Case Report

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대한영상의학회지

Immunoglobulin G4-Related Disease Involving the Pterygopalatine Fossa, Mimicking Invasive Aspergillosis: A Case Report and Literature Review

침습아스페르길루스증으로 오인된 날개입천장오목에 발생한 면역글로불린 G4 관련 질환: 증례 보고 및 문헌 고찰

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We report a case of Immunoglobulin G4 (IgG4) related disease involving the pterygoplataine fossa. A 83-year-old male presented with left ocular pain and visual disturbance. CT showed an isodense soft tissue lesion in the left pterygopalatine fossa with bony sclerotic changes and erosion. MRI revealed an infiltrative soft tissue mass in the left pterygopalatine fossa as a T2 slightly low signal intensity and heterogeneous enhancement. The patient underwent left ethmoidectomy, and biopsy of the mass was conducted. The histopathological diagnosis was IgG4-related disease. In this case, it was difficult to differentiate invasive aspergillosis, which is common in immunocompromised patients, considering the patient's clinical history of diabetes mellitus. This report describes the imaging findings of IgG4-related disease mimicking invasive sinusitis such as invasive aspergillosis.

Index terms Immunoglobulin G4-Related Disease; Pterygopalatine Fossa; Immunocompetence

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INTRODUCTION

Immunoglobulin G4 (IgG4)-related disease is an autoimmune condition in which

IgG4 plasma cells infiltrate into tissues as a result of increased serum IgG4 levels. The predominantly involved organs are the pancreas, salivary gland, and biliary tract (1, 2). In the head and neck region, the salivary, lacrimal, and pituitary gland are frequently affected. It is very rare for IgG4-related disease to invade the nasal cavity or paranasal sinus. This case study aims to describe the imaging features of IgG4-related disease primarily involving the pterygopalatine fossa.

CASE REPORT

An 83-year-old male came to our clinic with left ocular pain and visual disturbance for a month. The patient had suffered from Type II diabetes mellitus and hypertension. He had no fever. White blood cell and eosinophil counts were in normal range. The neurological examination revealed mild limitation in the left lateral gaze. Nasal endoscopy showed polypoid mass-like lesion at the left middle meatus without mucopus. Based on clinical history of diaetes mellitus, sino-orbital infection such as invasive aspergillosis was suspected rather than inflammatory or tumorous condition.

On facial CT, isodense soft tissue mass was observed in the left pterygopalatine fossa with sclerotic change and erosion of the adjacent bone. There was accompanying soft tissue infiltration along the left optic nerve and extraocular muscles, suggesting orbital infiltrations. Mucoperiosteal thickening of the left maxillary, ethmoid, and sphenoid sinuses were also observed (Fig. 1A, B). We first suspected invasive sinusitis such as invasive aspergillosis, involving pterygopalatine fossa and other differential diagnosis are inflammatory pseudotumor (IPT) and lymphoma. MRI was performed to evaluate the characteristics and extent of this lesion. The mass-like lesion of the left pterygopalatine fossa presented heterogenous low signal intensity on T2-weighted images (T2WI) (Fig. 1C) and heterogeneous enhancement on contrast enhanced T1-weighted images (T1WI). It showed infiltrating nature and extended to the left cavernous sinus, left orbital muscles, and left masticator space (Fig. 1D). A diffuse enhancement along the left optic nerve and thickening of extraocular muscles (medial, inferior and lateral rectus muscles) were accompanied. There was no evidence of involvement in lacrimal gland and salivary gland.

For diagnosis, we performed endoscopic sinus surgery. Left ethmoidectomy and biopsies of the sinonasal mass at nasal cavity were conducted. During the surgery, we noted an irregularly shaped pale brown-colored mass filling the left posterior ethmoid sinus. This mass was eroding the medial wall of the orbit. The part of the mass was removed. On hematoxylin-eosin staining, the sinonasal mass showed dense lymphoplasmacytic infiltration with fibrosis. Immunostaining for IgG4 showed the majority of plasma cells positive for IgG4 (Fig. 1E). Serum IgG4 concentration was checked after the surgery, showing upper limit of a normal range (85.7 mg/dL, reference range 3.9–86.4). The findings above met the 2019 American College of Radiology/European League Against Rheumatism classification criteria for IgG4-related disease (1). After surgery, the patient was treated with prednisolone.

Fig. 1. Imaging findings in an 83-year-old male with IgG4-related disease involving pterygoplatine fossa.
A, B. Axial images show an isointense soft tissue lesion in the left pterygopalatine fossa with sclerotic change and erosion of the adjacent bone (arrow, left image). Infiltration into the left orbital apex (arrow, right image) was accompanied (A). An isodense soft tissue lesion in the left pterygopalatine fossa infiltrates through the inferior orbital fissure (black arrows) into the orbital apex and extends to the maxillary sinus with bony erosion (white arrow) on sagittal image (B).
C, D. MRI. T2-weighted axial image (C) shows focal low signal intensity area (arrow) within mixed iso- to hypointense mass-like lesion occupying the left pterygopalatine fossa. Contrast enhanced T1-weighted images (D) reveal enhancement at the left pterygopalatine fossa infiltrating into the left cavernous sinus, left orbital muscles (black arrows), and left masticator space through the pterygomaxillary fissure (dashed arrow).
IgG4 = immunoglobulin G4

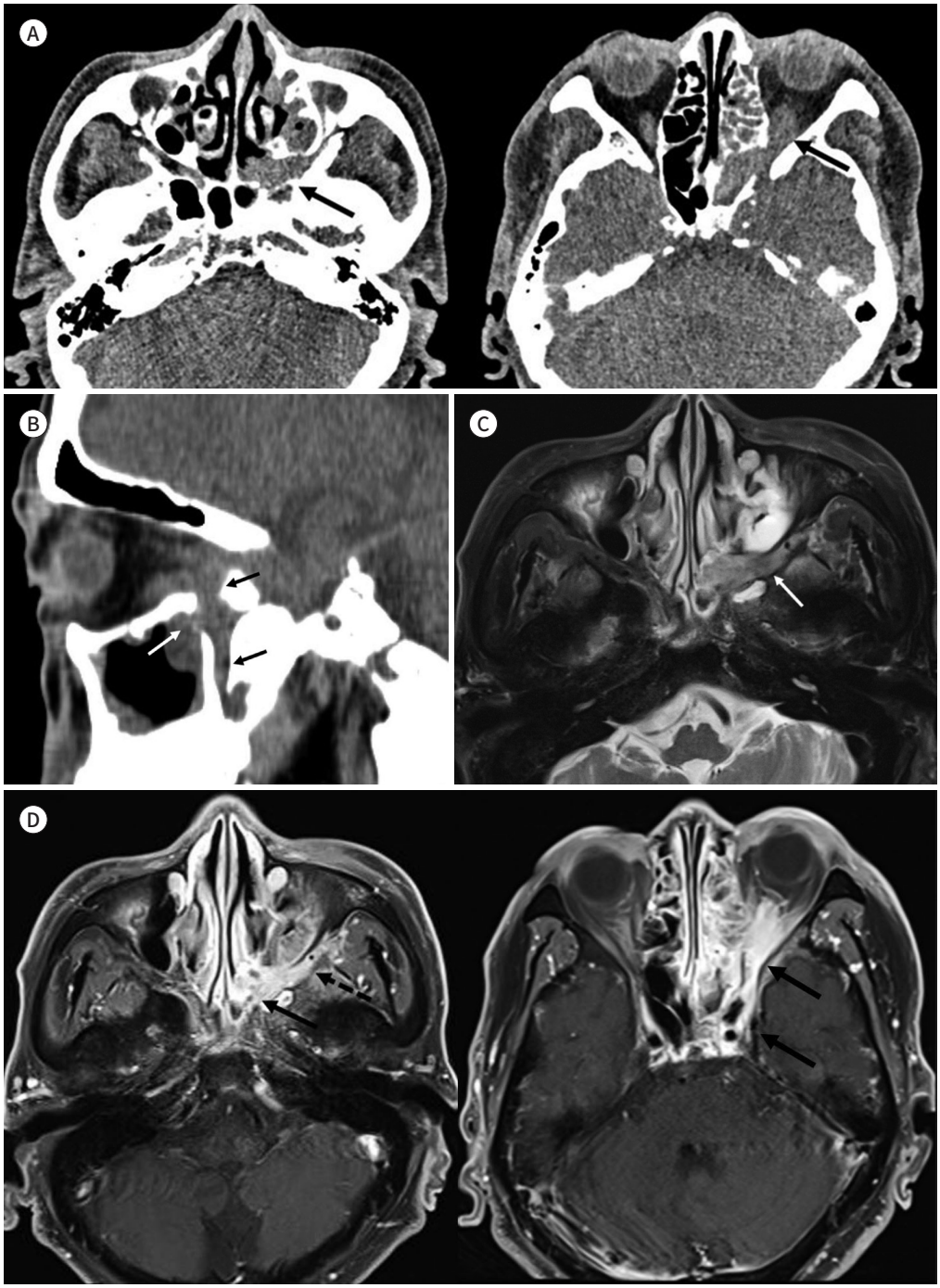
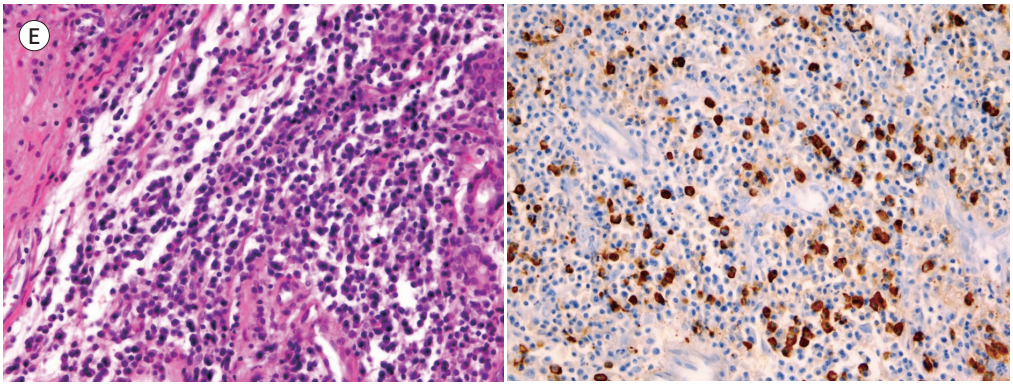


Fig. 1. Imaging findings in an 83-year-old male with IgG4-related disease involving pterygoplatine fossa.
E. Photomicrograph images of a sinonasal mass. Hematoxylin-eosin staining (× 400) shows dense lymphoplasmacytic infiltration with fibrosis (left). Immunostaining for IgG4 (× 400) reveals that the majority of plasma cells appear positive for IgG4 (right). Pathologic diagnosis was IgG4-related disease.
IgG4 = immunoglobulin G4



DISCUSSION

IgG4-related disease is an autoimmune condition in which IgG4 plasma cells infiltrate into tissues with increased serum IgG4 levels, causing tissue fibrosis and resulting dysfunction of the organ. This disease entity recently has been noticed and reported (2, 3). Clinical symptoms vary depending on the involved organ. Symptoms presented when sinonasal space or orbit involved with ocular pain, visual disturbance, headache, and chronic rhinitis, as noted in our case.

There are two different terminology "IPT" and "IgG4-related disease" commonly used in clinical field, sharing the pathologic feature of fibrosis and infiltration of lymphocytes and plasma cells. Although the relationship between IPT and IgG4-related disease has no definitely been established, recently IgG4-related disease is thought to form part of the spectrum of IPT according to the IgG4 level (4).

There are some differentials that should be considered when there is a lesion with T2 low signal intensity in the pterygopalatine fossa, such as lymphoma, invasive aspergillosis, Wegener's granulomatosis, and solitary fibrous tumor. Lymphoma shows soft tissue lesion with homogeneity and tends to remodel bone rather than lytic destruction which is different from our case (5, 6). Considering the clinical presentation of immune compromised state with DM, invasive aspergillosis was on the top of clinicians' differential diagnosis. But, laboratory findings favored non-infectious disease.

IgG4-related disease is known to show intermediate attenuation on CT and relatively low signal intensity on T1 and T2 weighted MR images (2). But usually, imaging findings of IgG4-disease seem quite non-specific and it is difficult to differentiate from other diseases by imaging. However, this case may reveal images of IgG4 related disease, slightly different from the typical imaging findings of invasive aspergillosis, which may facilitate in differentiating. On MRI, this case showed as T2 iso to hypointense soft tissue mass. Conversely, the typical MR findings of invasive aspergillosis are extremely low signal intensity in T2WI images with bright homogenous enhancement in post-contrast T1WI (7-9). Additionally, in this case, or-

bital involvement was overt and appeared extensively, but the presented clinical symptom was quite mild and normal laboratory findings compared with the extent involved. This may be another differential point from invasive aspergillosis involving orbit which may accompany more aggressive clinical symptom such as sudden vision loss.

In conclusion, it is notable that when T2 slightly low signal intensity mass in pterygopalatine fossa shows infiltrating enhancement feature, radiologists should also be alert to the possibility that IgG4-related disease can mimic invasive aspergillosis.

Author Contributions

Conceptualization, all authors; data curation, S.J.Y., K.J.Y.; investigation, S.J.Y., K.J.Y.; project administration, K.J.Y.; resources, all authors; supervision, K.J.Y.; visualization, all authors; writing—original draft, S.J.Y.; and writing—review & editing, S.J.Y., K.J.Y.

Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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REFERENCES

1. Wallace ZS, Naden RP, Chari S, Choi HK, Della-Torre E, Dicaire JF, et al. The 2019 American College of Rheumatology/European League Against Rheumatism classification criteria for IgG4-related disease. *Ann Rheum Dis* 2020;79:77-87
2. Fujita A, Sakai O, Chapman MN, Sugimoto H. IgG4-related disease of the head and neck: CT and MR imaging manifestations. *Radiographics* 2012;32:1945-1958
3. Horger M, Lamprecht HG, Bares R, Spira D, Schmalzing M, Claussen CD, et al. Systemic IgG4-related sclerosing disease: spectrum of imaging findings and differential diagnosis. *AJR Am J Roentgenol* 2012;199:W276-W282
4. Ryu G, Cho HJ, Lee KE, Lee JJ, Hong SD, Kim HY, et al. Clinical significance of IgG4 in sinonasal and skull base inflammatory pseudotumor. *Eur Arch Otorhinolaryngol* 2019;276:2465-2473
5. Koeller KK. Radiologic features of sinonasal tumors. *Head Neck Pathol* 2016;10:1-12
6. Kim SH, Mun SJ, Kim HJ, Kim SL, Kim SD, Cho KS. Differential diagnosis of sinonasal lymphoma and squamous cell carcinoma on CT, MRI, and PET/CT. *Otolaryngol Head Neck Surg* 2018;159:494-500
7. Kim E, Kim JY, Cho JH, Kang C. Image features of aspergillosis arising from the pterygopalatine fossa: a case report and literature review. *Oral Radiology* 2014;30:192-195
8. Katsura M, Mori H, Kunimatsu A, Sasaki H, Abe O, Machida T, et al. Radiological features of IgG4-related disease in the head, neck, and brain. *Neuroradiology* 2012;54:873-882
9. Wang Z, Feng R, Chen Y, Duan M, Wang M, Jin Z, et al. CT features and pathologic characteristics of IgG4-related systemic disease of submandibular gland. *Int J Clin Exp Pathol* 2015;8:16111-16116

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침습아스페르길루스증으로 오인된 날개입천장오목에 발생한 면역글로불린 G4 관련 질환: 증례 보고 및 문헌 고찰

손진영[1] · 김지영[2*] · 조진희[3] · 이은정[4]

본 증례는 좌안의 통증과 시력저하를 호소한 83세 남자 환자에게서 진단된 면역글로불린 G4 (immunoglobulin G4; 이하 IgG4) 관련 질환의 CT와 MRI 소견에 대한 보고이다. 이는 CT에서 주변의 골미란을 동반한 날개입천장 오목의 등밀도 연부조직 종괴로 관찰되었으며 이 병변은 자기공명영상 T2 강조영상에서 등신호에서 저신호강도를 보이며 비균질한 조영증강을 보였다. 환자는 벌집굴 절제술과 조직검사를 시행하였고 조직학적 검사상 IgG4 관련 질환으로 진단되었다. 환자는 기저질환으로 당뇨병이 있었고, 이런 면역저하 환자군에서 흔하게 보이는 침습적 진균성 부비동염과의 영상학적 감별이 어려운 경우였다. 저자들은 침습적 진균성 부비동염과 유사한 영상 소견을 보였던 IgG4 관련 질환의 증례를 문헌 고찰과 함께 보고하고자 한다.

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