A case of allergic fungal rhinosinusitis

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Abstract

Allergic fungal rhinosinusitis (AFRS) is a type of paranasal sinusitis, the cause of which is related to an allergic reaction to fungal infection. This condition has been called “allergic mucin”, characterized by the presence of eosinophils concentrated in the mucosal fluid. AFRS in Japan is still rare or might be un-/misdiagnosed by the lack of proper recognition of this specific condition. In this report, we describe a case of AFRS caused by Alternaria fungi infection. The case is 40-year-old woman. The first recommendations in the treatment for AFRS are surgery. However, when postoperative treatment is not appropriate, the recurrence rate may increase. In this case, we did operation for two times. The nature of AFRS is intractable, and careful follow-up and additional treatments may be necessary even after surgery.

Key words: Alternaria, allergic fungal rhinosinusitis, AFRS

Introduction

Allergic fungal rhinosinusitis (AFRS) is a type of paranasal sinusitis, the cause of which is related to an allergic reaction to fungal infection. Fungi grow in the mucous membrane lining the nasal cavity, remain there, and never cause systematic invasion. This condition has been called “allergic mucin” characterized by the presence of eosinophils concentrated in the mucosal fluid. Dematiaceous fungi and Aspergillus have often been reported as the causative agents. The number of reports concerning AFRS has been increasing worldwide after Millar’s1 first report in 1981, but AFRS in Japan is still rare or might be un-/misdiagnosed by the lack of proper recognition of this specific condition. In this report, we describe a case of AFRS caused by Alternaria fungi infection, along with a literature review.

Case report

Case: 40-year-old woman
Chief complaint: Stuffy nose
Medical history: The patient visited our ear, nose, and throat (ENT) department of Kinki University Hospital since she had been aware of a stuffy nose for 1 year. Nasal endoscopic inspection revealed polyp formation occupying the

Fig. 1 CT images before the first operation
entire left nasal cavity. Plain Computed Tomography (CT) imaging confirmed the presence of soft tissue densities located in the left frontal, ethmoidal, and maxillary sinuses, and bone destruction in part of the left maxillary sinus (Figure 1). Bone defects were present in some parts of the maxillary sinus cavity, which suggested the possibility of malignant disease.

Past history: Aspirin asthma
Serologic examination: Leukocytes, 7,200/μl (eosinophils: 5.9%); total serum IgE, 684 IU/ml (standard level: under 295 IU/ml); specific IgE value (standard level: under 0.34 IU/ml) to Aspergillus: 1.51, Alternaria: 10.0, Candida: 1.23.

Surgical findings: Endoscopic sinus surgery was conducted under general anesthesia. Polyps were first removed with a micro-debrider, and then the opening of the left maxillary, ethmoidal, and frontal sinuses was completed, which made it possible to discern that the fungal body looked like a peanut-butter lump in those sinuses.

Clinical course after surgery: The ethmoidal sinus became re-congested within 2 months after surgery, but no additional therapy was conducted. When the patient visited again two years after surgery, the recurrence of AFRS was indicated on the opposite side (Figure 2). She decided to receive a second surgery under general anesthesia. Almost the same procedure was performed to complete the opening of the right maxillary, ethmoidal, and frontal sinuses.

Prognosis: Although the patient experienced bilateral ethmoidal sinusitis within one month after the second surgery, conservative treatment with Montelukast sodium (nasal steroid drop) and daily sinus washing with saline at home gradually reduced mucosal swelling. Postoperative CT imaging indicated some mucosal swelling in the bilateral frontal sinuses, while other sinuses remained clear (Figure 3). The patient is now free from any subjective complaints.

Pathological examination: HE staining demonstrated an accumulation of infiltrated eosinophils and Charcot-Leyden crystals in the allergic mucin collected during the surgery (Figure 4), while Grocott stain detected a conidial chain and hyphal wall (Figure 5). Finally, Alternaria fungi were separated from the submitted specimen culture.
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Discussion

AFRS is a type of paranasal sinusitis, the cause of which is related to an allergic reaction to fungal infection. Millar's report, entitled: "Allergic aspergillosis of the maxillary sinuses", in 1981, is the first document in the literature. In 1989, the condition was defined as "Allergic fungal sinusitis", since non-Aspergillus fungi also produce the same pathology. Since then, a number of reports on AFRS have been published in Europe and the United States, while this specific condition has not well recognized in Japan. In 2006, the criterion for AFRS was proposed based on the guidelines of 5 medical associations including AAAAI (American Academy of Allergy Asthma and Immunology), in which “rhinosinusitis” was classified into the following four types:
1) Acute presumed bacterial rhinosinusitis (ABRS)
2) Chronic rhinosinusitis without nasal polyps (CRsNP)
3) Chronic rhinosinusitis with nasal polyps (CRsNP)
4) Allergic fungal rhinosinusitis (AFRS)

Detailed criteria for AFRS diagnosis including symptoms and objective documentations are provided as well (Table 1). The present case satisfies most symptoms and documentations and can be easily diagnosed according to these criteria.

It was reported that the prevalence of AFRS was 4-10% among the cases of chronic rhinosinusitis requiring surgery in Europe and the United States. There are a few domestic reports: the prevalence was 3.9% in Matsuwaki's and 8.3% in Nakayama's report, respectively. According to Cody's report, the AFRS causative agents are rated as follows: Bipolaris: 22.5%, Curvularia: 19.6%, Aspergillus: 14.7%, Dreichslera: 4.9%, Alternaria: 3.9%, Cladosporium:

Table 1 Criteria for AFRS (AAAAI, 2006)

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<th>Pattern of symptoms</th>
<th>Symptoms present For ≥12 wk</th>
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| Symptoms for diagnosis               | Requires ≥1 of the following symptoms:
|                                      | 1. Anterior and/or posterior drainage |
|                                      | 2. Nasal obstruction         |
|                                      | 3. Decreased sense of smell  |
|                                      | 4. Facial pain/pressure/fullness |
| Objective documentation              | Requires:                   |
|                                      | 1. Endoscopy to document presence of allergic mucin (pathology showing sparse fungal hyphae with degranulating eosinophils) and inflammation, such as edema of middle meatus or ethmoid area or NPs |
|                                      | 2. Evidence of rhinosinusitis by CT or MRI |
|                                      | 3. Evidence of fungal specific IgE (skin test or in vitro blood test) |
|                                      | 4. No histologic evidence of fungal invasion when risk factors for invasive fungal disease are present |
| Other possible documentation(Not required): | 1. Fungal culture |
|                                      | 2. Total serum IgE level     |
|                                      | 3. Imaging by more than one technique (CT or MRI) highly suggestive of AFRS |
Although AFRS caused by Alternaria was not common in the USA, further analyses is necessary to clarify the causative fungi in Japan. In the diagnosis of AFRS, identifications of allergic mucin, viral existence, and Charcot-Leyden crystals are critical. In our case, HE staining demonstrated the accumulation of eosinophils and Charcot-Leyden crystals in allergic mucin (Figure 4), and Grocott staining demonstrated the fungi in the specimen (Figure 5).

The first recommendations in the treatment for AFRS are surgery and systemic steroid administration. Conservative therapies such as applications of topical steroid, antifungal agents, and anti-leukotriene agents are also recommended. Surgical treatment is most effective to remove allergic mucin and the causative fungi, while preserving the normal mucous membrane and promoting drainage/ventilation of the paranasal sinuses. However, when postoperative treatment is not appropriate, the recurrence rate may increase. In this case, the treatment after the first surgery was just observation without any medication. As a result, recurrence of the disease occurred not only on the affected side, but also on the opposite side within two years. Therefore, the openings of the paranasal sinuses were fully dilated in the second surgery to prepare for postoperative treatment. Mucosal swellings in bilateral ethmoidal sinuses remained after one month. With applications of topical steroid drop, Montelukast sodium (anti-leukotriene), and daily washing of the sinuses with saline, mucosal swelling gradually improved. The patient has continued the same conservative treatments, and is still free from any objective symptoms, although CT scanning revealed some soft tissue densities in the frontal sinuses (Figure 3).

There have been a few reports on AFRS in Japan, indicating the possibility that a number of undiagnosed and misdiagnosed cases exist. It is unfortunate to say that this disease is not well recognized by pathologists and ENT doctors. Therefore, it is important for ENT doctors to ask pathologists whether or not the pathologies of AFRS and fungi are present in specimens collected during surgery. The nature of AFRS is intractable, and careful follow-up and additional treatments may be necessary even after surgery.

Conclusion

A case of allergic fungal rhinosinusitis (AFRS) caused by Alternaria fungi was reported. There is only one report except our report in Japan which caused by Alternaria fungi, as long as we researched. After two surgeries and subsequent conservative therapies, her AFRS became well controlled. The nature of AFRS is intractable, and careful follow-up and additional treatments may be necessary even after surgery.

References