

# *Childhood Illnesses of the Salivary Gland*

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## **Clinical Case**

*Natália is 3 years old and has received all the routine vaccines. She presents periodic swelling on the left side of the parotid region, which, according to the mother, is 'similar to mumps'.*

## **Introduction**

A swelling of the salivary glands is unusual during childhood and represents a diagnostic and therapeutic challenge for the pediatrician and the otorhinolaryngologist.

The salivary glands are groups of acinar units, made of serous and mucinous components, arranged in lobular formations (major) connected by connective tissue. They produce saliva and play an important role in the lubrication and protection of the teeth and buccal mucosa; they are part of the process of deglutition and digestion.

The salivary glands are divided into major (3 pairs) and minor (the number varies). The parotids are the largest glands and have an anterior location, close to the mastoid apophysis and to the external auditory canal. The submandibular glands are smaller, but are responsible for producing 2/3 of the salivary. They are located below the horizontal ramus of the mandible, and are medially in close contact with the floor of the mouth. There are also sublingual glands and several smaller salivary glands in the oral cavity. Saliva production is a result of nervous stimuli of the parasympathetic and sympathetic systems.

## **Investigation**

A careful history should always be the first step for patients with history of any salivary gland swelling. Some questions should be answered in order to achieve the diagnostic reasoning. Has anyone else noticed? If the doctor is not sure whether the condition is real, since the swelling is not always present when examining a patient, it is important to verify if someone else besides the mother and the father has described the swelling, or if pictures have been taken of the child when presenting the condition. Has the child received all the immunizations? Even though immunizations do not exclude some infectious processes, including mumps, it tends to guide the diagnostic reasoning to other etiologies, especially in recurrent processes, such as the example presented above. Do any other complaints precede/succeed the swelling? Prodromes of viral infections support

this etiologic reasoning, just as a systemic impairment (fever, worsening of the clinical picture) may indicate bacterial infections, and joint complaints indicate autoimmune processes.

The presence of symptoms related to the facial nerve should always point in the direction of a process with high risk of malignancy. Does the patient feel any local pain or is the region red? Local inflammatory characteristics usually occur with most of the acute and recurrent processes, and they are quite nonspecific, with the exception of very intense manifestations that follow brief distensions (obstruction) and bacterial infections. On the other hand, growths presenting less intense non-intermittent flogistic signs should obviously direct the diagnostic towards tumors (benign and malignant). Does the swelling decrease spontaneously and completely? It contributes to the differential diagnosis between specific and nonspecific inflammatory pictures and expansive processes (benign or malignant). What are the medications being used (antihistamines, psychotropics)? These medications can provoke changes in the salivary glands due to a reduction in the saliva production. Are there any other health problems (metabolic, cirrhosis, B12 deficiency, Sjogren's syndrome and granulomatosis)? These diseases, among others, can deteriorate the gland tissue and cause chronic sialadenitis.

#### **Differential diagnosis**

- Viral parotiditis (mumps)
- Recurrent parotiditis of childhood
- Sialolithiasis
- Rheumatic diseases (Sjogren's syndrome) – uncommon in children
- Atypical infections (tuberculosis, actinomycosis, syphilis, gonorrhea, cat scratch disease)
- HIV/AIDS
- Cystic fibrosis
- Benign tumors
- Malignant tumors

#### **Mumps**

Even though illnesses of the salivary glands are unusual in children and most of the cases are infections caused by the mumps virus, this is not the etiology of every process. In such a case, why not consider viral parotiditis for clinical presentations such as Natália's?

There has been a dramatic decrease in the incidence of the disease since the implementation of the mumps vaccine in 1967. Differently from the clinical case presented, mumps cases have typical systemic viral manifestations; 75% present a bilateral manifestation and one episode yields permanent immunity. Therefore there are no cases of recurrent sialadenitis. Despite being uncommon, other viruses can present tropism for the salivary gland tissue (cytomegalovirus and coxsackie). As a consequence, when viral manifestations coincide with a swelling of the salivary glands, it is necessary to determine the type of virus involved, if possible, using serologic tests (IgM and IgG).

#### **Physiopathogenesis of chronic sialadenitis**

Although there are several possible etiologic factors, the main elements are the

reduction in saliva production and/or obstruction with salivary stasis. With the diminished salivary flow, an ascending infection of the oral cavity occurs. The infections lead to multifocal ductal irregularities that progress to partial stenoses. These partial stenoses lead to salivary retention and ductal dilation, which will then lead to more infections, yielding the formation of mucus-pus, creating a salivary retention cycle (infection), with variable recurrence intervals.

- **Acute sialodenitis**
  - Dehydration/obstruction
  - Hyposalivation
  - Stasis
  - Oral bacteria
  - Toxic elements from the ducts to the parenchyma
  - Multiple abscesses
  - Other theories
- **Chronic sialodenitis**
  - Duct ectasis
  - Stenoses
  - Reduction of secretory tissue
  - Stasis
  - Oral bacteria
  - Toxic elements from the ducts to the parenchyma
  - Multiple abscesses
  - Other theories
- **Supplementary tests**
  - Plain X-ray

It is not very useful. It can show stones in the parotid duct, which are radiopaque in approximately 40% of the X-rays, while in the submandibular gland where stones are more common, only 20% are radiopaque.

- Echography

It is relatively sensitive to some of the cystic/solid processes, and it is widely used as it is not an invasive procedure and it is readily available. In the specific case of chronic sialodenitis, this exam may present variable diagnostic abilities for duct sialectasis, and it is considered similar to sialography by some authors. It should be stressed that negative results do not exclude the condition (false-negatives).

- Sialography

It is an invasive exam, due to the manipulation of the ductal system and the need of contrast agents. The identification of ‘sausage’ deformities or contrast leak into the parenchyma is a sign of degenerative chronic process of the ductal system. It can be used to classify the degrees of deformity in the ductal system.

- CT scan

As the concurrent use of contrast, the parenchyma increase as a consequence of fibrosis and inflammatory infiltrate can identify ductal ectasia more precisely than sialography. It is more useful in cases of differential diagnosis with tumoral lesions.

- MRI

Many authors consider it as a possible diagnostic gold standard, because it is able to assess both the glandular parenchyma and the ductal system without contrast, since saliva can be used instead (Sialo-MRI). Besides cost, its limitations include the need for skilled professionals, capable of interpreting the results, and the length of the exam. Its duration makes it hard for children to undertake it, unless sedated.

- Sialendoscopy

This is a new method that enables the visualization of the ductal system, as well as the removal of organic and inorganic debris (stones) and dilation of a portion of the ductal system. It is also considered invasive since it requires the dilation of the ductal system, and the introduction of an endoscope and a device to remove debris. The procedure requires sedation of pediatric patients and has a high cost. In spite of these potential limitations, sialendoscopy represents a new hope in the management of pathologies associated with the salivary glands. In addition to making the diagnosis possible, it allows the treatment by removing different types of debris, which are contributing to the physiopathogeny of the process. When available, sialendoscopy becomes a mandatory stage in the cases in which the children cannot have a normal life, due to frequent episodes of recurrence; the gland removal is an alternative.

### **Recurrent parotiditis**

With the exception of acute mumps, recurrent parotiditis of childhood is the most common diagnosis in chronic presentations for patients in this age range. It is more common in 3 to 6-year old boys. It presents as a swelling of the gland with moderate hyperemia and pain, which lasts from 4-7 days to months, alternating with periods without symptoms for days to months. Its etiology is unknown, and there are theories related to genetic, allergic and autoimmune processes (Sjogren's syndrome) and also immunodeficiency. Therefore, a recurrent status of parotiditis prior to the actual diagnosis of recurrent parotiditis of childhood should be investigated as to the possible underlying pathological process, such as Aids and autoimmune processes.

The unilateral impairment is more frequent, and it can occasionally lead to an intraglandular abscess. Clear saliva associated with white flakes is excreted through the main duct. The existence of pus characterizes a suppurative parotiditis, but does not exclude a diagnosis of recurrent parotiditis. The bacteria involved can usually be found in the oral cavity, but they are not very aggressive (staphylococcus and streptococcus). Even though it is not considered a sialolithiasis, organic debris accumulated in the ductal system lead to stasis, in the same way as an inorganic sialolith. However, differently from the sialolithiasis that often occurs in the submandibular gland, being one of the major causes of submandibulectomy, the process under discussion affects more frequently the parotid glands.

According to the literature, the diagnostic ability of echography may vary between 18 and 86%. It is routinely indicated because it is not very invasive. On the other hand, when sausage-type images (resulting from deformities of the ductal system – pinpoint ductal sialectasis) are identified by sialography, it is considered as

diagnostic for the chronic inflammatory process of the gland. However, it is an invasive exam, and, in addition to manipulation, there is the risk of an allergic reaction to the contrast, and an experienced radiology team is required to assist the patient. In the CT scan, density is increased by the cellular infiltrate and fibrosis, and the exam is not very useful to assess this specific diagnosis. It is more useful in the differential diagnosis when one suspects of tumors.

The introduction of contrast before the tomography generates a sialo-CT, and it is possible to assess more accurately any ductal alterations, which are characteristic of the chronic obstructive process. Considered by many as the gold standard, sialo-MRI allows the identification of the typical ductal alterations without contrast, because saliva can be used instead. The most reliable findings on the ductal system conditions are obtained through the “vily T2-weighted 3D FSE” exams, while alterations in the glandular parenchyma are obtained through “2D T2-weighted images”. The disadvantages of MRI would be the amount of time necessary to undergo the exam, the difficulty some clinics have shown in obtaining clear images of the ductal system, the cost and the claustrophobic patients. Histologically, biopsies of chronic processes are similar to those of Sjogren’s syndrome. The clinical picture reverts quite often during adolescence, but if not, it can advance to necrosis of the acinar cells and progressive substitution of the healthy tissue by lymphoid cells and fiber adipose tissue.

Up to a few years ago, as a result of frequent presentations that excluded other specific diagnosis and the fact that noninvasive actions did not mean clinical relief for the patient, doctors recommended removal of the affected salivary gland. In the last few years, however, the technological advance achieved with the miniaturization of endoscopic equipment provided the endoscopic treatment of the salivary glands. In addition to contributing to the diagnosis, by allowing several pathologies to be seen (stones, stenoses, etc), the procedure permits dilations of the stenoses, washing out the ductal contents and instillation of anti-inflammatory and anti-infection medication, as well as insertion of temporary stents in the salivary ductal system. In spite of the unmistakable evidence of its beneficial effects when compared to other invasive therapies, this procedure has been more used because of its non-invasiveness in relation to the complete removal of the gland.

- Rheumatic diseases (Sjogren’s syndrome)

Rare among children, this autoimmune condition should be included in the differential diagnoses because an early diagnosis is important and the salivary glands swelling is often part of a clinical status, even without xerophthalmia and xerostomy. The lymphocytic invasion of the glandular tissue is the histological finding, which, in association with other clinical aspects, leads to the diagnosis.

- HIV/AIDS

The impairment of the salivary glands (especially the parotid glands) in pediatric patients with HIV/AIDS is not that uncommon. Even though it usually happens in immunocompromised patients and is considered a sign of a worse prognosis, it can also be one of the first manifestations of the virus’ presence in the body. Its management is the same as that of cases of unknown etiology.

### Summary

If a recurrent parotiditis of childhood is suspected, parotid massage and analgesia should be provided as needed. Parents should be reassured until a diagnosis can be defined. In spite of the limitations, an echography is recommended because it is non-invasive and has the possibility of diagnosing solid and cystic lesions and ductal changes. If another exam that requires anesthesia is necessary and considering the diagnostic hypothesis and equipment availability, the recommendation of a SIALORESONANCE or a SIALENDOSCOPY should be discussed. In addition to helping the diagnosis by the ductal system visualization, it is important to bear in mind that endoscopy can play a role in the treatment through washing and instillation of antibiotics and cortico-steroids, with or without the insertion of a “silaostent” to maintain an appropriate drainage.

From the serologic point-of-view, immunodeficiencies (HIV) and autoimmune diseases (Sjogren’s syndrome), although rare, should not be ruled out. The older the patient is at the onset of the salivary gland inflammation, the higher is the possibility of an autoimmune condition as an etiologic factor. Surgery (total parotidectomy, removal of the submandibular gland) is the last resource for recurrent incapacitating clinical cases.

### Recommended readings

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