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A rare case of Kussmaul Disease (Sialodochitis Fibrinosa)

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Abstract

Sialodochitis fibrinosa (or commonly known as Kussmaul Disease) is a rare salivary gland disease characterized by recurrent salivary gland swelling and pain as a result of mucofibrinous plugs. Typically patients have a history of multiply recurrent glandular swelling, dehydration and/or decreased salivary flow, thick secretions from Stensen's or Wharton's duct, and/or history of allergic diseases. Retention of mucofibrinous plugs may lead to acute suppurative parotitis and chronic sialadenitis ultimately. The diagnosis is one of exclusion, and treatment is based on symptomatology and largely supportive.

Introduction

Sialodochitis fibrinosa is characterized by recurrent salivary gland swelling caused by obstruction of the salivary ductal system by mucofibrinous plugs. Diagnosis of sialodochitis fibrinosa is difficult, and may be easily confused with either acute inflammatory disorders (viral or bacterial infection) or chronic conditions (Sjögren's Syndrome) that may subsequently cause salivary gland swelling. Although literature on the disease is sparse, prior cases implicate an allergic association. The etiology of the disease is not well known and treatment is largely supportive, including sialagogues, rehydration, anti-histamines, and/or enlargement of the duct orifice. Here, we present a rare case of sialodochitis fibrinosa, discussing the patient's history, symptoms, diagnosis, and treatment.

Case Study

A 56-year-old woman presented with complaints of intermittent left parotid and submandibular swelling that has recurred over the past 25 years. The swelling was associated with pain without any overlying erythema. Swelling and pain are worsened by food and improved by compression of gland. She did not have complaints of significant xerostomia. The patient tried multiple sialogogues over the years with minimal improvement of her symptoms. She denied dysphagia, odynophagia, hoarseness, hemoptysis, hematemesis

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or weight loss. She had similar symptoms on the right but were less severe in frequency and duration.

On physical exam, she had diffuse swelling of her left parotid and submandibular gland that is tender upon palpation. She did not have facial palsy. She does not have any palpable cervical nodes. Intraoral examination was unremarkable except for palpation of the parotid duct, which expressed thick mucoid secretions without purulence. Flexible fiberoptic examination of the larynx was unremarkable except for thick secretions throughout the aerodigestive tract. MRI was performed to further examine her swelling and revealed dilation of the parotid and submandibular ductal system without evidence of malignancy (Figure 1).

Discussion

First reported by Kussmaul in 1879, sialodochitis fibrinosa (or commonly known as Kussmaul Disease) is a rare salivary gland disease characterized by recurrent salivary gland swelling and pain as a result of mucofibrinous plugs.[1] Obstruction of the outflow or collecting duct by these plugs results in proximal swelling of the gland and a dilated ductal system, affecting either the parotid or submandibular glands. Very few cases have been described in the literature, making it difficult to establish clear diagnostic criteria for Kussmaul disease. However, patients found to have sialodochitis fibrinosa do typically have some combination of findings: a history of multiply recurrent glandular swelling, dehydration and/or decreased salivary flow; thick secretions from Stensen's or Wharton's duct; history of allergic diseases, increased blood eosinophil or serum IgE; histopathological changes of duct thickening; or stromal infiltration by lymphocytes or eosinophils. Retention of mucofibrinous plugs may lead to acute suppurative parotitis and chronic sialadenitis ultimately.

The diagnosis of sialodochitis fibrinosa is difficult as many of the symptoms and findings are nonspecific. Thus it is necessary to rule out the more common causes of salivary gland swelling such as sialolithiasis, Sjögren's syndrome or salivary gland tumors. Additionally, certain drugs, such as isoproterenol, ethambutol, iodine compounds and heavy metals can cause salivary gland enlargement as a side effect. Visualizing mucous plugs at the salivary duct orifice, grossly or on imaging is more specific for Kussmaul's disease. Laboratory tests and imaging can also prove beneficial in ruling out autoimmune etiologies and revealing the presence of stones or calculi as the cause of swelling.

The etiology of sialodochitis fibrinosa is not well understood, and unfortunately, our understanding of the disease is based on isolated cases reported throughout the literature. Although several mechanisms have been proposed, many of the reported cases have illustrated an allergic association. Almost a century ago, Von Reuss presented the case of a 16-year-old girl whose parotid swellings coincided not only with her menses, but interestingly, also with peripheral eosinophilia.[2] Waldbott and Shea noted cases of salivary gland swelling in relation to certain food allergies, where avoidance of specific foods diminished symptoms.[3] As a result, they postulated that allergic parotitis was either a result of an allergic reaction in the gland itself, or due to thick mucus production causing

duct obstruction. Harkness presented two similar cases of recurrent submandibular gland swelling that could not be attributed to any known disease process.[4] Histological examination of one patient showed dense eosinophilic infiltration, while the second patient had peripheral eosinophilia of 8%. Both of these patients demonstrated a decrease in swelling and symptoms in response to antihistamines, lending further credence to an underlying allergic mechanism. Bacterial infection may be a secondary etiology or consequence in sialodochitis fibrinosa. Although most reports implicate an underlying allergic cause for the disease pathology, several cases have identified α -streptococcus in mucous plug analysis.[5,6]

The management of sialodochitis fibrinosa is largely dependent on the severity of the disease and symptomology. Rehydration with gentle massage to help expel the thick mucous and plugs may reduce swelling and pain. Enlarging the duct orifice to enable plug expulsion and irrigation of the gland with a saline or steroid solution may also be utilized. Sialogogues may be added to treatment regimens to help improve salivary flow and aid in plug expulsion. Antihistamines and/or steroids may prove beneficial, especially considering the allergic underpinnings associated with the disease. In rare cases, patients may develop significant parotid gland enlargement, for which parotidectomy may prove beneficial.

In considering the case of our patient, we unexpectedly did not find any medical history of allergic diseases or food allergies. Physical intraoral exam showed evidence of mucofibrinous plugs at the duct orifice along with thick mucoid discharge. Additionally, CT imaging confirmed the diagnosis by illustrating a large plug in the parotid gland and a dilated ductal system. As a result, blood eosinophilia was not performed, nor was histopathology of the duct or surrounding stroma.

With regards to treatment for our patient, massage did not provide relief for her symptoms. Additionally, sialogogue usage (Cevimeline) resulted in hallucinations and sleep disturbances as side effects. The patient noted that her use of Benadryl had (paradoxically) provided moderate pain relief. As a result, we tested both Robinul and Scopolamine to further control her symptoms; however the patient stated that neither treatment options were effective in decreasing pain or swelling. We next tried Botox injections with limited success. However lidocaine injections seemed to offer temporary relief of her pain and symptoms, so she opted to undergo left sided greater auricular nerve transection. Although this procedure did temporarily resolve her complaints and symptoms, her symptoms did reappear after six months.

References

1. Kussmaul, I. Anfallweise auftretende Speichelgeschwulst. Vol. 15. Berlin Klin Wschr; 1879.
2. Von Reuss A. Jahrbuch für kinderheilkunde. 1909; 70 n.p.
3. Waldbott GL, Shea JJ. Allergic parotitis. The Journal of allergy. Jan; 1947 18(1):51–54. [PubMed: 20282282]
4. Harkness P. Submandibular salivary disease: a proposed allergic aetiology. The Journal of laryngology and otology. Jan; 1995 109(1):66–67. [PubMed: 7876744]
5. Cohen L. Recurrent Swelling of the Parotid Gland; Report of a Case Due to Allergy. British dental journal. Jun 1.1965 118:487–490. [PubMed: 14290852]

6. Yoshioka YMK, Sakoda K, et al. Sialodochitis fibrinosa of the parotid glands: a case report. *Kokuintoka Gakkaishi*. 1997; 9:409–416.

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Figure 1.

Contrast Enhanced MRI scan of the head and neck showing typical findings in, Kussmaul's Disease. Panel A shows an Axial T1 weighed image with dilated parotid ducts (arrows). Panel B shows an Axial T2 weighed image highlighting the same ducts (arrows). Panel C shows a coronal T1 weighed image showing dilated parotid and submandibular ducts (arrows).