



## Intracranial Inflammatory Pseudotumor with Left Internal Carotid Artery Stenosis due to Immunoglobulin G4-Related Disease: A Case Report and Literature Review

Chenyang Zhao<sup>1#</sup>, Wenyu Liu<sup>1#</sup>, Xueqin Chen<sup>2</sup>, Dong Zhou<sup>1\*</sup> and Xintong Wu<sup>1\*</sup>

<sup>1</sup>Department of Neurology, West China Hospital, Sichuan University, PR China

<sup>2</sup>Department of Pathology, West China Hospital, Sichuan University, PR China

<sup>#</sup>These authors contributed equally to this work and share the first authorship

### Abstract

**Background:** Immunoglobulin G4-Related Disease (IgG4-RD) is an autoimmune-mediated inflammatory fibrotic disease that can cause organ enlargement and tissue destruction. It can involve any organ but is most common in the pancreas, bile ducts, etc. An intracranial IgG4-associated inflammatory pseudotumor, as described in the case presented here, is unusual.

**Case Report:** A 27-year-old male was admitted to hospital due to visual loss in the left eye for 11 years and left-sided facial numbness for 1 year. A mass located in the left orbit, cavernous sinus, infratemporal fossa, and parapharyngeal space was revealed by Magnetic Resonance Imaging (MRI). The left interior carotid artery was encased by the lesion. Surgery was performed for biopsy. The pathological findings showed diffuse lymphoplasmacytic infiltration with lymphoid follicles and fibrous tissue hyperplasia. Immunostaining identified many IgG4+ cells, with up to 30/High Power Field (HPF). The IgG4 serum level was also high. The patient was diagnosed with IgG4-RD and given steroid therapy. Four months after surgery and the therapy, MRI results showed that the lesion had been resolved.

**Conclusion:** Although this patient met the diagnostic criteria and had many similarities with previously reported cases of IgG4-RD, there were also many new manifestations, such as different imaging features, stenosis of the internal carotid artery, and so on. These findings suggest that our understanding of the disease is insufficient, and that future studies with a larger sample size and long-term observation follow-up are needed.

**Keywords:** IgG4-related disease; IgG4-related inflammatory pseudotumor; Pathology; Surgery

### Introduction

Immunoglobulin G4-Related Disease (IgG4-RD) is an autoimmune-mediated inflammatory fibrotic disease that can cause organ enlargement, tissue destruction, and even organ dysfunction. The annual incidence is 3.1 cases per 100,000 people and it occurs most often in older males [1]. In 2003, Kamisawa et al. [2,3] first proposed “IgG4-related systemic disease” as a new clinicopathological systemic entity. The pathology of this disease is characterized by lymphoplasmacytic infiltration that includes rich, IgG4-positive plasma cells organized in a storiform pattern of fibrosis [4] and obliterative phlebitis. IgG4-RD can involve any organ and is common in the pancreas, bile ducts, lungs, kidneys, etc. Several cases of IgG4-associated inflammatory pseudotumor have been reported, but intracranial IgG4-associated inflammatory pseudotumors are uncommon. Their clinical and radiological features are very similar to those of tumors, particularly meningioma. It is thus easily misdiagnosed and surgically removed while IgG4-related inflammatory pseudotumors are sensitive to steroid therapy. Therefore, we herein report this case and review previous cases in order to enhance our understanding of the disease and improve diagnostic accuracy.

### Case Presentation

#### Medical history

The patient was a 27-year-old male who was admitted to our hospital due to visual loss in the left eye for 11 years and left-sided facial numbness for 1 year. Eleven years ago, he suffered from decreased vision in the left eye with proptosis and pain. He went to the local hospital and after

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#### \*Correspondence:

Xintong Wu, Department of Neurology,  
West China Hospital, Sichuan  
University, Chengdu, PR China,  
E-mail: 36853839@qq.com

Dong Zhou, Department of Neurology,  
West China Hospital, Sichuan  
University, Chengdu, PR China,  
E-mail: zhoudong66@yahoo.de

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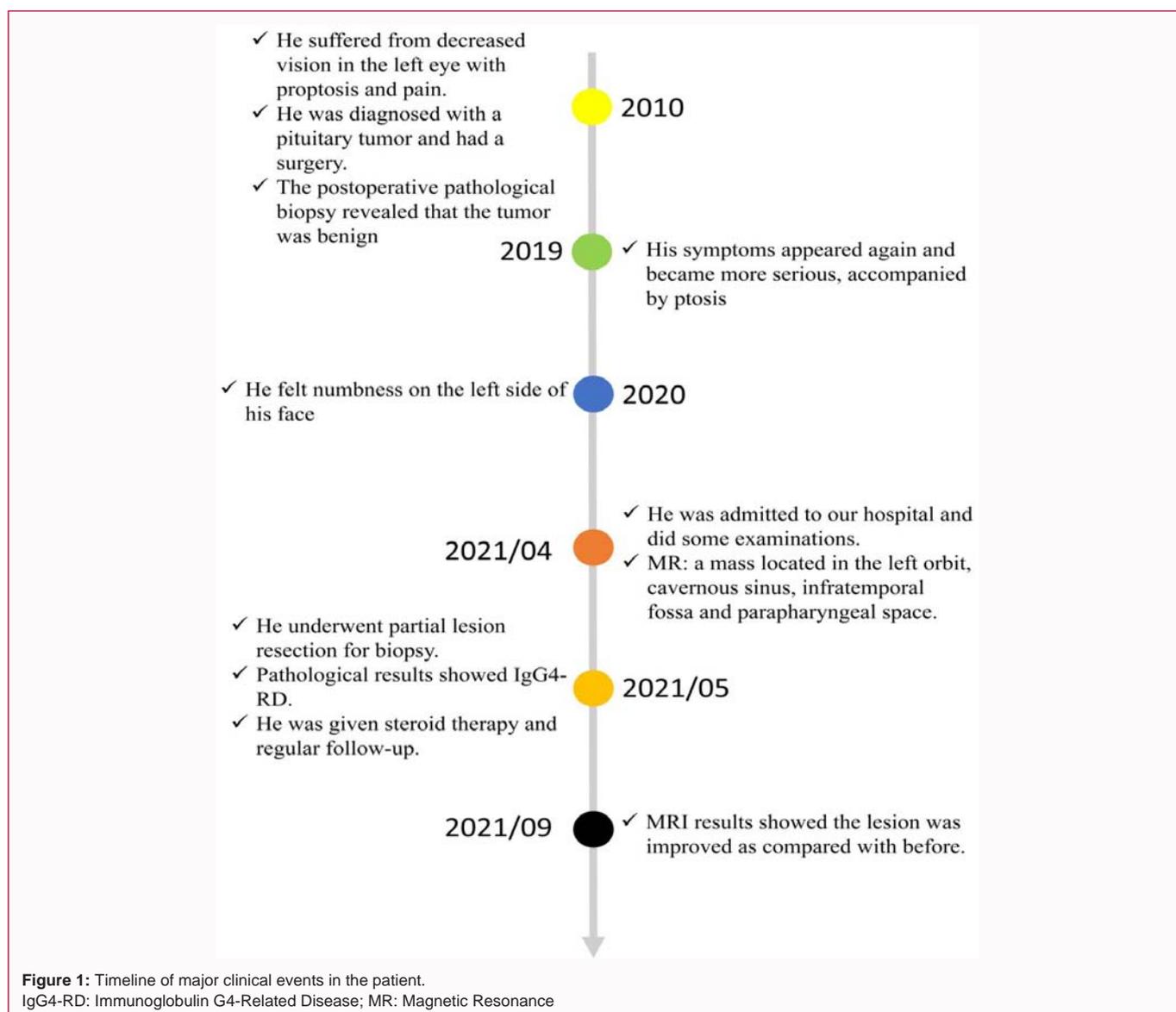
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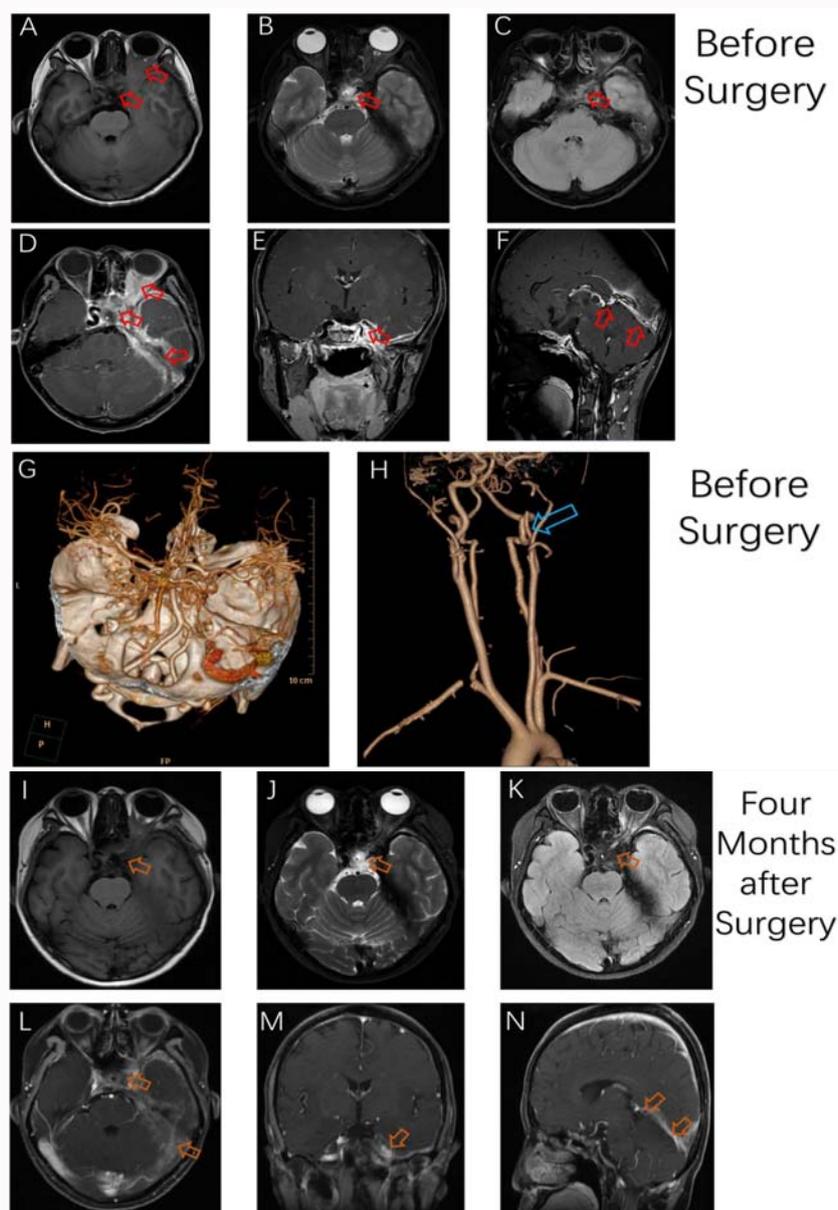
the examinations; he was diagnosed with a pituitary tumor. Gamma knife surgery through the sella turcica region was performed, however, results of the postoperative pathological biopsy revealed that the tumor was benign (details are not available). After discharge, the patient's symptoms were relieved. Nevertheless, 2 years ago, the patient's symptoms appeared again and became more serious, accompanied by ptosis. One year ago, he felt numbness on the left side of his face. He had no past history of autoimmune disease or other illness and no familial history of note (Figure 1).

Physical examination showed that the left pupil diameter was about 5 mm and the light reflection disappeared. The vision of his left eye decreased and the eyeball was fixed. The feeling of his left face and the force of the left masseter were weak. Magnetic Resonance Imaging (MRI) revealed a mass located in the left orbit, cavernous sinus, infratemporal fossa, and parapharyngeal space. It involved the optic nerve and the extraocular muscles. Combined with his surgical history, tumor, especially meningioma was highly suspected. Computed Tomography Angiography (CTA) revealed that the left anterior carotid artery was encased by the lesion, and had become narrow (Figure 2).

**Cerebrospinal fluid:** immunoglobulin 0.1280 g/L (0.005-0.041), protein 0.48 g/L (0.15-0.50). Serum: immunoglobulin 17.2 g/L (8.00-15.50), IgG4 1.990 g/L (0.035-1.500), prolactin 36.60 ng/mL (4.6-21.4), testosterone 0.747 ng/mL (2.50-9.08), Luteinizing Hormone (LH) 1.5 IU/L (1.7-8.6), Estradiol (E2) <5.0 pg/mL (11.3-43.2). Antinuclear Antibody (ANA) + (1:100 granular type). Other routine laboratory tests showed no abnormal data. Chest, abdominal, and pelvic CT findings were unremarkable.

### Surgical biopsy and pathological results

After a thorough whole-body evaluation, the patient underwent partial lesion resection craniotomy. During the operation, there was no obvious destruction of the skull, however, the dura mater of the left anterior middle skull base was thickened and partially destroyed. Its texture was tough and the blood supply was average. Then the lesion of the dura mater was removed and sent for biopsy. The neuropathological findings showed diffuse lymphoplasmacytic infiltration with lymphoid follicles and fibrous tissue hyperplasia. Immunostaining showed EMA (plasma cell +), SSTR2 (-), and many IgG4+ cells were identified, with up to 30/HPF (Figure 3).



**Figure 2:** Before surgery, brain MRI images showed intracranial mass encased the left cavernous sinus, invaded the left retrobulbar space anteriorly, and was indistinctly demarcated with the left optic nerve and extraocular muscles (as red arrows showed). To the left, the lesion invaded the left temporal and tentorium, and the meninges at the base of the posterior cranial fossa thickened. The mass extended down to the left infratemporal fossa and left parapharyngeal space, and the left internal jugular vein and sigmoid sinus were involved. The mass showed predominantly isointense signal on T1 (A) and hyperintense signal on T2-weighted image (B); and it was homogeneously enhanced after contrast MRI (D-F). CTA showed stenosis of the left internal carotid artery (G-H). Four months after surgery, compared with the previous MRI images, the meningeal thickening was slightly relieved, and the thickening and enhancement of the left optic nerve and extraocular muscles were less than before (I-N). MRI: Magnetic Resonance Image; CTA: Computed Tomography Angiography.

### Postoperative treatment and follow up

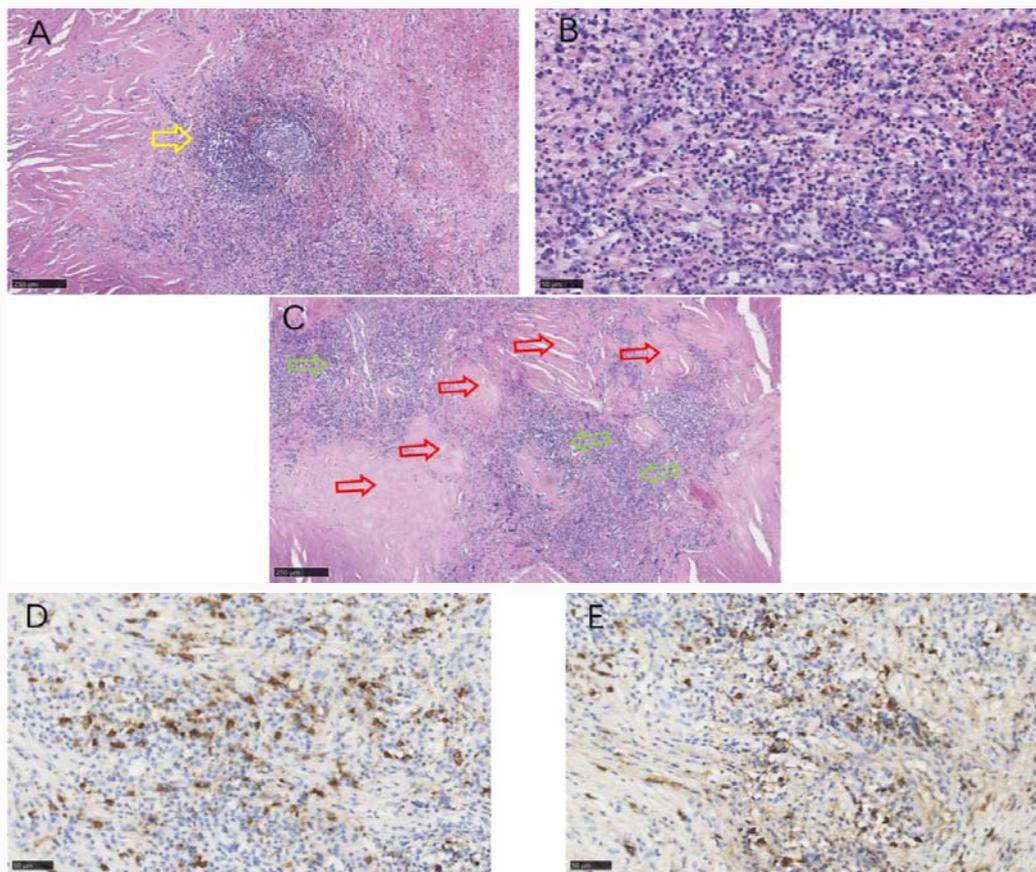
Finally, the patient was diagnosed with IgG4-RD based on the clinical manifestations, laboratory examinations, and pathological biopsy results. He was given steroid therapy and regular follow-up. Four months after treatment, MRI results showed the left cavernous sinus area, the thickened meninges, the involved optic nerve, and the extraocular muscles were all improved as compared with before (Figure 2).

### Discussion

Immunoglobulin G4-Related Disease (IgG4-RD) is an immune-mediated fibro-inflammatory condition characterized by organ

enlargement and elevated serum IgG4 levels. It can involve multiple organs, such as the pancreas, bile duct, liver, gastrointestinal tract, lacrimal gland, salivary gland, paranasal sinus, thyroid, artery, peritoneal tissue, lung, kidney, lymph nodes, etc. [1]. In intracranial lesions due to IgG4-RD, central nervous system involvement has been reported less frequently while hypophysitis and pachymeningitis are generally well known. In addition, intracranial pseudotumors are rare [5]. To our knowledge, 28 cases of intracranial pseudotumor due to IgG4-RD have been reported (Table 1).

In terms of diagnosis and treatment, Umehara et al. [6] proposed the first diagnostic criteria in 2011. Then, in 2015, an expert consensus on the diagnosis and treatment of IgG4-RD was published, and the



**Figure 3:** Pathological results showed diffuse lymphoplasmacytic infiltration with lymphoid follicles and fibrous tissue hyperplasia. A) Yellow arrow indicated lymphoid follicles. B) Lymphocyte and plasma cell infiltration. C) Red arrows represented fibrous tissue hyperplasia, and green arrows indicated lymphocyte and plasma cell infiltration.

article emphasized the importance of histopathological examination of IgG4-RD. Furthermore, glucocorticoid therapy is the first choice for initial treatment for all active patients with IgG4-RD. However, rituximab may be considered in patients with relapse or inability to tolerate glucocorticoid therapy [7]. The 2019 ACR/EULAR classification criteria for IgG4-RD consist of entry, exclusion, and inclusion criteria and are high sensitivity and specificity [8]. In 2021, Umehara et al. [9] published a revised version of the comprehensive diagnostic criteria. Despite these advances, the diagnosis of intracranial pseudotumor due to IgG4-RD can be difficult, regardless of which diagnostic criteria are chosen. This is in part because the description is not sufficient and the specificity is not high enough. Thus, patients with the disease are easily misdiagnosed with tumors or infections, and treated with unnecessary surgery. Therefore, the need for more relevant case reports is essential to aid in the further understanding of the clinical features, imaging, and prognosis of intracranial pseudotumor due to IgG4-RD.

In this work, we searched English and Chinese databases and included 28 cases of intracranial pseudotumor due to IgG4-RD in the study (Table 1). We observed some points when comparing our case with previous reported cases. First, according to a cross-sectional study conducted by Zen et al. [10], IgG4-related disease typically affects elderly males. The average age of the 28 patients we included was 52. A 96-year-old while our case is a 27-year-old male. This indicated that age is not a critical factor for diagnosis. Second, a neoplasm could be located anywhere in the brain, and could be

single or multiple. The corresponding clinical manifestations varied according to the location of the lesion. The common symptoms were headache, visual disturbance, limb weakness, facial numbness, and so on. In this case, the disease course was long and the scope of lesion involvement was extensive, from the anterior to the orbit, involving the optic nerve and extraocular muscles, and inferior to the parapharyngeal space. Based on previous reports, MR imaging of the lesion usually demonstrates isointense signal on T1-weighted imaging, isointense/hypointense signal on T2-weighted imaging, and is uniformly enhanced after contrast MRI, which is possibly explained by the combination of fibrosis and attenuated cellularity [11-20]. However, our case showed slightly hyperintense signal on T2-weighted imaging. The texture of the lesion, in this case, was hard, consistent with previous reports. Third, pseudotumors involved large vessels in 7 cases [14,18-21]. Most studies reported that IgG4-RD usually involved thickening around arteries without compression or occlusion [14,19,20,22,23]. Some researches thought this phenomenon could explain why inflammatory infiltrates with abundant plasma cells were seen around arteries in the affected regions [19,24-26]. But, in this patient, it was clear that the left internal carotid artery was compressed by the lesion throughout the whole process, and was obviously narrower than the contralateral side. There was also an obvious narrowing of the lumen of the C5 segment of the left internal carotid artery and the wall was rough. Fourth, the immunoglobulins and proteins were significantly elevated in the cerebrospinal fluid of this patient. In previous reports, only one case described cerebrospinal fluid with protein levels within the normal

**Table 1:** Reported cases of IgG4-RD with intracranial pseudotumor lesions.

Case	Report	Age, Sex	Symptoms	Lesion (MR Image)	Intracranial Multiple Lesions	Extracranial Lesions	Tumor Consistency	Encasing Artery	Serum IgG4 (mg/dl)	CSF	Histopathological Findings
1	Wong et al. [6]	77,M	Blurred vision	Pituitary tumor with suprasellar extension and optic nerve compression	N	Pancreas, gall bladder	ND	N	720	ND	Prominent plasmacytic infiltration with a sprinkle of lymphocytes and an area of sclerosis
2	Lui et al. [7]	52,F	Left homogenous superior quadrantanopia	Right lateral ventricle (Contrast enhancing in T1 -weighted magnetic resonance imaging)	N	N	ND	ND	ND	ND	Inflammatory cell-rich (lymphocytes and plasma cells) lesions with a small number of benign-looking spindle cells; Obliterative phlebitis; IgG4-positive plasma cells at a density of 36 to 47 positive cells per HPF
3		45,M	Progressive left-sided weakness	Right frontal dural-based tumor	N	N	ND	ND	ND	ND	
4		26, F	Severe headache, left eye discomfort with diplopia, left ear pain	Extensive enhancing dural thickening over left frontotemporal lobes with extension through the temporal bone into left orbit and infratemporal fossa	N	N	ND	ND	ND	ND	
5	Lindstrom et al. [8]	53,M	Chronic lower neck headaches	Posterior fossa tumor	N	N	ND	ND	ND ANA +(speckled; 1:80)	ND	Lymphoplasmacytic inflammation, fibrosis, and phlebitis; IgG4+ cells/HPF 26.8
6	Kim et al. [9]	43,M	Headache and progressive motor weakness in the right upper extremity	Irregular-shaped extra-axial mass in the left frontal area near the corpus callosum and cingulate gyrus(Heterogeneous enhancement in T1-weighted MR images obtained with Gd contrast)	Y	N	Hard	ND	ND	ND	Thickened fibrous tissue with marked inflammatory cell (mature lymphocytes and plasma cells) infiltration; IgG4+ cells/HPF 72.4
7	Katsura et al. [10]	59,F	Left-sided facial numbness	Homogeneously enhancing soft-tissue mass involving the skull base along the second and third divisions of the left trigeminal nerve (T2-weighted image demonstrates a hypointense mass)	N	N	ND	ND	ND	ND	nerve fibers surrounded Inflammatory infiltration comprising predominantly B and T lymphocytes with moderate fibrosis; IgG4+ cells/HPF 128, IgG4+/IgG+ plasma cells (71%)
8		61,F	Facial paresthesia	Cavernous sinus to the pterygopalatine fossa, infraorbital canal, and masticator space (Hypointense T2-weighted images; homogeneous enhancement)	N	N	ND	ND	145	ND	Diffuse lymphoplasmacyte and high IgG4+ plasma cell infiltration (>50 IgG4+ cells per high-power field and IgG4+/IgG+ cell ratio >30%) with fibrosis or sclerosis
9		64,M	Diplopia	Orbital apex and to the cavernous sinus (Iso- to hypointense T2-weighted images)	N	Lacrimal gland, lung and thoracic paravertebral area	ND	ND	3280	ND	
10	Katsura et al. [11]	55,M	Facial paresthesia	Cavernous sinus to orbit, pterygopalatine fossa, and masticator space (Hypointense T2-weighted images; homogeneous enhancement)	N	N	ND	ND	ND	ND	
11		65,M	Facial paresthesia	Cavernous sinus to the pterygopalatine fossa, intracranial dura mater(Hypointense T2-weighted images; homogeneous enhancement)	N	Lacrimal glands and thoracic paravertebral area	ND	ND	121	ND	
12		62,M	Superior nasal quadrant anopsia	Anterior clinoid process and posterior fossa dura mater (Hypointense T2-weighted images; homogeneous, gradual enhancement)	N	Pancreas	ND	Vertebral artery penetrating the mass	405	ND	
13	Moss et al. [12]	36,F	Headaches, double vision worse in left gaze, and numbness of the left forehead and cheek	Extra-axial mass in the left middle cranial fossa involving the left cavernous sinus	Y	N	ND	ND	ND	ND	Fragments of leptomeninges and dura involved by dense chronic inflammatory infiltrate composed predominantly of plasma cells with abundant macrophages and lymphocytes; IgG4+ cells per high-power field 43 and IgG4+/IgG+ cell ratio 20%

14	Moss et al. [12]	50,F	Headaches, double vision worse in the left gaze, and visual loss in the left eye	Left petrous bone extending into the left cavernous sinus, orbital apex, middle cranial fossa, the floor of the anterior cranial fossa, cerebellopontine angle, tentorium, and superior nasopharynx	N	N	ND	ND	ND	12 WBC/ $\mu$ L (95% monocytes), 2 RBC/ $\mu$ L, 45 mg/dL protein and 40 mg/dL glucose. CSF cytology did not reveal malignant cells	Lymphoplasmacytic infiltration in a background of dense fibrosis or sclerosis; over 50 IgG4 positive plasma cells per high power field and an IgG4/IgG ratio of 80%
15	Nishino et al. [13]	67,M	Headache, occasional left-sided facial numbness, and enlargement of the lachrymal and parotid glands	Intracranial, extra-axial, dural-based mass lesions in the bilateral Sylvian fissures, bilateral trigeminal nerves, and pituitary stalk	Y	Lachrymal glands, parotid glands, kidney	ND	Bilateral internal carotid arteries (arteritis)	3410	ND	Infiltration of lymphocytes and plasma cells with high ratios of IgG4+/IgG1+ plasma cells (>1)
16	Okano et al. [14]	62,M	Visual disturbance and quadrantanopia of the right eye	Right paraclinoid, right Meckel's cave, left foramen magnum (Homogeneously enhanced on contrast-enhanced T1W)	Y	Pancreas, submandibular gland, kidney	Hard	ND	0.000405	ND	Predominantly infiltration by inflammatory cells, such as lymphocytes, plasmacytes, and eosinophils; numerous IgG4-positive plasma cells that had infiltrated into the lesions
17	Noshiro et al. [15]	39,M	Right blurred vision and proptosis	Right optic nerve, retrobulbar space, cavernous sinus, bilateral maxillary nerves (Isointense on T1-weighted imaging, hypointense on T2-weighted imaging, and homogeneously enhanced on contrast-enhanced T1W)	N	Paravertebral tissue; Kidney	ND	ND	88300	ND	Plasmacyte and lymphocyte infiltration and dense fibrosis, which encased peripheral nerves; IgG4+/IgG+ cell ratio >40%
18	Rice et al. [16]	46,M	Fatigue, weight loss (25 kg), headache, right-sided facial numbness, and diplopia on gaze to the right	Carotid canal, jugular foramen, and foramen ovale	N	N	ND	ND	ND	ND	Diffuse plasma cell rich, chronic inflammatory cell infiltrate; prominent stromal fibrosis/hyalinization, fat necrosis with focal granulation tissue, and numerous IgG and IgG4-positive plasma cells
19	Tang et al. [17]	50,M	Abducens paralysis of the right eye.	Right upper clivus area (Isointense signal on T1-weighted MRI isointense/hypointense signal on T2-weighted MRI; uniformly enhanced after contrast MRI)	N	N	soft	ND	13.1	ND	Fibroinflammatory lesion with strands of sclerosing fibrosis mixed with lymphocytes and plasma cells; numerous IgG4-positive plasma cells
20	Goulam-Houssein et al. [18]	70,M	Diplopia, seizures	Frontal- temporal lesion (Isointense signal on T1-weighted MRI; homogeneous enhancement)	N	N	ND	ND	152	ND	Inflammatory cell infiltration of lymphocytes, plasma cells, and eosinophils forming nodules, on a background of a sclerotic fibrocollagenous stroma; obliterative phlebitis; IgG4/IgG ratio 91%
21		54,M	Headaches, left eye ptosis, vision loss	Cavernous sinus and Meckel's cave (Fat-suppressed T2WI: diffuse hypointense; post-gadolinium T1WI: avid diffuse enhancement)	Y	N	ND	Smooth narrowing cavernous ICA	92.7	The elevated protein of 1.55 g/L	IgG-staining plasma cells in the dura
22		28,F	Headaches and sudden loss of vision in the left eye and painful eye movements	Suprasellar mass (post-gadolinium sagittal T1WI: enhancing)	N	N	ND	Occlusion of ICA	88	ND	Lymphoplasmacytic infiltration with IgG4-positive plasma cells throughout
23	Kuroda, et al. [19]	76,F	N	Around the medulla, cerebellum, and middle fossa (Isointensity on T1- and T2-weighted imaging; post contrast MRI showed diffuse enhanced nodules)	Y	N	Hard	Encasing VA butis not occluded or stenosed	330	ND	Diffuse lymphoplasmacytic infiltration with lymphoid follicles; IgG4+ cells >400 per high-power field and an IgG4/IgG cell ratio >80%
24	Liu et al. [20]	50,F	Headache, dysphagia, and dysphonia	Middle upper clivus area (Isointense signal in T1-weighted MRI image; hypointense signal in T2-weighted MRI image; uniformly enhanced after contrast MRI scan)	N	N	Hard	Attaching to basal artery	ND	ND	Composed of sclerosing fibrosis associated with dense lymphoplasmacytic infiltration; over IgG4-positive cells per high-powered field and a high IgG4/IgG ratio

25		58,M	Abdominal pain and distension, diarrhea	Sellar space	N	Pancreas	ND	ND	225	ND	Large number of lymphocytes and plasma cells infiltration
26		40,M	Polydipsia, polyuria, weight loss, lumbago pain	Pituitary, bilateral cavernous sinus	N	Pancreas, kidney, ureter, lymph node	ND	ND	2510	ND	IgG4+ cells per high-power field >50 and IgG4+/IgG+ cell ratio 80%
27	Meng et al. [21]	43,M	Thirsty, polydipsia, polyuria, eyelid swelling	Circular nodules in the pituitary	N	Lacrimal gland, lymph node	ND	ND	3270	ND	IgG4+ cells per high-power field >50 and IgG4+/IgG+ cell ratio 40%
28		52,M	Eyelid swelling, seizure, eye pain, vision loss, buccal and submandibular swelling	Dura mater, right cavernous sinus occupied	N	Lacrimal gland, submandibular gland, parotid gland	ND	ND	6100	ND	IgG4+ cells per high-power field 30 and IgG4+/IgG+ cell ratio 40%
29	Present case	27,M	Visual loss, facial numbness	Dura mater, left orbits, cavernous sinus, infratemporal fossa, and parapharyngeal space	N	N	Hard	ICA stenosis	1720	Immunoglobulin 0.1280 g/L, protein 0.48 g/L.	Diffuse lymphoplasmacytic infiltration with lymphoid follicles and fibrous tissue hyperplasia. IgG4+ cells up to 30 /HPF

N: None; ND: Not Described; M: Male; F: Female; ICA: Internal Carotid Artery; VA: Vertebral Artery; MRI: Magnetic Resonance Imaging; CSF: Cerebrospinal Fluid; ANA: Antinuclear Antibody; WBC: White Blood Cell; RBC: Red Blood Cell; T1WI: T1-Weighted Image; T2WI: T2-Weighted Image

range [22]. Whether elevated immunoglobulin in cerebrospinal fluid of patients with IgG4-RD intracranial pseudotumor has a diagnostic value as in IgG4-related hypertrophic pachymeningitis [27] requires further study. In addition, our patient showed ANA+ (1:100 granular type), and one patient of previous reports also showed ANA + (speckled; 1:80), which was not mentioned in other reports. Fifth, in all 28 cases thus far, 11 cases had extracranial organ involvement, suggesting that, when intracranial pseudotumor is highly suspected to be related to IgG4-RD, particular attention should be paid to the screening of other organs such as the pancreas, lung, kidney, etc [14-16,21,28-31]. Importantly, if uncontrolled lesions remain, it may lead to irreversible damage to certain organs, such as aortitis, retroperitoneal fibrosis sclerosing cholangitis, pachymeningitis, and pericarditis.

For the treatment of IgG4-RD, it is currently believed that steroids are the first-line choice. The response to steroid therapy can vary depending on the affected organ and the severity of fibrosis. In this case, it could be seen that the patient achieved tumor reduction when compared with the previous imaging.

## Conclusion

This patient met the diagnostic criteria and was thus diagnosed with IgG4-RD. This case has many similarities with those previously reported; however, there were also many new manifestations, which suggest that our understanding of the disease is not enough. In the future, more studies with a larger sample size and long-term observation and follow-up will help improve our comprehension of this disease and improve patient outcomes.

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