

World Journal of *Clinical Cases*

World J Clin Cases 2022 June 26; 10(18): 5934-6340



MINIREVIEWS

- 5934 Development of clustered regularly interspaced short palindromic repeats/CRISPR-associated technology for potential clinical applications
Huang YY, Zhang XY, Zhu P, Ji L
- 5946 Strategies and challenges in treatment of varicose veins and venous insufficiency
Gao RD, Qian SY, Wang HH, Liu YS, Ren SY
- 5957 Diabetes mellitus susceptibility with varied diseased phenotypes and its comparison with phenome interactome networks
Rout M, Kour B, Vuree S, Lulu SS, Medicherla KM, Suravajhala P

ORIGINAL ARTICLE**Clinical and Translational Research**

- 5965 Identification of potential key molecules and signaling pathways for psoriasis based on weighted gene co-expression network analysis
Shu X, Chen XX, Kang XD, Ran M, Wang YL, Zhao ZK, Li CX
- 5984 Construction and validation of a novel prediction system for detection of overall survival in lung cancer patients
Zhong C, Liang Y, Wang Q, Tan HW, Liang Y

Case Control Study

- 6001 Effectiveness and postoperative rehabilitation of one-stage combined anterior-posterior surgery for severe thoracolumbar fractures with spinal cord injury
Zhang B, Wang JC, Jiang YZ, Song QP, An Y

Retrospective Study

- 6009 Prostate sclerosing adenopathy: A clinicopathological and immunohistochemical study of twelve patients
Feng RL, Tao YP, Tan ZY, Fu S, Wang HF
- 6021 Value of magnetic resonance diffusion combined with perfusion imaging techniques for diagnosing potentially malignant breast lesions
Zhang H, Zhang XY, Wang Y
- 6032 Scar-centered dilation in the treatment of large keloids
Wu M, Gu JY, Duan R, Wei BX, Xie F
- 6039 Application of a novel computer-assisted surgery system in percutaneous nephrolithotomy: A controlled study
Qin F, Sun YF, Wang XN, Li B, Zhang ZL, Zhang MX, Xie F, Liu SH, Wang ZJ, Cao YC, Jiao W

- 6050** Influences of etiology and endoscopic appearance on the long-term outcomes of gastric antral vascular ectasia

Kwon HJ, Lee SH, Cho JH

Randomized Controlled Trial

- 6060** Evaluation of the clinical efficacy and safety of TST33 mega hemorrhoidectomy for severe prolapsed hemorrhoids

Tao L, Wei J, Ding XF, Ji LJ

- 6069** Sequential chemotherapy and icotinib as first-line treatment for advanced epidermal growth factor receptor-mutated non-small cell lung cancer

Sun SJ, Han JD, Liu W, Wu ZY, Zhao X, Yan X, Jiao SC, Fang J

Randomized Clinical Trial

- 6082** Impact of preoperative carbohydrate loading on gastric volume in patients with type 2 diabetes

Lin XQ, Chen YR, Chen X, Cai YP, Lin JX, Xu DM, Zheng XC

META-ANALYSIS

- 6091** Efficacy and safety of adalimumab in comparison to infliximab for Crohn's disease: A systematic review and meta-analysis

Yang HH, Huang Y, Zhou XC, Wang RN

CASE REPORT

- 6105** Successful treatment of acute relapse of chronic eosinophilic pneumonia with benralizumab and without corticosteroids: A case report

Izhakian S, Pertzov B, Rosengarten D, Kramer MR

- 6110** Pembrolizumab-induced Stevens-Johnson syndrome in advanced squamous cell carcinoma of the lung: A case report and review of literature

Wu JY, Kang K, Yi J, Yang B

- 6119** Hepatic epithelioid hemangioendothelioma after thirteen years' follow-up: A case report and review of literature

Mo WF, Tong YL

- 6128** Effectiveness and safety of ultrasound-guided intramuscular lauromacrogol injection combined with hysteroscopy in cervical pregnancy treatment: A case report

Ye JP, Gao Y, Lu LW, Ye YJ

- 6136** Carcinoma located in a right-sided sigmoid colon: A case report

Lyu LJ, Yao WW

- 6141** Subcutaneous infection caused by *Mycobacterium abscessus* following cosmetic injections of botulinum toxin: A case report

Deng L, Luo YZ, Liu F, Yu XH

- 6148** Overlapping syndrome of recurrent anti-N-methyl-D-aspartate receptor encephalitis and anti-myelin oligodendrocyte glycoprotein demyelinating diseases: A case report
Yin XJ, Zhang LF, Bao LH, Feng ZC, Chen JH, Li BX, Zhang J
- 6156** Liver transplantation for late-onset ornithine transcarbamylase deficiency: A case report
Fu XH, Hu YH, Liao JX, Chen L, Hu ZQ, Wen JL, Chen SL
- 6163** Disseminated strongyloidiasis in a patient with rheumatoid arthritis: A case report
Zheng JH, Xue LY
- 6168** CYP27A1 mutation in a case of cerebrotendinous xanthomatosis: A case report
Li ZR, Zhou YL, Jin Q, Xie YY, Meng HM
- 6175** Postoperative multiple metastasis of clear cell sarcoma-like tumor of the gastrointestinal tract in adolescent: A case report
Huang WP, Li LM, Gao JB
- 6184** Toripalimab combined with targeted therapy and chemotherapy achieves pathologic complete response in gastric carcinoma: A case report
Liu R, Wang X, Ji Z, Deng T, Li HL, Zhang YH, Yang YC, Ge SH, Zhang L, Bai M, Ning T, Ba Y
- 6192** Presentation of Boerhaave's syndrome as an upper-esophageal perforation associated with a right-sided pleural effusion: A case report
Tan N, Luo YH, Li GC, Chen YL, Tan W, Xiang YH, Ge L, Yao D, Zhang MH
- 6198** Camrelizumab-induced anaphylactic shock in an esophageal squamous cell carcinoma patient: A case report and review of literature
Liu K, Bao JF, Wang T, Yang H, Xu BP
- 6205** Nontraumatic convexal subarachnoid hemorrhage: A case report
Chen HL, Li B, Chen C, Fan XX, Ma WB
- 6211** Growth hormone ameliorates hepatopulmonary syndrome and nonalcoholic steatohepatitis secondary to hypopituitarism in a child: A case report
Zhang XY, Yuan K, Fang YL, Wang CL
- 6218** Vancomycin dosing in an obese patient with acute renal failure: A case report and review of literature
Xu KY, Li D, Hu ZJ, Zhao CC, Bai J, Du WL
- 6227** Insulinoma after sleeve gastrectomy: A case report
Lobaton-Ginsberg M, Sotelo-González P, Ramirez-Renteria C, Juárez-Aguilar FG, Ferreira-Hermosillo A
- 6234** Primary intestinal lymphangiectasia presenting as limb convulsions: A case report
Cao Y, Feng XH, Ni HX
- 6241** Esophagogastric junctional neuroendocrine tumor with adenocarcinoma: A case report
Kong ZZ, Zhang L

- 6247** Foreign body granuloma in the tongue differentiated from tongue cancer: A case report
Jiang ZH, Xu R, Xia L
- 6254** Modified endoscopic ultrasound-guided selective N-butyl-2-cyanoacrylate injections for gastric variceal hemorrhage in left-sided portal hypertension: A case report
Yang J, Zeng Y, Zhang JW
- 6261** Management of type IIIb dens invaginatus using a combination of root canal treatment, intentional replantation, and surgical therapy: A case report
Zhang J, Li N, Li WL, Zheng XY, Li S
- 6269** Clivus-involved immunoglobulin G4 related hypertrophic pachymeningitis mimicking meningioma: A case report
Yu Y, Lv L, Yin SL, Chen C, Jiang S, Zhou PZ
- 6277** *De novo* brain arteriovenous malformation formation and development: A case report
Huang H, Wang X, Guo AN, Li W, Duan RH, Fang JH, Yin B, Li DD
- 6283** Coinfection of *Streptococcus suis* and *Nocardia asiatica* in the human central nervous system: A case report
Chen YY, Xue XH
- 6289** Dilated left ventricle with multiple outpouchings – a severe congenital ventricular diverticulum or left-dominant arrhythmogenic cardiomyopathy: A case report
Zhang X, Ye RY, Chen XP
- 6298** Spontaneous healing of complicated crown-root fractures in children: Two case reports
Zhou ZL, Gao L, Sun SK, Li HS, Zhang CD, Kou WW, Xu Z, Wu LA
- 6307** Thyroid follicular renal cell carcinoma excluding thyroid metastases: A case report
Wu SC, Li XY, Liao BJ, Xie K, Chen WM
- 6314** Appendiceal bleeding: A case report
Zhou SY, Guo MD, Ye XH
- 6319** Spontaneous healing after conservative treatment of isolated grade IV pancreatic duct disruption caused by trauma: A case report
Mei MZ, Ren YF, Mou YP, Wang YY, Jin WW, Lu C, Zhu QC
- 6325** Pneumonia and seizures due to hypereosinophilic syndrome – organ damage and eosinophilia without synchronisation: A case report
Ishida T, Murayama T, Kobayashi S
- 6333** Creutzfeldt-Jakob disease presenting with bilateral hearing loss: A case report
Na S, Lee SA, Lee JD, Lee ES, Lee TK

LETTER TO THE EDITOR

- 6338** Stem cells as an option for the treatment of COVID-19
Cuevas-González MV, Cuevas-González JC

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The primary aim of *World Journal of Clinical Cases* (*WJCC*, *World J Clin Cases*) is to provide scholars and readers from various fields of clinical medicine with a platform to publish high-quality clinical research articles and communicate their research findings online.

WJCC mainly publishes articles reporting research results and findings obtained in the field of clinical medicine and covering a wide range of topics, including case control studies, retrospective cohort studies, retrospective studies, clinical trials studies, observational studies, prospective studies, randomized controlled trials, randomized clinical trials, systematic reviews, meta-analysis, and case reports.

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Clivus-involved immunoglobulin G4 related hypertrophic pachymeningitis mimicking meningioma: A case report

Yang Yu, Liang Lv, Sen-Lin Yin, Cheng Chen, Shu Jiang, Pei-Zhi Zhou

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Abstract

BACKGROUND

Immunoglobulin G4 related disease (IgG4-RD) is a fibroinflammatory disease with markedly elevated serum IgG4 levels and fibrous tissue proliferation, accompanied by numerous plasma cells. IgG4 related hypertrophic pachymeningitis (IgG4-RHP) is relatively rare and indistinguishable from other phymatoid diseases before the operation. The risk of long-term immunosuppression needs to be balanced with disease activity.

CASE SUMMARY

A 40-year-old man presented with headache and bilateral abducent paralysis. He was also diagnosed with pulmonary tuberculosis 10 years ago and was on regular treatment for the same. Before the operation and steroid therapy, the patient was suspected of having tubercular meningitis at a local hospital. A clivus lesion was found *via* brain magnetic resonance imaging (MRI) at this presentation. He was preliminarily diagnosed with meningioma and underwent Gamma Knife Surgery. Transnasal endoscopic resection was performed to treat deterioration of nerve function. Postoperative pathologic examination suggested IgG4-RD. Moreover, the serum IgG4 was elevated at 1.90 g/L (reference range: 0.035-1.500 g/L). After steroid therapy for 2 mo, the lesion size diminished on MRI, and the function of bilateral abducent nerves recovered.

CONCLUSION

IgG4-RHP is relatively rare and indistinguishable before the operation. Elevated serum IgG4 levels and imaging examination help in the diagnosis of IgG4-RHP. Surgery is necessary when lesions progress and patients start to develop cranial nerve function deficit.

Key Words: Immunoglobulin G4 related disease; Hypertrophic pachymeningitis; Immunoglobulin G4 related hypertrophic pachymeningitis; Clivus; Case report

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Core Tip: Immunoglobulin G4 related disease (IgG4-RD) is a fibroinflammatory disease with markedly elevated serum IgG4 levels and fibrous tissue proliferation, accompanied by numerous plasma cells. It is known to affect multiple organs. IgG4-related hypertrophic pachymeningitis (IgG4-RHP) is relatively rare and indistinguishable from IgG4-RD before the operation. Herein, we present a rare case of IgG4-RHP with intact magnetic resonance imaging and pathologic images. The case highlighted the differential diagnosis with other phymatoid lesions such as meningioma, fungal infection, and tuberculosis and the importance of comprehensive multidisciplinary treatment. Surgery becomes necessary when lesions progress and patients start to develop cranial nerve function deficit.

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INTRODUCTION

Immunoglobulin G4 related disease (IgG4-RD) was initially noticed in patients with autoimmune pancreatitis in 2001 and formally named in 2010, classified as sarcoidosis with different manifestations in several organs and the same pathological characteristics[1,2]. The main characteristic of IgG4-RD is elevated levels of serum IgG4. Moreover, the lesions are often tumescent with abundant IgG4-positive plasma cells and fibrosis. Such inflammatory lesions can be seen in the pancreas, kidney, lungs, salivary glands, and other organs. Specifically, the conditions of IgG4-RD in the central nervous system are meningitis and hypophysitis[3]. As for the IgG4-related hypertrophic pachymeningitis (IgG4-RHP), the clinical and imaging manifestation is similar to that of meningioma, posing a challenge for preoperative diagnosis[4,5]. Additionally, the time and scope of operation should be considered carefully. Finally, this disease is related to some bacterial infections, such as tuberculosis. And we need to weight the pros and cons between these infections and corticosteroid therapy for IgG4-RD. Herein, we report a rare case with IgG4-RHP at the clivus area mimicking meningioma and discuss the relevant literature.

CASE PRESENTATION

Chief complaints

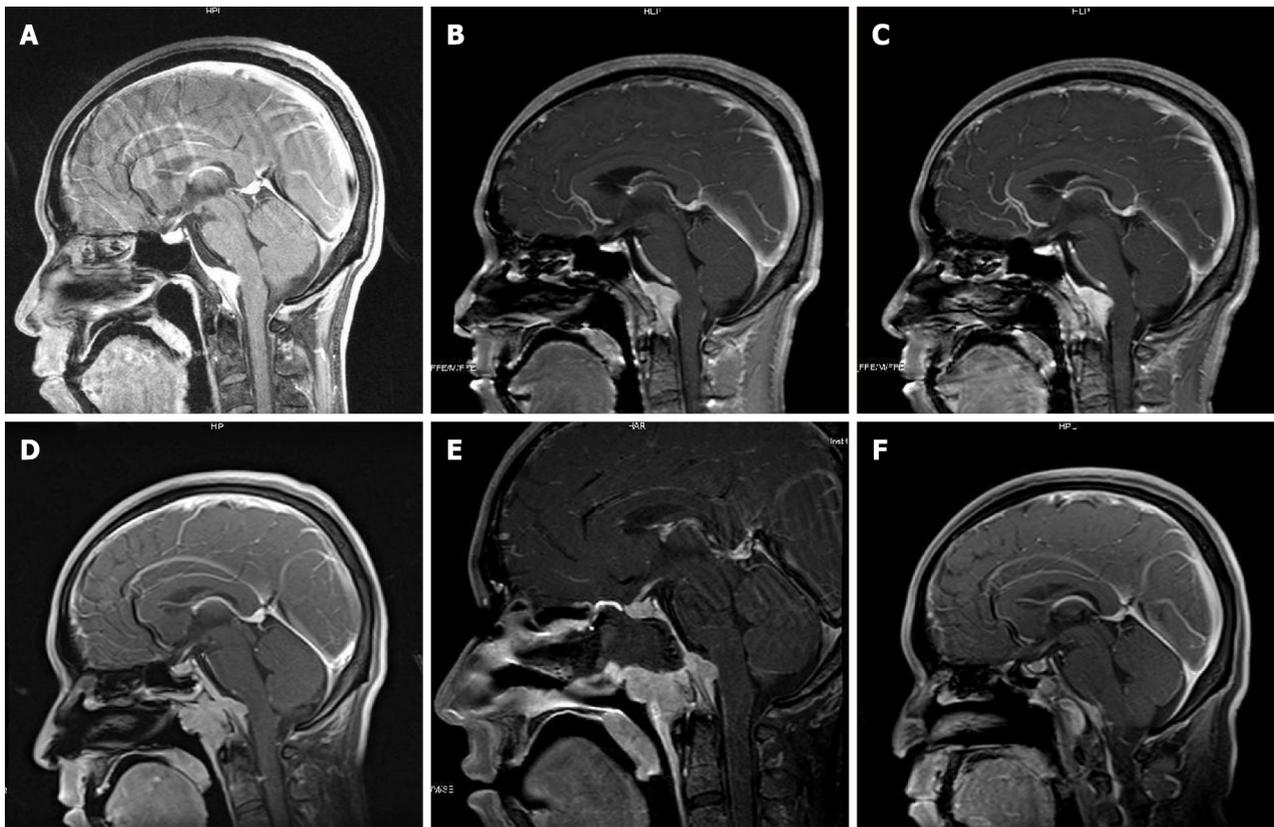
A 40 year-old man was admitted for headache, bilateral temporal visual field defect, and limited abduction in both eyes.

History of present illness

Five years before the present admission, the patient started to experience discontinuous and aggravating headache. Owing to symptomatic deterioration, the patient was admitted to the neurology department of a local hospital. Because the patient also had a history of pulmonary tuberculosis, he was suspected of having tuberculosis meningitis and treated with anti-tuberculosis drugs at the local hospital. However, the symptom did not alleviate. Upon presentation to our hospital, the patient underwent a brain magnetic resonance imaging (MRI) scan that showed the presence of a clival lesion measuring $2.6 \times 1 \text{ cm}^2$ with isointense signal on T1-weighted (T1WI) and T2-weighted (T2WI) imaging; accordingly, he was diagnosed with meningioma. The lesion was homogeneously enhanced on contrast MRI with a dural tail sign (Figure 1). Because there was no cranial nerve function defect, the patients chose to undergo Gamma Knife Surgery at a dose of 11 Gy at the 45% isodose line, and regular follow-up was planned.

History of past illness

The patient had suffered from pulmonary tuberculosis 11 years ago and accepted standard anti-tuberculosis treatment for 1 year.



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Figure 1 Lesion alteration on the sagittal view. A: This image shows the condition when the patient was first admitted to the tuberculosis department diagnosed as having a meningioma (December 2016); B: Before the performance of Gamma Knife Surgery (March 2017); C: Six months after the performance of Gamma Knife Surgery (September 2017); D: Preoperative image (November 2019); E: Postoperative image (January 2020); F: Two months after operation (March 2020).

Personal and family history

No other particular personal and family history was reported.

Physical examination

This patient showed right abducens paralysis, hoarse voice, bitemporal hemianopsia, and slight swallowing difficulty. No other positive signs were found.

Laboratory examinations

Lumbar puncture was performed and we found that the number of karyocytes (mainly mononuclear cells) and protein levels in cerebrospinal fluid had risen (Table 1).

After pathological results showed IgG4-RD, further systemic evaluation was performed to find other lesions associated with IgG4-RD. The serum IgG level was 17.20 g/L (reference range: 8.00-15.50 g/L), and the serum level of IgG4 was 1.90 g/L (reference range: 0.035-1.500 g/L). Tuberculosis associated gamma interferon release assay showed positive results with TB-IGRA (T-N) at 414.21 pg/mL.

Imaging examinations

After admission, routine laboratory testing and preoperative preparation were carried out. A repeat brain MRI scan showed that the lesion became larger, measured 3.8 cm × 2.9 cm × 2.9 cm, and compressed the adjacent brain stem (Figure 1). Further, small pneumatoceles in the upper lobe of the right lung were detected by thorax computed tomography (CT). Moreover, the examination of visual field confirmed binocular hemianopia (Figure 2). No other positive results were found.

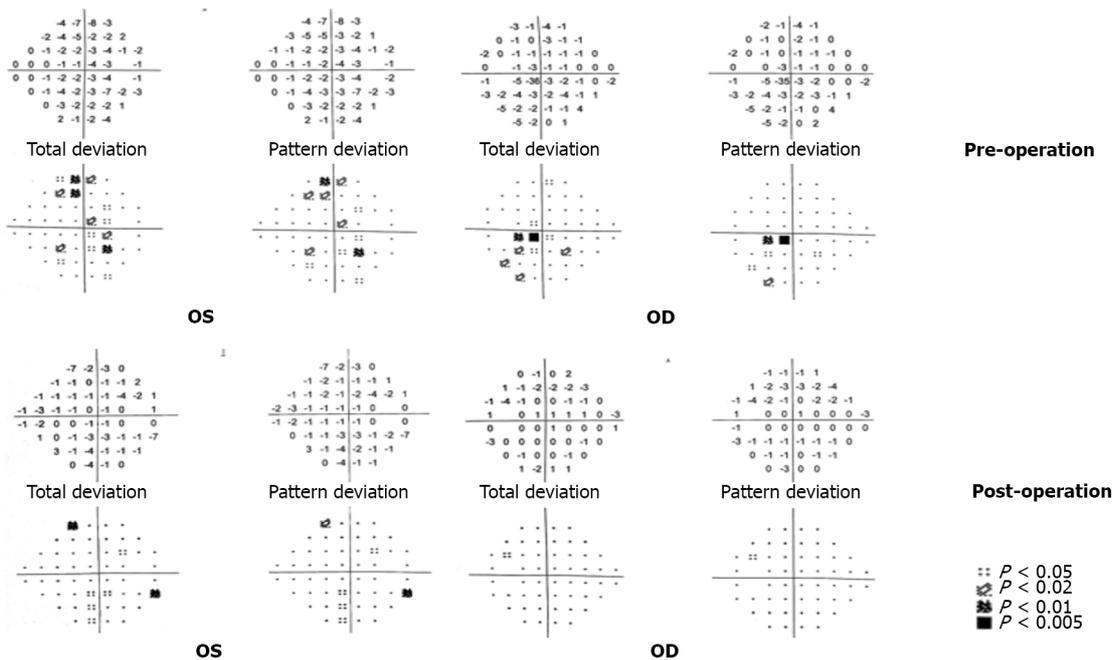
FINAL DIAGNOSIS

The postoperative pathology confirmed the proliferation of fibrous tissue accompanied by numerous lymphocytes and plasma cells, which is displayed in Figure 3. Immunohistochemical staining showed positive results for CD138 and IgG4. Gene rearrangement test showed negative results for IgH. Thus,

Table 1 Examination results of cerebral fluid through the years

	2014	2016	2020
General characters	Normal	Normal	Normal
Karyocytes (10 ⁶ /L)	120	90	1
Mononuclear cells (%)	97.0	92.0	-
Multinucleate cells (%)	3.0	8.0	-
Pus cells	Unseen	Unseen	Unseen
Protein quantification (g/L)	1.02	0.69	0.55
Glucose (mmol/L)	2.55	4.07	3.95
Chloride (mmol/L)	127.2	124.4	127
Acid-fast bacillus	None	None	None
Ink staining	None	None	None
Gram staining	None	None	None
Fungal and bacterial culture	Undone	Negative	Negative
TB-DNA	Undone	Negative	Negative

Reference value: Karyocytes (< 10 × 10⁶/L), pus cells (none), protein quantification (0.15-0.45 g/L), glucose (2.5-4.4 mmol/L), and chloride (120-130 mmol/L).



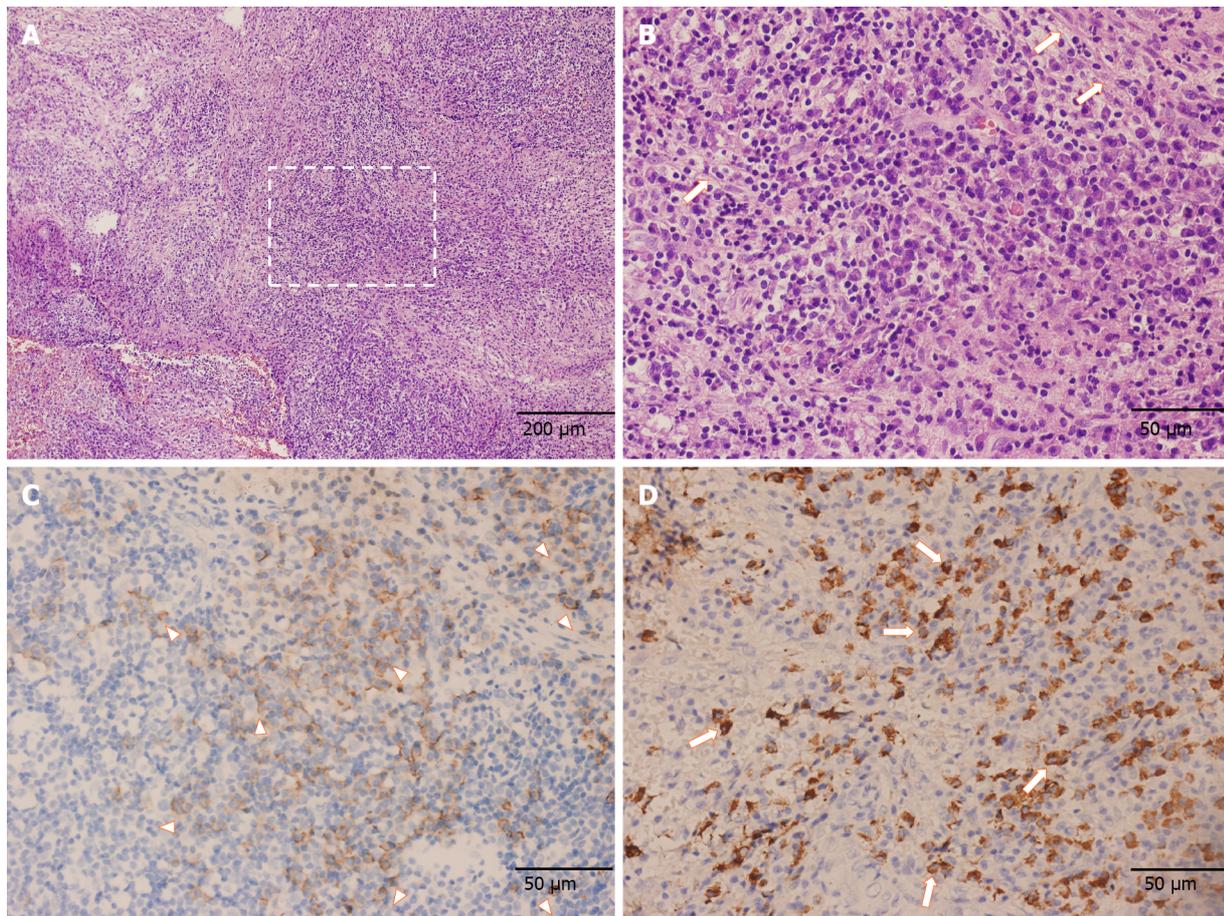
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Figure 2 Examination of visual field before and after the operation. The alteration of the visual field before and after the operation is shown. The second row is the result at 4 mo after the operation.

IgG4-RD was finally diagnosed.

TREATMENT

The patient underwent transnasal endoscopic approach resection which aimed to partially remove the lesion for pathology analysis and alleviate the headache caused by meningeal tension. During the operation, we found that the lesion extended to the sphenoid sinus and nasopharynx without a clear



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Figure 3 Pathological features of the resection part indicating the diagnosis of immunoglobulin G4 related hypertrophic pachymeningitis.

A and B: Proliferation of fibrous tissue accompanied by numerous lymphocytes and plasma cells as shown by hematoxylin and eosin staining (A: $\times 100$; B: $\times 400$); C: Immunohistochemical staining exhibited an increased number of CD138-positive plasma cells ($\times 400$); D: Large number of immunoglobulin G4-positive plasma cells as shown by immunohistochemical staining [~ 200 per high power field ($\times 400$)].

boundary. Notably, the local mucosa was edematous and tight. The clivus bone had been partially damaged, and the clivus epidural was thicker. The intraoperative frozen section examination revealed the proliferation of spindle cells accompanied by many lymphocytes and plasma cells.

For the diagnosis of IgG4-RD, solu medrol was administrated at a dose of 80 mg per day, and methotrexate was administrated at 10 mg every week. Famotidine, calcium carbonate, and vitamin D3 tablets were prescribed against adverse reactions during the treatment. After discharge from the hospital, the solu medrol was tapered over 4 wk to 50 mg per day.

OUTCOME AND FOLLOW-UP

The patient confirmed that his headache and hoarse voice gradually improved after 1 mo. The follow-up was arranged 3 mo after the operation, which showed that the abduction movement could be achieved for binocular vision. Brain MRI showed that the residual lesion obviously shrunk (Figure 1). The change for bilateral visual fields is displayed in Figure 2.

DISCUSSION

IgG4-RD is a condition that affects multiple organs, and its clinical manifestations often vary across different organs. Reportedly, several kinds of bacterial infection can be causative factors for this disease related to stimulation with Toll-like receptor ligands[6,7]. Several previous studies have also reported the comorbidity of IgG4-RD with tuberculosis, as seen in our patient[8-11].

IgG4-RD of the CNS is mainly related to IgG4-related hypertrophic pachymeningitis and hypophysitis. Among them, IgG4-RHP is relatively rare, with the primary clinical manifestation of headache and other nerve function disabilities. Moreover, it was apparent that the cranial nerve

function could partially recover once the disease was in remission. At the first onset of the disease, multi-organ disease is not widespread (57%)[6]. Therefore, regular follow-up and systemic evaluation is crucial.

Through this case, we summarize the differential diagnoses of IgG4-RHP, such as meningioma, tuberculosis meningitis, fungal meningitis, and metastatic tumor. Furthermore, the complete MRI images showed the lesion alteration during treatment. However, there are limited reports of this rare disease in the literature. Higher evidence-based studies are needed to promote the diagnosis and treatment of IgG4-RHP.

Measuring the serum concentration of IgG4, radiological examination, and pathological screening are important for diagnosis. It is difficult to distinguish IgG4-RHP and meningioma before the operation and pathologic examination. The serum level of IgG4 can facilitate diagnosis, but it does not always show an increase. As reported by Wallace *et al*[12], the sensitivity and specificity of serum IgG4 were 90% and 60%, respectively. Moreover, the negative predictive value and positive predictive value of the serum IgG4 assay were 96% and 34%, respectively, which could be helpful and convenient to exclude the diagnosis of IgG4-RD related to the CNS[12]. It is also helpful to distinguish tuberculosis and IgG4-RD based on the fact that serum IgG4 does not significantly increase in tuberculosis[13]. Further, imaging results could be a crucial clue for preoperative diagnosis. Lumbar puncture provides the necessary information for differentiation from CNS infections and malignant tumors. IgG4 levels in cerebrospinal fluid have been reported to be elevated[14]. However, the concentration of IgG4 in cerebrospinal fluid could not distinguish this disease from other inflammatory pachymeningitis[6].

Radiology examination plays an essential role in diagnosis. The lesion could be observed as linear dural thickening or a bulging mass. The linear dural thickened lesion appears both in the brain and spine. The tumoral lesion is frequently located in the clivus area. The heterogeneity was observed on MRI because of active inflammation. Typically, T1WI MRI would exhibit a hyperintense or isointense signal. Hypertrophic pachymeningitis usually shows thickening meninges and hypointensity on T2WI MRI, while it would become relative hyperintense when the inflammation aggravates[3,4,6,7,15]. The lesion would be homogeneously enhanced on enhanced MRI. In this case, the lesion showed an isointense signal on T1WI and T2WI and was homogeneously enhanced on contrast MRI with a dural tail sign. CT showed that the skull was involved apparently and the lesion appeared hyperdense when contrast-enhanced CT was performed. In case of a meningioma, CT frequently displays that the lesion is isodense or has slightly higher density with a round, leafy, or flat shape[3,6]. Calcification becomes visible in some tumors[6]. Meningioma has similar characteristics as an IgG4-RHP lesion. T1WI often shows isointense or mildly hypointense signal, and T2WI usually shows isointense or mildly hyperintense signal. Besides, the meningioma could be markedly characterized by the tail of the meninges.

It is advisable to focus on some characteristics to help distinguish between meningioma and IgG4-RHP. We noticed that the symptoms of IgG4-RHP were severe and diverse, while those of meningioma were not as varied. These symptoms were due to inflammatory irritation and compression of the adjacent nerves and dura mater[16]. Another characteristic of IgG4-RHP was that the tail signal was broader than meningioma on MRI for the diffuse inflammation along with the dura mater. The meningioma lesion seems relatively confined and phymatoid compared with IgG4-RHP. Moreover, the IgG4-RHP lesion frequently involves extracranial parts.

Other diseases, such as metastatic tumors and fungal infections, should also be considered. It was observed that metastatic tumors could spread and proliferate along the meninges, causing various severe symptoms. In this situation, the history of malignant tumor provided clues to the diagnosis. Likewise, a CNS fungal infection can show similar features, which can be identified by examining the cerebrospinal fluid.

CNS tuberculosis is another antidiastole. Patients with tuberculous meningitis often have a fever, headache, and focal neurological symptoms. And tuberculous meningitis is often secondary to pulmonary or intestinal tuberculosis. As for radiology examination, CT often exhibits nodular or punctate calcifications and hydrocephalus, and enhanced scans are often accompanied by meningeal strengthening. MRI frequently shows a hypointense T1WI signal and hyperintense T2WI signal. The enhancement scan could display irregular bar or nodular strengthening lesions of the meninges. Cerebrospinal fluid is essential for the diagnosis of tuberculous meningitis. Moreover, TB-IGRA could facilitate this diagnosis.

The purpose of the operation was not only to perform a biopsy but also to alleviate symptoms. We know that the lesion would stretch meninges and then cause headache. Similarly, the lesion compresses cranial nerves to cause relevant symptoms. The resection can reduce meningeal tension, release compression, and finally alleviate headache and nerve deficits. Further, it is suitable to use the transnasal endoscopic approach for a clival lesion in IgG4-RHP. When the lesion is too broad to remove completely, it is sensible to leave some parts in order to maintain the integrity of the dura mater, which can prevent severe complications such as cerebrospinal fluid leakage and intracranial infection.

Glucocorticoids and immunosuppressants can be used for the non-surgical treatment, such as prednisolone (0.6 mg/kg/d) for 4 wk. The dose of steroid was gradually decreased through 3-6 mo and the dose was finally maintained at 2.5 to 5.0 mg/d for 3 years[17]. Other immunosuppressants should be considered, such as methotrexate, cyclophosphamide, mycophenolate mofetil, and azathioprine[6,7].

Another consensus recommended utilizing calcium carbonate and vitamin D3 tablets to prevent glucocorticoid-induced osteoporosis[18,19]. Additionally, it is essential to exclude some latent infections before using glucocorticoids and immunosuppressants. In this case, the patient had a history of tuberculosis and we performed the chest CT and TB-IGRA to ensure the absence of any current underlying infection. In future, when similar patients with the imaging characteristics described in this report are encountered, measurement of serum IgG4 levels may be helpful for diagnosis.

CONCLUSION

IgG4-RHP is a relatively rare disease that seems complicated to diagnose preoperatively. The purpose of surgery is to obtain the specimens required for pathological examination and plan the follow-up treatment. It is essential to perform a rigorous follow-up and systematic assessment of the whole body.

FOOTNOTES

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