Case of Suspected Sialodochitis Fibrinosa (Kussmaul’s Disease)

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Abstract

Here we report a case of Kussmaul’s disease, or sialodochitis fibrinosa. This rare disease is characterized by recurrent swelling of the salivary glands, which then discharge clots of fibrin into the oral cavity. An 80-year-old man with a history of allergic rhinitis visited our department with the chief complaint of pain in the bilateral parotid gland area on eating. An initial examination revealed mild swelling and tenderness in this region, and indurations could be felt around the bilateral parotid papillae. Pressure on the parotid glands induced discharge of gelatinous plugs from the parotid papillae. No pus was discharged, and there were no palpable hard objects. Panoramic X-ray showed no obvious focus of dental infection, and there was no calcification in the parotid gland region. Magnetic resonance imaging revealed segmental dilatation of the main ducts of both parotid ducts, with no signs of displacement due to sialoliths or tumors, or of abnormal saliva leakage. Two courses of antibiotic therapy resulted in no improvement. During treatment, gelatinous plugs (fibrin clots) obstructing the left parotid duct were dislodged by massage, which prevented further blockage by encouraging salivary outflow. The obstruction persisted in the right parotid duct, however. Therefore, the distal portion of the right parotid duct was partially resected and the opening into the mouth enlarged, which, in combination with massage, prevented further obstruction. The pain and swelling of the parotid gland and discharge of gelatinous plugs improved, with no further recurrence at 12 months postoperatively. This case is presented along with a review of the relevant literature.

Key words: Sialodochitis fibrinosa — Kussmaul’s disease — Fibrin clot — Allergy — Salivary gland

Introduction

First reported in 1879, Kussmaul’s disease, or sialodochitis fibrinosa, is rare. It is characterized by episodic swelling of the salivary glands, which then discharge clots of fibrin into the oral cavity\textsuperscript{5}). Although firm diagnostic criteria remain to be established, a number
of studies have listed the following as typical findings in this disease: (1) episodic swelling of the salivary glands; (2) discharge of plugs containing numerous eosinophils from the main ducts; (3) elevated levels of eosinophils and IgE; (4) concomitant presence of other allergic disorders; (5) stenosis and irregular dilatation of the main duct evident on ptyalography; and (6) hypertrophy of the main ducts, edema, and lymphocyte infiltration of the interstitium surrounding the salivary ducts.

Here we report a case of suspected sialodochitis fibrinosa.

**Case Report**

The patient was an 80-year-old man who presented at our hospital in October 2012 with the chief complaint of bilateral pain in the parotid gland area. The patient had first noticed swelling and tenderness in the area of the left parotid gland only. Subsequently, he had experienced pain bilaterally in this area when eating. His previous medical history revealed allergic rhinitis, an enlarged prostate gland, surgical intervention for thyroid cancer, and mumps in childhood. His temperature on the initial visit was 36.0°C. A blood test showed the following results: CRP, 0.01 mg/dl; WBC, 5,500/dl (neutrophils 65.2%, eosinophils 1.3%, basophils 0.3%, monocytes 5.0%, lymphocytes 28.2%); RBC, 5×10⁶/dl; and platelet count, 25×10⁴/dl.

A physical examination revealed that the patient had no difficulty in opening his mouth. Mild swelling and tenderness were observed bilaterally in the parotid gland area. Indurations could be felt around the parotid papillae bilaterally. Pressure on the parotid glands bilaterally resulted in discharge of gelatinous plugs from the parotid papillae (Fig. 1). No discharge of pus was observed. Therefore, the bilateral parotid gland area was investigated in more detail by panoramic X-ray (Fig. 2) and MRI (Fig. 3). The results showed no obvious focus of dental infection, nor any tumorous lesions in the buccal or parotid areas on either side. Sialoliths were also absent. Retrograde oral bacterial infection via the parotid papillae was suspected, but administration of 750 mg/day amoxicillin for 2 weeks did not improve the symptoms.

**Treatment and Course**

At his initial visit, the patient complained of bilateral pain in the parotid gland area. He had no difficulty in opening his mouth. Mild swelling and tenderness were observed bilaterally in the parotid gland area. Indurations could be felt around the parotid papillae bilaterally. Pressure on the parotid glands bilaterally induced discharge of gelatinous plugs from the parotid papillae (Fig. 1). No discharge of pus was observed. Therefore, the bilateral parotid gland area was investigated in more detail by panoramic X-ray (Fig. 2) and MRI (Fig. 3). The results showed no obvious focus of dental infection, nor any tumorous lesions in the buccal or parotid areas on either side. Sialoliths were also absent. Retrograde oral bacterial infection via the parotid papillae was suspected, but administration of 750 mg/day amoxicillin for 2 weeks did not improve the symptoms.
Swelling of the bilateral parotid gland area subsequently recurred, and pressure on the parotid glands induced discharge of gelatinous plugs from the parotid papillae. On the 15th day, the patient was switched to 200 mg/day clarithromycin and instructed to be conscientious in massaging the parotid glands. The pain was alleviated with discharge of the gelatinous plugs, but reappeared at approximately weekly intervals. On the 45th day, the symptoms in the left parotid gland improved, and by the 60th day, the pain had disappeared. On the right side, however, although improving somewhat, the pain persisted.

On the 100th day (February 2013), the patient noticed worsening of the pain in the right parotid gland area. The area around the right parotid papilla was tender, and indurations could be felt in that region. Detailed examination of the bilateral parotid gland area by computed tomography (CT)
(Fig. 4) and FDG-PET/CT (Fig. 5) revealed no dental infection, tumorous lesion, or pressure on the parotid glands due to such a lesion; sialoliths were also absent. Retrograde oral bacterial infection via the parotid papillae was suspected, and 750 mg/day amoxicillin was administered for 2 weeks. Although the symptoms improved somewhat, the tenderness did not resolve. Salivary outflow from the right parotid papilla was poor, and a lacrimal duct bougie was therefore inserted. This resulted in discharge of gelatinous plugs. No pus was discharged.

On the 130th day (March 2013), the area around the right parotid papilla was anesthetized with 1.2 ml of 1% lidocaine containing 1/100,000 epinephrine. A lacrimal duct bougie was then inserted via the parotid papilla and a sharp-pointed scalpel used to resect the distal portion of the parotid duct immediately above the bougie, enlarging the opening of the parotid duct into the mouth. Clarithromycin was administered for 2 weeks postoperatively at 200 mg/day; salivation was induced by taste stimulation, and parotid gland massage conscientiously carried out. Postoperatively, the pain and swelling of the parotid gland and discharge of gelatinous plugs improved, with no recurrence on either side as of March 2014.

The differential diagnoses for sialodochitis fibrinosa include mumps, sialolithiasis, recurrent parotiditis, purulent parotiditis, Sjögren’s syndrome, and periparotid tumor. In the present case, blood tests revealed no indications of inflammation, ruling out mumps, recurrent parotiditis, purulent parotiditis, and Sjögren’s syndrome. Sialolithiasis, parotid tumor, and periparotid tumor were ruled out based on the CT (Fig. 4), MRI (Fig. 3), and PET/CT (Fig. 5) findings. Bilateral sialodochitis fibrinosa was diagnosed based on the recurrent swelling of the parotid glands, the discharge of gelatinous plugs from the parotid papillae (Fig. 1), the segmental dilatation of the parotid ducts evident on MRI (Fig. 3), and the patient’s history of allergic rhinitis.

**Discussion**

Kussmaul described sialodochitis fibrinosa
as “a disease in which the formation of fibrinous plugs within the salivary ducts causes the ducts to become blocked, causing recurrent painful swelling of the salivary glands”\(^5\). Relatively few Japanese cases have been reported\(^6\), but its diagnosis is relatively easy, and the actual number of patients may be greater than the number of reports would suggest.

Sialodochitis fibrinosa is characterized by recurrent swelling of the salivary glands, which may also be painful\(^4,5,9-11\). Pressure on the salivary glands when swollen induces discharge of white fibrin clots, which then alleviates any pain. Most frequently occurring in the parotid glands, followed by the submandibular glands, it has also been reported to occur bilaterally\(^4,5,9-11\). Although rarely seen in children, onset may occur at any time between young adulthood and old age, and it is more common in women\(^3,5,10-12\).

The etiology of this disease remains to be fully clarified. However, it tends to occur in the parotid glands, it is often seen where there is a history of disorders such as allergic rhinitis and bronchial asthma, and eosinophil counts are elevated, which all suggests the involvement of allergy\(^1,3,4,8,9,11,12\). In a case in which the symptoms improved when antigens were eliminated, the condition was described as allergic parotiditis\(^4,5,8,9,11\).

Sela et al. carried out basic experiments using human serum in rat parotid gland to produce an animal model of allergic parotiditis\(^6,12\). Histological examination revealed both acute inflammation and degeneration of the salivary gland parenchyma, which suggests the predominance of type III hypersensitivity in this condition.

Few histopathological investigations into this disease have been published. However, some studies have reported the presence of eosinophils, neutrophils, and fibrin clots containing detached epithelium in the salivary ducts, while others have described hypertrophy of the salivary duct epithelium, as well as hyalinization, edema, and lymphocyte infiltration of the surrounding interstitium\(^5,7\). It progresses from the main duct. Moreover, the majority of eosinophils are the EG2-positive active type, and there is marked eosinophil infiltration of the salivary duct interstitium, which suggests an allergic antigen-antibody reaction in the main salivary ducts, resulting in release of IL5 and other cytokines from T lymphocytes and other cells. This in turn induces aggregation of eosinophils, the formation of white gelatinous fibrin clots, obstruction of the salivary ducts, and swelling\(^3,4,9-11\).

In another report on a patient treated with total parotidectomy, it was noted that although swelling in the parotid gland area was alleviated postoperatively, white fibrin clots continued to be discharged from the opening of the Stensen’s duct. This suggests that sialodochitis fibrinosa is an inflammation of the salivary ducts, rather than of the salivary glands\(^5\).

Although firm diagnostic criteria remain to be established, various reports have noted the following as typical of this disease: (1) episodic swelling of the salivary glands; (2) discharge of plugs containing numerous eosinophils from the main ducts; (3) elevated levels of eosinophils and IgE; (4) concomitant presence of other allergic disorders; (5) stenosis and irregular dilatation of the main duct evident on ptyalography; and (6) hypertrophy of the main ducts, edema, and lymphocyte infiltration of the interstitium surrounding the salivary ducts\(^3,4,9,11\).

Drug therapy is the treatment of first choice. As an allergic reaction is implicated in sialodochitis fibrinosa, previous reports have described treatment with anti-allergic agents, antihistamines, proteases, and oral corticosteroids. Some patients do relapse, however, and topical injection of corticosteroids may be effective in such cases, although reports of patients being completely cured by this method are few. Surgical resection of the salivary glands has also been reported, but the salivary ducts must be removed at the same time as the glands themselves\(^4,10,11\).

In the present case, gelatinous plugs (fibrin clots) large enough to constitute an obstruction formed in the parotid duct. Here, massage proved effective in dislodging
these plugs and encouraging salivary outflow, preventing further blockage on the left side. Obstruction persisted on the right side, however. Part of the distal portion of the parotid duct was therefore resected and the opening into the mouth simultaneously enlarged, which, with subsequent continuation of massage, successfully prevented further obstruction. Although rather uncommon, sialodochitis fibrinosa should be borne in mind as a possible diagnosis in patients complaining of recurrent swelling of the salivary glands. Clinically, however, it is relatively easy to diagnose on the basis of its characteristic signs, which include discharge of white fibrin clots from the opening of the salivary ducts into the mouth and dilatation of the main duct evident on ptyalography.

More in-depth studies are required to elucidate the pathology of sialodochitis fibrinosa and establish a method of treatment.

**Conclusion**

A case of suspected sialodochitis fibrinosa was described.

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