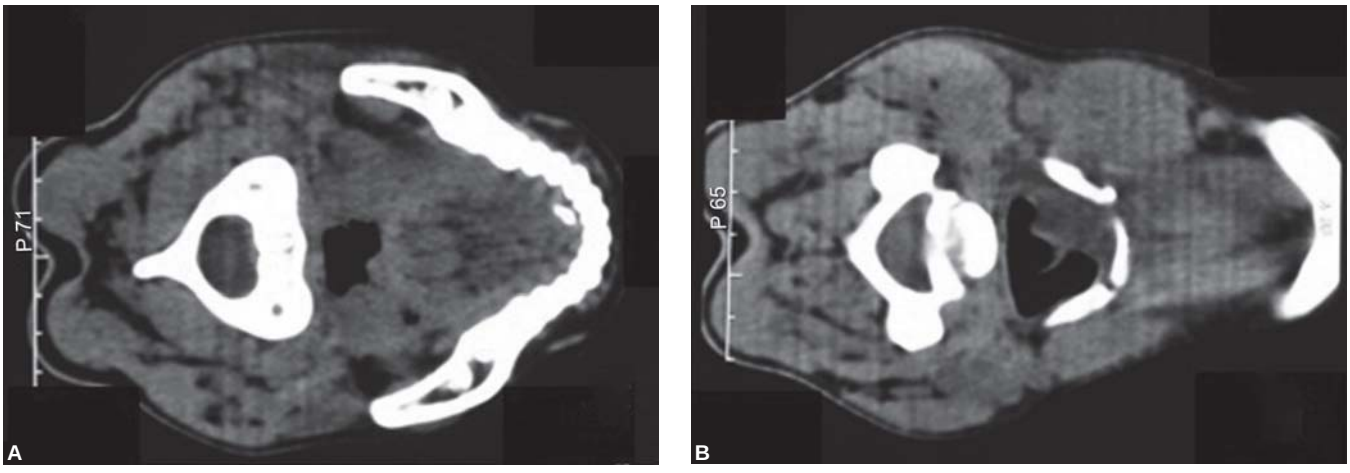
CASE REPORT

A female patient, 17 years old, presented to emergency of Oral and Maxillofacial Surgery at Hospital XV, Curitiba/PR, with augmentation in right submandibular region (Figs 1A to E). She reported an acute pain related to the area, with difficult to shallow and open the mouth. She has the diagnosis of RDEB and presented with many blisters, widespread bullae and atrophic scaring (Figs 1A to E). The limited mouth opening is the result of the obliteration of the buccal commissures related with blistering and posterior

Figs 1A to E: (A) Frontal initial aspect, (B) oblique initial aspect, (C) limited mouth opening, (D) inferior aspect and evident augmentation in right submandibular area,(E) scars in the left arm and the sindactilia

scars (Figs 2A and B). Since, her birth she have had been through three surgeries in her hands because of the sindactilia. These procedures have to be done under only sedation of the patient in another hospital that is expert in pediatric treatment, whether an intubation could lead into a obliteration of the airway path thus making a life-threatening condition.

Patient had another episode of augmentation in the same region, 1 year before. At that time an ultrasound examination of submandibular gland was done and a calculi was found at the submandibular duct (Fig. 3), this calculi was approximately 8.2 mm high by 33.7 mm inch wide. At that moment, the treatment of choice was an accompaniment.



Figs 2A and B: (A) Axial CT cone beam image showing the calculi, (B) axial CT cone beam slice showing the augmentation in the submandibular gland

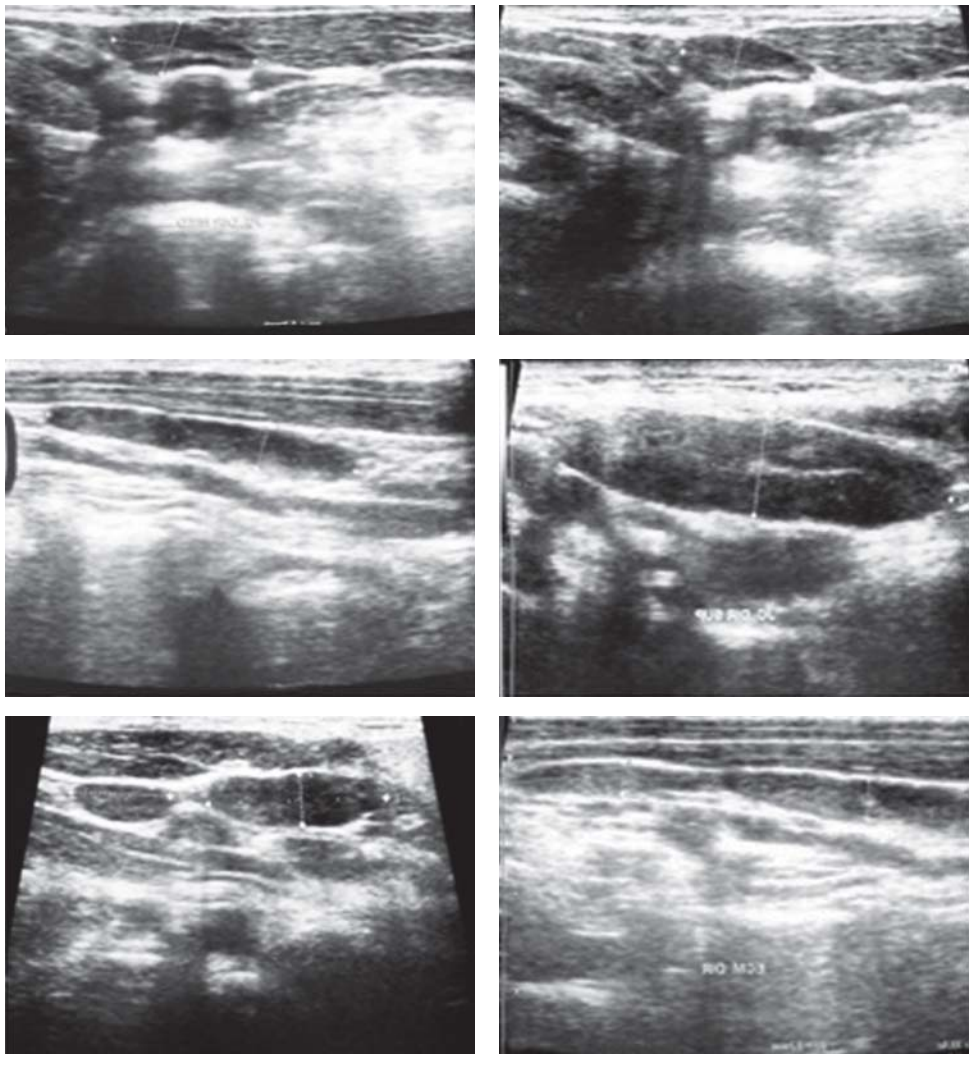


Fig. 3: Ultrasound examination of the calculi at the first episode

Because she was not complaining of pain and did not have any systemic commitment she was medicated with an anti-inflammatory.

Unfortunately at this second episode she was systemic compromised, she was with acute pain, had difficulty to swallow, a considerable augmentation in the submandibular region was present and to avoid a systemic widespread infection she has to be hospitalized. Multidisciplinary team (maxillofacial surgeon, the head and neck surgeon and the urologist) was involved in the treatment and decided to medicate the patient with: Saline solution, dextrose solution, ceftriaxone, clindamycin, ketoprofen, dipyrone. Computerized tomography (CT) of the region, laboratorial exams and ultrasound was requested.

At the axial slice of the CT a sialolith with about 8 mm per 7 mm associated with the submandibular duct became evident; it appears that it was at the last third of the duct (Fig. 4). At the laboratorial exams she appears with leukocytosis and a high mean corpuscular volume. Based in the risks associated with the epidermolysis, it was decided to try a lithotripsy to break the sialolith, since the service already tried before with success in another to cases. In this point of the treatment the patient was at the 5th day of hospitalization, she was stabilized; the augmentation became more restricted, she was not reporting pain and was accepting food.

When the patient was preparing to pass through the lithotripsy the sialoliths became superficial and the treatment plan had to be changed. Now the goal was to remove directly the sialoliths, just with local anesthesia. But a great challenge in this particular case was the fact that the patient has a little opening of the mouth and her tongue is collapsed with the floor of the oral cavity. The sialolith was completely

Fig. 4: After 4 months, the patient do not have any esthetic or functional alterations

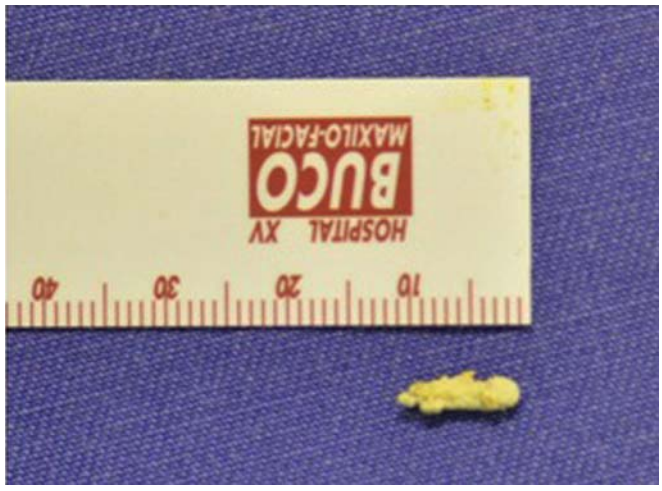


Fig. 5: The sialolith that was removed

removed (Fig. 5) and drainage of the saliva could be obtained.

After just 1 more day in hospital the patient continued the treatment at home. Seven days later she came back to a consultation without symptoms, just with a residual augmentation of the submandibular gland that was expected to recede. At the final CT the calculi were not anymore there and a regression of the augmentation of the submandibular gland was visible.

DISCUSSION

Individuals with severe generalized recessive dystrophic EB have extreme fragility of oral mucosa. This is usually evident after birth and it can interfere with a neonate's ability to suckle. Oral ulcerations can affect all areas of the mucosa, including the tongue. The lesions heal with scarring. The continual process of blister formation and healing with scarring results in marked changes in the oral architecture. The tongue looses the lingual papillae and becomes bound down to the floor of the mouth, known as ankyloglossia.[8-10] Moreover the progressive scars in the labial commissure could induce an obliteration, leading to a significant decrease in mouth opening.[11]

The alterations in the mouth floor which can lead into an obliteration of the submandibular duct, this situation create an ideal environment to calculi development. In this particular case oral manifestations of the EB end up hindering the treatment of the calculi. In order to avoid great damages that could worse the mouth obliteration and the ankyloglossia the lithotripsy turn into the first option of treatment.

In the past years, research has been dedicated to develop less invasive alternatives to conventional surgery to remove the sialolith. In gynecology, orthopedics and general surgery, the endoscopic procedures have proved been

successful. The removal of calculi from the urinary tract across the lithotripsy has revolutionized the urology. Nowadays the surgical intervention is required in less than 1% of cases.[12]

Since, the early 1980s the extracorporeal lithotripters have been used to ablate renal and biliary stones by the propagation of shock waves through the body using an acoustic generator. The first equipment could not focus the shock waves over a sufficiently small area to allow lithotripsy of salivary stones, without placing the teeth, eyes and brain at risk.[13] However, recent technical advances in the designing of electromagnetic and piezoelectric generators have allowed the shock waves to be focused over a zone 17 × 4 × 5 mm which has permitted their successful application to salivary gland lithotripsy *in vivo*.[14]

According to Yu CQ, 2007, there are three options in which we can treat patients with salivary stones: Removal through the oral cavity, interventional sialoendoscopy, and resection of the gland. Our choice depends on the size, shape, number and quality of the stones. The author suggests that the choice of type of treatment depends on the size of the stone, if the stone is under 4 mm of size the sialoendoscopy using the lithotripsy should be done, in other hand if the size of the stone overtake the 4 mm the operation is the first option.

It was first decided to make a lithotripsy because the maxillofacial service of the hospital has success with this technique to remove sialoliths in another two cases in the past. Iro H et al 1992, have described the success in removing a sialolith in the submandibular gland duct through the lithotripsy. This procedure is less invasive, once there is no incision and the procedure did not requires sedation. If there are any teeth sensibility during the lithotripsy a local anesthesia is required. The area of interest is placed into the sphere that propagates the shock waves and a concomitant tomography images guide the operator into the area of interest.

Another option could be to remove the calculi through an intraoral incision with local anesthesia. But in this type of patient the healing is not predictable and the ankyloglossia could be aggravated, once the healing process invariably lead into a severe scars. This technique is done under local anesthesia; a linear incision is done in the most prominent region followed by dissection and removal of the calcified mass.[15]

The last option and the most invasive one would be the excision of the sialolith through extraoral accesses. With this approach a numerous complications related with the EB could appear such as: Difficulty to healing the tissues, healing with a significant scar, difficulty in the recovery of the surgery. One important detail is that a general anesthesia will be required, and the intubation probably would be extremely difficult and we could deal with the real possibility of the obliteration in the upper air path.

CONCLUSION

EB is a very complex disease and the treatment could not be distinct. In respect to oral manifestations, the obliteration of the mouth and the ankyloglossia are the foremost challenges for the oral management of the patient. The oral morbidity associated with the disease is a result of the secondary effects of the disease, such as dental caries and periodontal disease. The treatment of sialolithiasis usually does not present major challenges, nevertheless if the sialolithiasis is associated with EB, the treatment became an extremely challenge. In this particular case the option of treatment was the less invasive. A specific treatment is required for each patient that has this condition; a protocol is difficult to be established once this condition have so different manifestations between the patients.

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