



IgG4-Related Disease Mimicking Nasal Malignant Carcinoma with Multiple Organ Metastases: A Case Report

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Abstract

Objectives: The IgG4-related disease often mimics different malignant diseases. We presented a case of IgG4-related disease associated with lesion in nasal cavity, lung and kidney which is easily misdiagnosed as nasal carcinoma with multiple metastases.

Method: Single case report.

Results: The patient was finally diagnosed as IgG4-related disease under the examination of IgG4 in the nasal tissue and serum.

Conclusion: IgG4-related disease has possibility to exhibit the nasal mass accompanied by multiple pulmonary and renal nodules which mimic malignant tumors with multiple metastases in different organs. The doctors should attach much importance to the diagnosis of IgG4-related disease compared to the other nasal diseases.

Keywords: IgG4; Nasal malignant carcinoma; plasmacytes

Introduction

IgG4-related disease was first reported by Hamano *et al.* [1] in 2001. The disease was clinically characterized by increased serum IgG4 level, infiltration of IgG4-positive plasmacytes and sclerosing lesions which generate multiple nodules in different organs [2] that it sometimes mimics malignant tumors [3-5]. To date, IgG4-related disease has been shown to be well associated with nasal diseases [6-8]. Herein, we describe a case of nasal mass with multiple organ lesions in IgG4-related disease, which is extremely easily to be misdiagnosed as malignant nasal tumor with multiple organ metastases.

Case Presentation

A 63-year-old man was admitted to our department of The First Affiliated Hospital, Sun Yat-sen University. He had an 8-month history of impaired vision in right eye and headache, and 4 month later he underwent nasal obstruction, hoarseness and severe dysphagia which made him keep a liquid diet. His previous CT scans revealed a mass in the left nasal cavity (Figure 1A) and multiple nodules in bilateral lung (Figure 1B) and kidney (Figure 1C). Previous nasal endoscopy also showed a left-side nasal neoplasm (Figure 1D) and vocal cord paralysis (Figure 1E). The patient was referred to several different hospitals and the nasal mass was mostly considered to be a nasal tumor with multiple metastases. However, percutaneous renal biopsies were performed and no malignant lesion was detected. The patient further underwent a nasal endoscopic surgery for antrostomy on left sinus and resection of left-side nasal neoplasm in Yangjiang Central Hospital, China. Surprisingly, the postoperative pathology revealed the nasal polyp on the left nasal sinus and chronic mucosal inflammation on rhino-pharynx. However, the patient didn't have any relieves from the symptoms of headache and decreased vision after the surgery. Then the patient came to our department for further diagnosis and treatment. The patients received both the computed tomography and magnetic resonance imaging in our hospital and they indicated diffuse inflammation in the pterygoid muscle, optic sheath, cerebellopontine area and cavernous sinus region.

From the above findings of his history and examinations, autoimmune diseases especially the IgG4 related disease were highly considered. Thus, further biopsy was performed from the nasal mass. The immunohistochemical examination indicated that there were many IgG (Figure 1F) and IgG4 (Figure 1G) positive cells in the nasal mass and the ratio of IgG4 and total IgG positive cells

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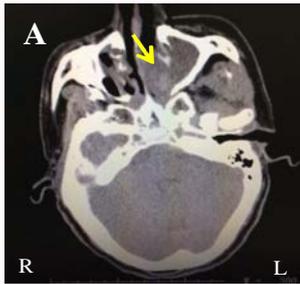


Figure 1: Pictures from a patient of IgG4-related disease.
A: CT images of nasal mass (yellow arrow).

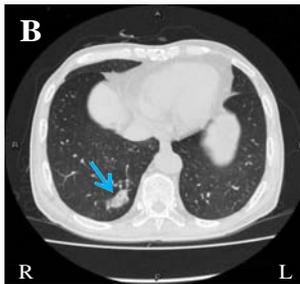


Figure 1B: Multiple pulmonary nodules (blue arrow).



Figure 1C: Multiple renal nodules (green arrows).



Figure 1D: The nasal endoscopic finding for the nasal mass (red arrow).

is more than 40% (Figure 1F-1G). Moreover, serum IgG4 was also measured and was found to increase to 21.10 g/L (The level of serum IgG4 more than 1.35 g/L is considered to be positive). The patient was finally diagnosed as IgG4-related disease after excluding some other possible diseases such as the lymphoma and nasopharyngeal carcinoma. After the systemic administration of steroid, the patient had a quick recovery.

Discussion

The IgG4-related disease has been described as a novel clinical entity [2] which is defined by diffuse or localized swelling or masses

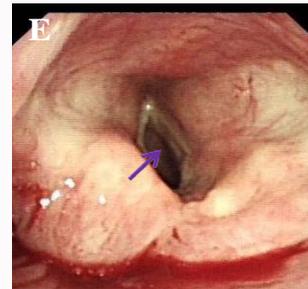


Figure 1E: The paralysis of the left vocal cord (purple arrow) can also be found under the endoscopy.

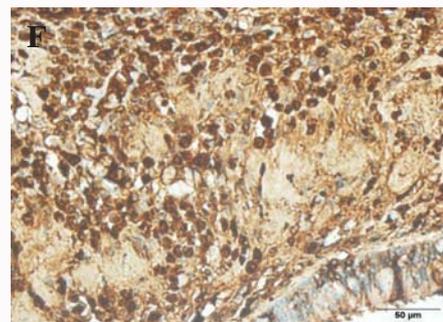


Figure 1F: The immunohistochemistry for the nasal tissue have shown the IgG-positive cell infiltration (400X).

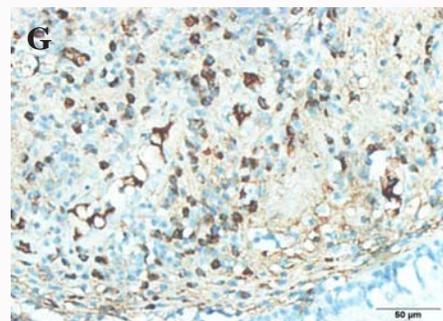


Figure 1G: IgG4-positive plasmacytes (400X) in the nasal tissue.

in single or multiple organs, high serum IgG4 level and infiltration of IgG4-positive plasmacytes in the target tissues [9]. The patient in our case was diagnosed definitely as IgG4-related disease based on the diagnostic criteria. Except for the nasal mass and pulmonary and renal nodules, the inflammation in the skull base and the complained of hoarseness and severe dysphagia should also be parts of the manifestations of IgG4-related disease.

In our case, the patient was found to have a nasal mass accompanied by multiple nodules in lung and kidney, which was highly suspected of malignant nasal carcinoma with multiple organ metastases. Though para-nasal lesion in IgG4-related disease that mimicking nasopharyngeal carcinoma has been reported since the year of 2010 [6,8], our case was the first one that exhibited the nasal mass accompanied by multiple organ nodules in IgG4-related disease, making it much more confusing to diagnose the disease simply based on the chief complaints and the auxiliary examinations if the serum level of IgG4 and immunochemistry examination for IgG4 in the nasal mass were not performed.

Before the final diagnosis of IgG4-related disease in our hospital,

the patient already underwent an endoscopic surgery for the resection of nasal mass in local hospital but has no any relieves from his complaints. However, he quickly recovered from the disease after the steroid was administrated systemically. It should be noted that the nasal mass in IgG4-related disease could become small with the treatment of steroid and an endoscopic surgery should be avoided. Therefore, we should strengthen our understanding and awareness for the diagnosis of the IgG4-related disease in nasal disease.

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