Sialosis: Recognizing a Growing Problem

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Abstract

Learning goals: At the conclusion of this presentation, the participants should be able to recognize the characteristic pathology, radiology and conditions associated with sialosis, and discuss current treatment options.

Objectives: Sialosis is chronic, non-inflammatory swelling of the salivary glands, predominantly affecting the parotid glands and associated with endocrine disorders, pharmacologic agents or nutritional deficiencies. We report case examples of sialosis including characteristic pathology and radiology as well as management options supported by a literature review.

Methods: Patient chart analysis and literature review.

Results: A 63 year old male with greater than twenty years of bilateral parotid gland swelling is presented. Workup of salivary swelling and ultimate diagnosis of sialosis will be discussed. Sialogram, MRI and gross pathology sections are reviewed. Decision-making regarding medical and surgical management in this case is discussed, including use of botulinum toxin injection, taympanic neurectomy and steroid infusions.

Conclusions: Sialosis is a likely underdiagnosed entity with classic histopathologic, radiographic and clinical findings. Recognition of this diagnosis and initiation of intraluminal steroid infusions can impact the clinical course of the disease.

Case Report

A 63 year old man presented with a 20+ year history of bilateral parotid gland swelling worse on the left. It began as painless swelling until 2002. At that time, the swelling became painful and treatment was first sought. He reported no history of dry mouth, dry eyes, or arthritis throughout this course. His past medical history included diabetes and hypertension, both of which were being treated. The pain was constant and dull, which was not exacerbated by eating and was not reproduced by massaging of the gland. A full work-up was undertaken at that time, including an autoimmune panel (SSA, SSB, RA, ANA) and a Shimier’s test, all of which were negative. In 2002, a minor salivary gland biopsy was taken from the right side of his lower lip. The biopsy was interpreted as “suggestive of Sjogren’s syndrome,” and was given a preliminary diagnosis of Sjogren’s syndrome at this time. His continued symptoms prompted an MRI scan in 2003 and again in 2004, which showed only diffuse enlargement of the glands, with no distinct masses, or areas of irregular enhancement. In 2005, he underwent left tympanic neurectomy, which led to temporary relief, with recurrence of swelling and pain about 1 month later. In 2006, Botox injections of the parotid glands were also unsuccessful, providing little to no relief. A sialogram was performed in 2010 showing hallmark signs of sialosis. Having failed other techniques of management, an intraductal triamcinolone instillation was offered. Ultimately, this led to a significant improvement in his pain, although minimal improvement was made in his swelling. A second lip biopsy (left side) was taken in 2012 confirming absence of Sjogren’s disease.

Introduction

Sialosis (sialadenosis) is a chronic, bilateral, diffuse, non-inflammatory, non-neoplastic painless swelling of the major salivary glands that primarily affects the parotid glands, but occasionally involves the submandibular glands and rarely the minor salivary glands (Scull 2008). The current case is presented to amplify upon previous work presented (Tokita 2013) and will present diagnostic workup used to aid in diagnosis. This patient also demonstrates the range of options available for management of sialosis – tympanic neurectomy, Botax injections and ultimately steroid infusion. The final option for management, parotidectomy, was ultimately not needed.

Discussion

Sialosis may be idiopathic but is also associated with chronic malnutrition, obesity, diabetes mellitus, alcoholism, liver disease, eating disorders and medication, particularly antihypertensives (Duggan, Chen). In this case, the patient had risk factors of diabetes and hypertension medications.

Initial workup in the case of chronic bilateral parotid swelling with new onset of pain should rule out infectious, neoplastic, or autoimmune etiologies. Minor salivary gland biopsy is an excellent tool to rule out Sjogren’s disease. Minor salivary glands, however, are not adequate for diagnosis of parotid sialosis. CT and MR demonstrated bilaterally swollen parotid glands in this patient. The finding on CT most suggestive of sialosis is fatty replacement of the affected gland along with enlargement of that gland. MRI is an excellent study to rule out neoplastic and inflammatory disease. Decreased arborization and splaying of ducts are the hallmarks of sialosis on sialography. While not performed in this case, sialendoscopy often reveals no significant intraluminal abnormalities associated with sialosis.

In our presentation, interventions for management of sialosis include tympanic neurectomy (Vasama), physically denervating the sympathetic input to the parotid, thus reducing function. Additionally, botulinum toxin injection was attempted, functionally performing chemodenervation of the glands to reduce function (Guntinas-Lichius). Parotidectomy could be considered for refractory cases, but even this does not guarantee success (Sadegh). This patient ultimately received benefit from triamcinolone (Kenalog-10®) infusion. The mechanism by which the infusion provided benefit is not clear in that sialosis is not an inflammatory disease process. This presentation is the first to report improvement of sialosis from steroid infusion.

The pathophysiology of sialadenosis remains unclear but the currently proposed hypothesis is that unbalanced autonomic input to the glands or abnormal stimulation as in diabetes or bulimia, leads to acinar hypertrophy.

Conclusions

Sialosis is a complex salivary pathologic process that may present a diagnostic challenge. The association with systemic diseases that are increasing -- such as diabetes mellitus and obesity -- will likely result in an increase in this condition. Key to the diagnosis is a history, physical exam, and radiographic imaging to exclude other inflammatory salivary diseases – supplemented in appropriate cases by serum evaluation and cytology (FNA). Sialography reveals classic patterns that can aid in the diagnosis of sialosis and exclude other causes of parotid swelling.

Management options used to address sialosis are varied and include observation, botulinum toxin A injection, tympanic neurectomy and parotidectomy. In this case, triamcinolone infusion ultimately provided relief after failure of response to left tympanic neurectomy and bilateral botulinum toxin injection.

References