Metastatic colorectal cancer patient with tumefactive demyelinating lesion from NMOSD

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Introduction: Tumefactive demyelinating lesion(TDL) is defined as a solitary demyelinating lesion in the brain greater than 2 cm. Neuromyelitis optica spectrum disorder(NMOSD) is an autoimmune disease of the CNS characterized by inflammatory lesions. We present a case of NMOSD with right upper extremity weakness with TDL of the brain on imaging during palliative FOLFOX chemotherapy(CTx).

Case presentation: A 59-year-old man had been receiving palliative FOLFOX CTx for metastatic colorectal cancer. After 6 cycles, he complained numbness of the rt hand. The patient was initially treated with pregabalin for peripheral neuropathy(PN). Soon, he visited the ophthalmology for the sudden loss of rt vision; no light perception. Further brain magnetic resonance(MR) imaging showed abnormal signals in the right optic nerve and left precentral gyrus lesion. It suggested unilateral optic neuritis(ON) and brain metastasis(Figure 1). After the multidisciplinary meeting, the brain lesion was considered a TDL rather than metastasis or primary glioma. Craniotomy for excisional biopsy was performed, and histopathological findings revealed abundant foamy macrophages signed by CD68 and reactive gliosis, which were compatible with TDL(Figure 2). Although serum AQP4-IgG test was negative, we could diagnose NMOSD since there were 2 core clinical characteristics; ON and symptomatic cerebral syndrome (typical brain lesion). Initial treatment with iv methylprednisolone 1 g for 3 days had no neurological improvement; therefore, the therapeutic plasma exchange was carried out 7 times for 2 wks. The patient gradually improved the rt-hand weakness but did not recover his vision. Follow-up MR imaging showed improved ON and nearly resolved TDL in the left precentral gyrus (Figure 3).

Discussion: NMOSD is a rare syndrome with less than 1% of demyelinating disease. The core clinical characteristics implicate CNS region. Our case presented with PN at the 1st onset, and then CNS involving symptoms occurs and showed negative AQP4-IgG. The unusual presentation of seronegative NMOSD represents a diagnostic challenge, and this case report will contribute to the few cases reported in the literature.

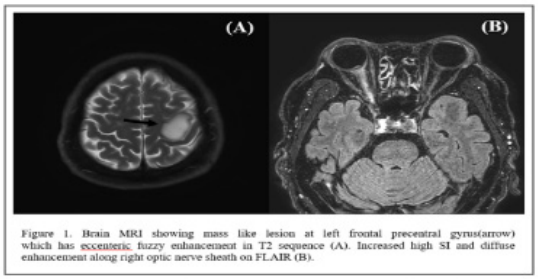

Figure 1. Brain MRI showing mass like lesion at left frontal precentral gyrus(arrow) which has eccenteric fuzzy enhancement in T2 sequence (A). Increased high SI and diffuse enhancement along right optic nerve sheath on FLAIR (B).

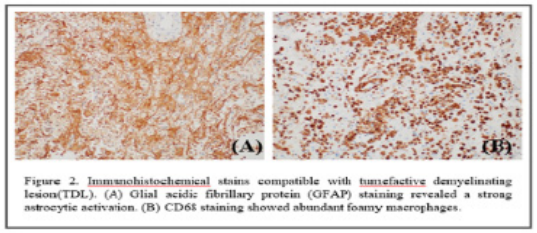

Figure 2. Immunohistochemical stains compatible with tumefactive demyelinating lesion(TDL). (A) Glial acidic fibrillary protein (GFAP) staining revealed a strong astrocytic activation. (B) CD68 staining showed abundant foamy macrophages.

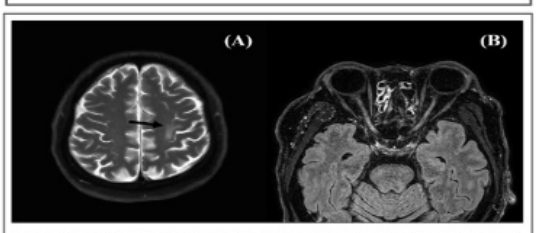

Figure 3. Following brain MRI showing improved inflammation of right optic nerve and left precentral gyrus. Small patchy gliotic lesion (arrow) in T2 sequence (A). Decreased right optic nerve sheath swelling, enhancement on FLAIR (B).