Letter to the Editor

Optic Neuritis as Isolated Presentation of Kikuchi-fujimoto Disease in Pediatric Patient

Arslan et al. Optic Neuritis as Isolated Presentation of Kikuchi-fujimoto Disease

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To the Editor
Kikuchi-Fujimoto disease (KFD), also known as histiocytic necrotizing lymphadenitis, is an idiopathic, rare, benign disease. It is self-limiting and characterised predominantly by cervical lymph node enlargement in young women especially of Asian decent (1,2).
A 12-year-old female patient was seen in ophthalmology clinic with the complaints of mild color deficiency, specifically red desaturation, in her right eye. The symptoms first began 1 month prior to examination. On ophthalmology examination; she had 20/20 vision in both eyes with no afferent pupillary defect (APD). Color vision was diminished in the right eye (5/10 color plates in the right eye, 10/10 in the left). Visual fields were full to confrontation. Automated visual field testing was unreliable in both eyes and failed to reveal a focal deficit in the right eye.
Extraocular muscle motility was normal. Her external exam and anterior segments in both eyes were unremarkable. Funduscopic exam was notable for optic nerve pallor on the right.
Laboratory testing showed elevated ESR/CRP levels but other blood test values were within the normal ranges as detailed on Table 1. Her past medical history and medications were unremarkable with the exception of persistent firm and intermittently painful submental and submandibular lymphadenopathy, which had been treated with multiple antibiotics. The year prior to presentation to ophthalmology, she had a submental lymph node biopsy, which resulted in the diagnosis of KFD. At time of presentation, she was not receiving any targeted treatment for this diagnosis.
Magnetic resonance imaging (MRI) of the brain and orbits with contrast showed moderate asymmetric T2 hyperintense signal of the right optic nerve; also asymmetric enhancement and thickening of the orbital and intracanalicular segments of the right optic nerve compared to the left, compatible with optic neuritis (figure 1). Brain parenchyma demonstrated normal signal intensity without evidence of mass, hemorrhage, midline shift, or abnormal enhancement.
She was started on oral prednisone 20 mg daily and tapered over 3 weeks. Follow-up MRI 4 months later revealed minimally asymmetric contrast enhancement of the right optic nerve substantially decreased compared to the prior study. There was also minimally asymmetric T2 hyperintense signal of the right optic nerve. Her symptoms were decreased but not resolved at the time of the MRI. Therefore her ophthalmologist decided to give her infusion of rituximab 375 mg/m2 for 4 weeks.
A follow-up ophthalmology exam after 1 year demonstrated a complete resolution with equal color vision in the eyes. Written informed consent was obtained from patient's parents.
KFD is a very rarely reported entity with ocular manifestations (only 22 patients including our case) in literature. To summarize; ocular manifestations of KFD were: uveitis, retinal vein vasculitis, optic neuritis (2 patients, including our patient), lacrimal gland involvement, oculomotor palsy, papillary conjunctivitis, eyelid edema, peri-orbital edema, subretinal macular infiltrate, conjunctival injection, choroidal edema, papillary edema, intraconical attack of apex. We present here a 12-year-old female patient with KFD and unilateral optic neuritis.
Optic neuritis is an acute inflammation of the optic nerve and one of the common causes of optic neuropathy. The causes of optic neuritis can be divided into infectious and noninfectious categories. Noninfectious category

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includes multiple sclerosis (MS), acute disseminated encephalomyelitis (ADEM), neuromyelitis optica (NMO), other systemic disorders such as lupus, or sarcoidosis. Exposure toxins and radiation can also cause optic neuropathy (3,4).

In our patient there were no findings of central nervous system demyelinating disease on MRI including MS, ADEM, or NMO. The brain MRI was normal. Her serum NMO-IgG test was negative. Her clinical presentation, clinical examination and laboratory tests did not support a diagnosis of rheumatologic or systemic diseases such as SLE, sarcoidosis and there was no prior history of radiation treatment. The negative serodiagnostic tests helped to rule out HSV, HIV and VZV. Therefore, her immunosuppressive-treatment responsive optic neuritis was thought to be related to KFD.

When evaluated together with the other optic neuritis case in the literature; KFD-induced lymphadenopathy and optic neuritis findings were at different times in both cases. While there was a period of one year in our case; it was simultaneous in the previous case. Additionally, The previous patient spontaneously recovered in 4 weeks. We used Rituximab and systemic steroid therapy and our patient recovered after one year (2).

In conclusion, clinicians should be aware that KFD may be associated with ocular findings, such as optic neuritis. Follow-up of KFD patients with ocular involvement is recommended for potential recurrence or long-term damage.

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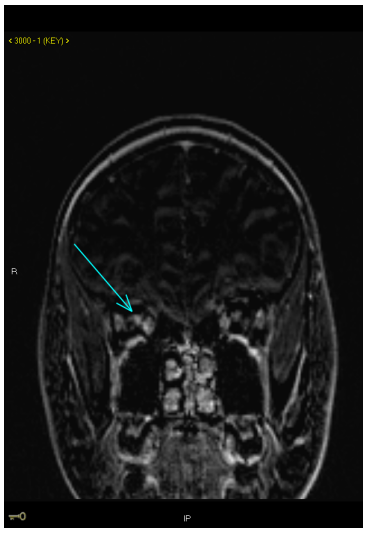

FIG 1. 12-year-old female with Kikuchi-Fujimoto Disease. Subtraction post contrast T1 weighted coronal image showed asimetric abnormal enhancement in right optic nerve (arrow).

TABLE 1. Blood test values of our patient

	At the diagnosis	**1 year later**
Perinuclear (P-ANCA)	negative	
Cytoplasmic, (c-ANCA)	negative	
Myeloperoxidase (MPO)	negative	
Proteinase-3 Antibody (PR3)	negative	
RNP Antibody, Serum	negative	negative
Smith Antibody, Serum	negative	negative
Anti-Ro(SS-A), Serum	negative	negative
Anti-La (SS-B), Serum	negative	negative
C3 Complement, Serum	121	140
C4 Complement, Serum	24	28
Anti-Nuclear Ab	negative	negative
Anti-DNA Screen, Serum	negative	negative
Rheumatoid Factor	negative	negative
Cyclic Citrulline Pep Ab	negative	negative
Erythrocyte Sed Rate	69	48
NMO-IgG, ELISA	negative	negative

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