MEGA JOURNAL OF CASE REPORTS

OPEN ACCESS

Case Presentation

Compiled Date: August 09, 2025

Stereotactic Radiosurgery for Isolated Intracranial IgG4 Disease: An Illustrative Case

Mariagrazia Nizzola, Department of Neurosurgery and Gamma Knife Radiosurgery, I.R.C.C.S. San Raffaele Scientific Institute, Vita-Salute University, via olgettina 60, 2013, Milan, Italy, Tel: 390226439426

Department of Neurologic Surgery, Mayo Clinic, Rochester, MN 55905, USA

Rhoton Neurosurgery and Otolaryngology Surgical Anatomy Program, Mayo Clinic, Rochester, MN 55905, USA

Abstract

Isolated intracranial IgG4 disease is a rare form of IgG4-related disease involving IgG4-positive plasma cell infiltration along the meninges, often mimicking other pathologies and leading to misdiagnosis. A 72-year-old man with left cranial nerve VI palsy was initially suspected of having a tentorial meningioma. Stereotactic radiosurgery with Gamma-Knife led to the resolution of symptoms and radiological improvement. A subsequent frontal meningeal lesion biopsy confirmed isolated IgG4 disease. This case highlights the diagnostic complexity of the condition and suggests that stereotactic radiosurgery may have a potential role in managing and improving neurologic symptoms related to intracranial IgG4 lesions.

Background and Importance

Isolated intracranial IgG4 disease, a rare subset of IgG4-related diseases, represents a unique challenge in both the neurology and immunology fields [1,2]. It is characterized by the infiltration of IgG4-positive plasma cells along the meninges of the central nervous system; it leads to a range of clinical manifestations that depend on the location of the inflammatory lesion and the compression of neural structures, leading to functional deficits [3]. Misinterpretations of the clinical and imaging findings are common, and they pose a significant diagnostic and therapeutic dilemma [4-7]. The advent of Stereotactic Radiosurgery with Gamma Knife (SRS-GK) has significantly changed the treatment approach for various intracranial lesions and conditions. In the context of isolated intracranial IgG4 disease, SRS-GK has not been described as a treatment option, but its application is associated with misdiagnosis, particularly with intracranial meningioma [8]. In this paper, we present a case report that illustrates a peculiar clinical scenario at the intersection of isolated intracranial IgG4 disease

and SRS-GK. By providing a detailed analysis of the patient's clinical course, radiological findings, treatment course involving SRS-GK, and subsequent clinical outcomes, we aim to provide insights into considerations involved in the integration of SRS-GK into the management of isolated intracranial IgG4-related lesions.

Clinical Presentation

A 72-year-old man was admitted to our department in June 2021 with a sudden onset of left-sided eye chemosis and diplopia due to left Cranial Nerve (CN) VI palsy. MRI revealed an enhancing mass within the left cavernous sinus, extending posteriorly along the left tentorial free edge, initially suggesting meningioma. The patient underwent stereotactic radiosurgery using Gamma Knife. The treatment planning system utilized was Leksell Gamma Plan (Elekta Instrument). At the three-month follow-up, complete resolution of CN VI palsy was observed, and the MRI results indicated a significant response to the initial treatment. In 2024, a subsequent meningeal lesion was identified along the frontal convexity, notably without any clinical symptoms. The immunohistochemical analysis of the cerebrospinal fluid produced non-diagnostic findings for any specific disease. Thus, a craniotomy and excision for histological examination were deemed necessary. The definitive histological evaluation subsequently revealed isolated intracranial IgG4, resulting in a referral to the rheumatology department for ongoing disease follow-up. The patient signed the informed consent form for personal data treatment for procedures and research, and all the procedures were performed according to ethical standards.

Discussion

The radio-responsiveness of IgG4-related disease has been demonstrated in several noteworthy clinical cases. Prominent examples include those with orbital involvement [9], a case involving the parotid gland [10], and an intracranial lesion, initially mistakenly identified as a meningioma, and successfully treated with linear accelerator therapy, yielding positive outcomes [8]. SRS-GK represents a precise and non-invasive form of radiation therapy and offers a targeted approach for treating intracranial lesions while minimizing damage to surrounding healthy tissue. Studies exploring the efficacy of SRS-GK in IgG4-related diseases are not described in the literature, as the standard immunomodulation therapy is very effective. This case report depicts the first application of SRS-GK in treating this disease, despite the initial misdiagnosis of the lesion as a meningioma. The application of SRS-GK in IgG4 disease is discussed from a retrospective standpoint. Moreover, the notable and complete resolution of neurological symptoms after SRS-GK treatment highlights the effectiveness of this approach in managing the correlates of neurological symptoms. Furthermore, this report emphasizes the crucial point that IgG4-related disease should be considered a potential differential diagnosis for intracranial lesions, especially when the clinical context is appropriate. While hypophysitis and pachymeningitis are commonly recognized conditions linked to the neurological manifestations of IgG4 disease, intracranial pseudotumor remains rare. To date, only a limited number of cases involving multiple pseudotumors have been documented [6,7,11,12]. Highlighting the diagnostic challenges and therapeutic successes associated with this case it reinforces the importance of a multidisciplinary approach in the assessment and management of isolated intracranial IgG4-related disease. This case not only broadens the treatment options for this complex neuroinflammatory disorder but also highlights the need for increased clinical awareness and careful diagnostic evaluation to achieve optimal patient outcomes. However, several considerations warrant further investigation and discussion. These include the optimal dosing and schemes for SRS-GK

in IgG4-related disease, long-term efficacy and safety outcomes, and the potential for disease recurrence.

Conclusion

This case aims to offer valuable insights into the current literature and emphasize the potential role of SRS-GK in treating isolated intracranial IgG4 disease, particularly when neurological deficits are present.

References

1. Goulam-Houssein S, Grenville JL, Mastrocostas K, et al. IgG4-related intracranial disease. Neuroradiol J. 2019;32(1):29-35.
2. Kamisawa T, Zen Y, Pillai S, Stone JH. IgG4-related disease. Lancet. 2015;385(9976):1460-71.
3. Esmaeilzadeh M, Dadak M, Atallah O, et al. IgG4-related hypertrophic pachymeningitis with tumor-like intracranial and intracerebral lesions. Acta Neurochir (Wien). 2022;164(10):2781-7.
4. Qing P, Lu C, Yan B, et al. Case report: IgG4-related intracranial lesions mimicking multiple sclerosis in a 14-year-old girl. Front Neurol. 2022;13:1007153.
5. Takeuchi S, Osada H, Seno S, Nawashiro H. IgG4-Related Intracranial Hypertrophic Pachymeningitis : A Case Report and Review of the Literature. J Korean Neurosurg Soc. 2014;55(5):300-2.
6. Ohyama K, Tanaka M, Nosaki Y, Yokoi T, Iwai K. IgG4-related Inflammatory Pseudotumor with Imaging Findings Similar to Meningioma. Intern Med. 2023;62(11):1665-9.
7. Liu X, Wang R, Li M, Chen G. IgG4-Related Inflammatory Pseudotumor Involving the Clivus: A Case Report and Literature Review. Front Endocrinol (Lausanne). 2021;12:666791.
8. Fogarty T, Choong D, Gill S, Harper C, Robbins P. Stereotactic radiosurgery for intracranial IgG4-related disease. J Med Imaging Radiat Oncol. 2019;63(5):707-710.
9. Andrew N, Kearney D, Selva D. IgG4-related orbital disease: a meta-analysis and review. Acta Ophthalmol. 2013;91(8):694-700.
10. Roos DE, Dreosti MV, James CL, Hissaria P. Radiotherapy for parotid IgG4-related disease. J Med Radiat Sci. 2019;66(1):66-69.
11. Okano A, Nakatomi H, Shibahara J, Tsuchiya T, Saito N. Intracranial Inflammatory Pseudotumors Associated with Immunoglobulin G4-Related Disease Mimicking Multiple Meningiomas: A Case Report and Review of the Literature. World Neurosurg. 2015;83(6):1181 e1-4.
12. Kuroda N, Inenaga C, Arai Y, Otsuki Y, Tanaka T. Intracranial Multiple Pseudotumor Due to Immunoglobulin G4-Related Disease without Other Lesions: Case Report and Literature Review. World Neurosurg. 2019;132:69-74.

Citation of this Article

Nizzola M. Stereotactic Radiosurgery for Isolated Intracranial IgG4 Disease: An Illustrative Case. Mega J Case Rep. 2025;8(8):2001-2004.

Copyright