中文題目：以視覺缺失表現的 IgG4 相關性疾病

英文題目：A case of IgG-4 related disease with visual defects presentation

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**Introduction:**

Immunoglobulin G4-related disease (IgG4-RD) is a regional or systemic fibroinflammatory disease of unknown etiology. It usually presents compressive or swelling lesions, but not always elevated serum IgG4 levels. The key histopathological appearances are a dense lymphoplasmacytic infiltration, particularly IgG4-positive cells, a storiform pattern of fibrosis and obliterative phlebitis. The disease may involve thyroid, pancreas, lymph nodes, salivary glands, pituitary glands, kidney and retroperitoneum. However, it is rare involvement of the optic nerve, and visual impairment is an uncommon symptom. Glucocorticoids are typically the first line of therapy. We present a case, who had blurred vision and bitemporal hemianopia was diagnosed IgG4 related disease during hospitalization. He exhibited good response to methylprednisolone and lesion was rapid subsided.

**Case presentation:**

The patient is a 34-year-old man who had received partial gastrectomy and cystogastrostomy for large pancreatic pseudocyst in 2019 and recurrent pancreatic pseudocyst infection. He visited our neurologic outpatient department for blurred vision and bitemporal hemianopia in recent one month. Initial differential diagnosis was pituitary gland tumor or other pathology as a result of optic chiasm lesion. Therefore, he was admitted to an ordinary ward and received an MRI for further evaluation.

Brain MRI showed bilateral hypertrophic pachymeningitis, hypophysis involved optic chiasma, and inflammation (pseudotumor) of bilateral optic nerve sheath. Autoimmune related disease was considered. Rheumatologist was consulted and an autoimmune panel showed negative of ANA, ENA, MPO, PR3 and RF. However, a high titer of IgG4(6480 mg/dL) was disclosed.

Later, whole body CT with/without contrast showed multiple enlarged lymph nodes at bilateral neck, axilla, mediastinum, retroperitoneum, and bilateral pelvic. It also revealed multiple poor-enhanced lesions in bilateral kidneys and mild enlargement of pancreas. Due to these abnormalities noted under CT image, she received a right kidney sono-guided biopsy for lesion in low echogenicity. The pathologic result showed a dense lymphoplasmacytic infiltration, storiform fibrosis and obliterative phlebitis. Immunohistochemistry stains show IgG4 to IgG ratio≥90% and IgG4 count >100 / HPF. The pathologic result combined with elevated serum IgG4 level, and he was diagnosed with IgG4-related disease.

He received methylprednisolone 20 mg BID IV and his visual problem and proptosis have resided significantly within days. He received methylprednisolone pulse therapy for 3 days and used azathioprine, hydroxychloroquine and prednisolone as maintenance treatment. At eight months follow up, we found that previous splenomegaly and lymphadenopathy have resolved along with a normal pancreatic size on CT scan.

**Conclusion:**

IgG4- related disease is a rare and fibroinflammatory disease of unknown etiology. It usually involved multiple organs, but some of patients have disease that is confined to single organ for many years and that would be misdiagnosed sometimes. In our presentation, the case has blurred vision and hemianopia initially. After image study and enlarged lymph nodes biopsy, the typical pathologic result was found. He also had high titers of serum IgG4. His visual problem is subsided and resolved lymphadenopathy and splenomegaly after corticosteroid treatment. Due to early diagnosis and treatment, the patient can well preserve his visual function.