



A Case of Allergic Fungal Rhinosinusitis Associated With Abducens Nerve Palsy

Kenzo Tsuzuki*, Kengo Hashimoto, Ken Okazaki, Kazutaka Kuroda and Masafumi Sakagami

Department of Otolaryngology - Head and Neck Surgery, Hyogo College of Medicine, Japan

Abstract

Background: Allergic Fungal Rhinosinusitis (AFRS) is characterized by Chronic Rhinosinusitis (CRS) with nasal polyps (CRSwNP), type I and III hypersensitivity reaction, production of allergic mucin with abundant eosinophils, and non-invading fungi. Ophthalmic complications due to cranial neuropathies are uncommon in AFRS. A case of AFRS with abducens nerve paralysis is reported.

Patient Description: A 66-year-old man presented with worsening diplopia and nasal obstruction. Ophthalmologists diagnosed left abducens nerve paralysis. Edematous and polypoid mucosa with viscous discharge was observed in the left middle and superior nasal meatuses. Blood examination showed hyperglycemia, eosinophilia, and antigen-specific IgE positive to *Aspergillus*. Opacification of a high-density area with sphenoid bony deficiency was seen on computed tomography and iso- and low intensities were observed on T1- and T2- weighted magnetic resonance imaging in the left maxillary, posterior ethmoid and sphenoid sinuses. Left Endoscopic Sinus Surgery (ESS) was performed urgently under general anesthesia. Polypoid mucosa with viscous contents was observed in the maxillary, posterior ethmoid and sphenoid sinuses during ESS. Histopathological examination of the sphenoid sinus showed *Aspergillus* in the contents and marked eosinophilic infiltration without fungal infiltration into the mucosa. Consequently, the definitive diagnosis was AFRS. At postoperative 3 months, ophthalmologists diagnosed the diplopia as cured. Currently 6 months after surgery, there is no evidence of suspected recurrence or exacerbation.

Conclusion: AFRS should be suspected when nasal polyposis with viscous discharge and cranial nerve paralysis are seen, and it should be carefully treated surgically.

Keywords: Allergic Fungal Rhinosinusitis (AFRS); Abducens nerve paralysis; Endoscopic Sinus Surgery (ESS)

Introduction

Allergic Fungal Rhinosinusitis (AFRS) is characterized by Chronic Rhinosinusitis (CRS) with nasal polyps (CRSwNP), a type I (raised IgE) and possibly type III hypersensitivity reaction, production of allergic mucin with abundant eosinophils, and non-invading fungal hyphae [1]. Approximately 7% to 10% of operations for CRS are due to AFRS in the USA [2]. AFRS is thought to be refractory CRS. For a definite diagnosis, there are the classic criteria of Bent and Kuhn [3] and the clinical guidance of the American Academy of Allergy, Asthma & Immunology (AAAAI) [4]. Taken together, we diagnose AFRS when all six essential items of both criteria are fulfilled: (1) Sustained CRS symptoms for more than 12 weeks (such as nasal discharge, nasal obstruction, olfactory disorder, or facial pain); (2) Characteristic findings of rhinosinusitis on Computed Tomography (CT) or Magnetic Resonance Imaging (MRI); (3) Presence of allergic mucin on endoscopy (pathologically confirmed presence of fungi and mucosal infiltration of eosinophils); (4) Edema/polyp formation of the middle nasal meatus or middle turbinate on endoscopy; (5) Type I hypersensitivity to fungi (elevated blood serum levels of specific IgE to fungi or positive intracutaneous test); and (6) Pathologically ruled out infiltration of fungi into the surrounding mucosa. Previous studies reported that, in AFRS, ophthalmic complications due to cranial neuropathies are uncommon [5,6]. A case of AFRS that presented with diplopia due to left abducens nerve palsy is reported.

Case Presentation

A 66-year-old man presented to Hyogo College of Medicine with diplopia and nasal obstruction. His nasal obstruction was treated by nearby ENT clinics for more than three months. Because double vision appeared in addition to the exacerbation of nasal symptoms one month prior to his visit,

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*Correspondence:

Kenzo Tsuzuki, Department of Otolaryngology – Head and Neck Surgery, Hyogo College of Medicine, 1-1 Mukogawa, Nishinomiya, Hyogo 663-8501, Japan, Tel: 81798456493;

Fax: 81798418976;

E-mail: kenzo@hyo-med.ac.jp

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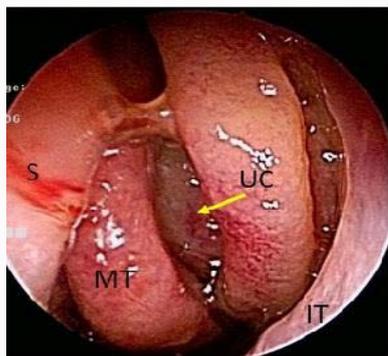


Figure 1: Preoperative left nasal endoscopic findings. Edematous and polypoid mucosa is observed in the left middle nasal meatus (arrow). MT: Middle Turbinate; IT: Inferior Turbinate; S: Nasal Septum; UC: Uncinate Process

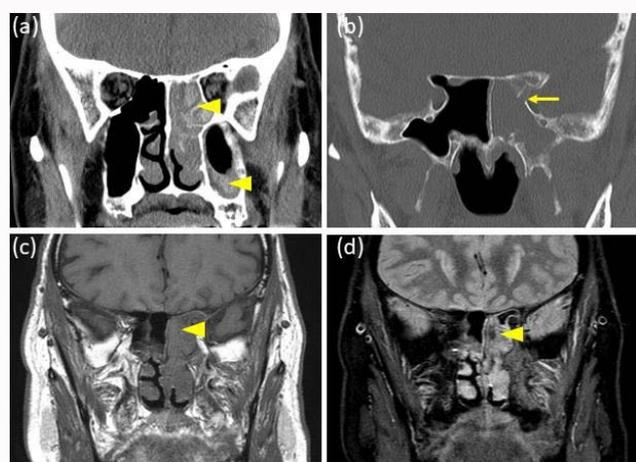


Figure 2: Preoperative findings on sinonasal imaging. CT (a, b) and MRI (c, d): Arrowheads indicate the viscous effusion suggesting eosinophilic mucins in the maxillary sinus, posterior ethmoid sinus, and sphenoid sinus on the left side (a, c, d); the arrow indicates a thin bony deficiency in a part of the left sphenoid bone around the superior orbital fissure (b).

he was referred to our department for treatment. On consultation with the Department of Ophthalmology in our hospital, the double vision was diagnosed as left abducens nerve palsy. The patient also had diabetes mellitus.

Edematous and polypoid mucosa was observed in the left middle nasal meatus on endoscopy (Figure 1). Around the left superior nasal meatus and superior nasal turbinate, edematous and polypoid mucosa with viscous discharge was also observed. No fungi were detected in the viscous discharge on culture. No abnormality was observed in the right nasal cavity. Blood examination showed hyperglycemia (144 mg/dl), HbA1c-NGSP (7.0%), Eosinophilia (24.4%), and Antigen-specific IgE positive to *Aspergillus* (Class 2). The antigen-nonspecific IgE level (53.3 IU/ml) was within the normal range (<173 IU/ml). Beta-D-glucan was also within the normal range (<11.0 pg/ml). CT showed opacification with a High-Density Area (HDA) in the maxillary sinus, posterior ethmoid sinus, and sphenoid sinus on the left side (Figure 2a). A thin bony deficiency was observed in part of the left sphenoid bone around the superior orbital fissure (Figure 2b). T1- and T2-weighted MRI showed iso- and low intensities, respectively, in the left posterior ethmoid sinus, and sphenoid sinus (Figure 2c and 2d). No intracranial lesions were observed.

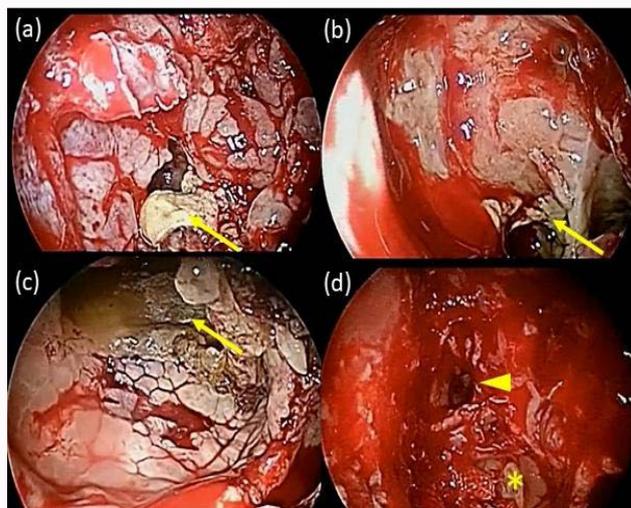


Figure 3: Surgical findings. The polypoid mucosal swelling and viscous contents in the posterior ethmoid sinus (a), sphenoid sinus (b), and maxillary sinus (c). The anterior ethmoid sinus showed edematous mucosal swelling without viscous contents (d, asterisk). Inflammation was not observed in the frontal sinus (d, arrowhead). Arrows indicate viscous contents.

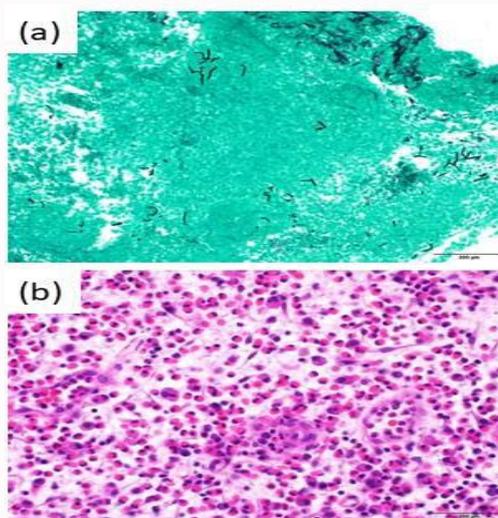


Figure 4: Histopathological findings in the sphenoid sinus. (a) Grocott stain shows the presence of *Aspergillus* in the sphenoid sinus contents. (b) H&E stain shows marked eosinophilic infiltration without fungal infiltration in the sinus mucosa. Scale bars: 200 μm.

In order to remove and decompress the mucinous lesions immediately, left Endoscopic Sinus Surgery (ESS) was performed urgently under general anesthesia for the abducens nerve palsy due to sphenoid sinusitis on the day of the initial visit to our department in the hospital. Blood loss was less than 10 mL. Surgical findings of the paranasal sinuses showed polypoid mucosal swelling and viscous contents in the posterior ethmoid sinus, sphenoid sinus, and maxillary sinus (Figure 3). Although the anterior ethmoid sinus showed edematous mucosal swelling, there was no retention of viscous contents. Inflammation was not observed in the frontal sinus. Histopathological examination of the specimens of the sphenoid sinus during ESS showed *Aspergillus* in the contents (Grocott stain, (Figure 4a) and marked eosinophilic infiltration without fungal infiltration into the sinus mucosa (Hematoxylin-Eosin (H&E) stain, (Figure

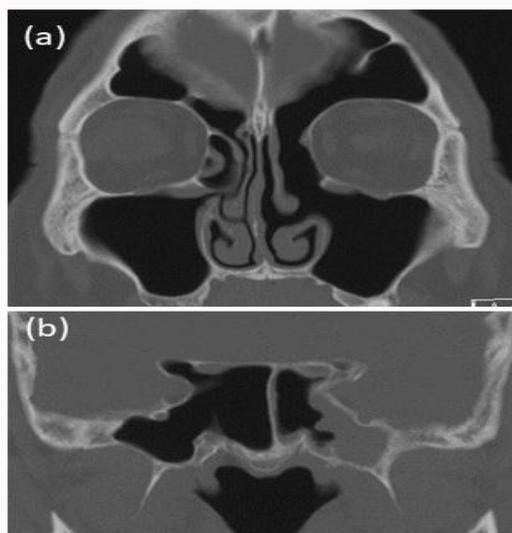


Figure 5: Postoperative CT findings.

Three months after ESS with the diplopia considered cured, no abnormality is found in the maxillary, ethmoid, and frontal sinuses (a), except for partial mucous membrane swelling remaining in the left sphenoid sinus (b).

4b)). Since this case fulfilled all six essential items of the diagnostic criteria (mentioned above), a definite diagnosis of AFRS was made. No antifungal medicine was administered before and after surgery. Corticosteroids (steroids) were not given systemically because of the history of diabetes mellitus. Since the patient's postoperative course was uneventful, he was discharged on postoperative day 5. Three months after ESS, the ophthalmologists declared the diplopia cured. On endoscopy and CT, partial mucous membrane swelling remained in the left sphenoid sinus, but no abnormality was found in other paranasal sinuses (Figure 5). Currently, 9 months after ESS, there have been no findings suggestive of recurrence or exacerbation.

Discussion

A case of abducens neuropathy due to AFRS was reported, since, to the best of our knowledge, there have been no case reports of abducens neuropathy due to AFRS in Japan according to a previous nationwide report [7]. A total of 37 cases of AFRS fulfilling the diagnostic criteria of AAAAI have been reported in Japan, and 8% of them had ophthalmic symptoms (3/37 cases: 2 cases with exophthalmos and one case with decreased vision) [7]. Although there was a report that 3% (3/94) of AFRS cases in the USA had abducens paralysis, its occurrence is thought to be uncommon [6].

As mechanisms causing cranial neuropathies due to rhinosinusitis, inflammation affecting adjacent nerves and mechanical compression, which can cause direct nerve compression and/or a circulatory disorder due to vascular compression, can be considered. The abducens nerve is the most common cranial nerve to be affected due to its medial anatomical location in the cavernous sinus and proximity to the sphenoid sinus, followed by the optic nerve [8]. In AFRS, the volume of allergic eosinophilic inflammatory mucinous products of fungal colonies increases gradually and their expansion causes bony demineralized erosion and nerve compression [9]. Approximately 20% of patients with AFRS demonstrated bone erosion on CT scan [10], and male patients were reported to be at higher risk of bone erosion than female patients [11]. Therefore, the pathogenesis of diplopia in the present case could be the result of inflammation of

the sphenoid sinus spreading directly to the abducens nerve and expansive compression of the abducens nerve due to eosinophilic mucin produced by the fungi with sphenoid bone destruction.

With respect to the management of Fungal Rhinosinusitis (FRS), accurate early diagnosis before treatment is very important, because the medical treatment strategies for infectious and non-infectious (AFRS) diseases differ. For infectious FRS, immune suppressors, such as steroids, can never be used, because they can stimulate and exacerbate fungal infiltration. In cases of invasive type, antifungal therapy is required until serum beta-Dglu can levels decrease to within normal limits [12]. On the other hand, for AFRS, steroids are indicated and useful, with careful attention to Cushingoid side effects, where as the benefits of antifungal therapy are still unclear [13]. A combination of steroids and ESS is required, and oral steroids can subsequently lead to short-term postoperative improvement [13]. Although oral steroids could not be administered to the present case with diabetes mellitus, steroids might be required for future recurrence of AFRS.

In the differential diagnosis, Eosinophilic CRS (ECRS) is one of the most important diseases. Based on the Japanese Epidemiological Survey of Refractory Eosinophilic Chronic Rhinosinusitis (JESREC) study, ECRS is definitely diagnosed by the following four items: bilateral lesions; nasal polyps; ethmoid sinus-dominant; and blood eosinophilia [14]. The majority of ECRS cases have bilateral involvement, whereas less than 30% of AFRS cases show bilateral involvement in the future. Unilateral AFRS might become bilateral in the future. Some patients with unilateral AFRS certainly fulfill the JESREC criteria because of CRSwNP with severe eosinophilia (>10%), as shown in the present case. Patients with ECRS who fulfill the diagnostic criteria for AFRS might also exist. It is important when making the diagnosis to consider the possibility of AFRS by being sufficiently aware of the disease, though the pathogenesis of AFRS still remains controversial, with some uncertain issues [13].

In the present case, ESS was performed one month after the onset of ophthalmic symptoms, and fortunately the abducens paralysis improved. ESS is necessary in the earlier stage for rhinogenic ocular symptoms [10]. The ocular manifestations of AFRS can be reversible if promptly and appropriately addressed and decompression is performed [6,10]. Fortunately, no recurrence and good progress have been seen in the present patient. However, since AFRS is thought to be refractory and have a high possibility of recurrence, continuous careful long-term follow-up observation is required [15].

Conclusion

We encountered an uncommon case of AFRS that presented with abducens neuropathy, which has not been previously reported in Japan. Lessons learned from the present case are that what we should take into consideration the possibility of AFRS when we observe nasal polyposis with viscous nasal discharge and cranial nerve paralysis, and what ophthalmic symptoms should be treated surgically in the early stage.

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