Neurology Asia 2011; 16(1) : 81 – 83

CASE REPORTS

Encephalitis associated to Kikuchi-Fujimoto's disease in a young woman: A case report

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Abstract

Kikuchi-Fujimoto's disease or histiocytic necrotizing lymphadenitis affects predominantly young women and most of the cases have a benign and self-limited course with a complete remission within a few months. The involvement of the central nervous system is very unusual. We present the case of a young woman who developed temporal lobe encephalitis 3 months after the onset of Kikuchi-Fujimoto's disease. The patient was treated with corticosteroid and achieved a relative good outcome although there were remaining mild cognitive impairment and behavioral disturbances.

INTRODUCTION

Kikuchi-Fujimoto's disease (KFD) or histiocytic necrotizing lymphadenitis, was reported at first time in 1972, in Japan.[1,2] It is an idiopathic disease, that have been reported increasingly worldwide with higher frequency in young women and children.[3,4] The clinical picture is characterized by the presence of unilateral multiple cervical lymphadenopathy, with fever, malaise, weight loss, macular skin rash, arthralgia, myalgia, vomiting, diarrhea, hepatomegaly and splenomegaly. Laboratory findings include leukopenia, elevated erythrocyte sedimentation rate (ESR) and anemia.[3,4] Clinically, KFD can be mistaken for lymphoma or systemic lupus erythematosus (SLE), therefore the diagnosis should be confirmed by biopsy study. The involvement of the central nervous system (CNS) is not common. We report this case because it displayed an unusual association between KFD and temporal lobe encephalitis.

CASE REPORT

A 19-year-old Hispanic woman, with a history of 2 months of severe and persistent headache, fever, cervical pain, malaise and joint pain was admitted in her local hospital with the presumed diagnosis of infectious mononucleosis. She had no previous story of drug use, cat scratches, or contact with sick individuals. Physical examination showed unilateral posterior cervical lymphadenopathy, pharyngitis and a rash located in chest and abdomen. Laboratory findings included white blood cell counts of 3,000 per/mm^3 and ERS of 19 mm/h. Acute antibody titers against Toxoplasma gondii, and Epstein-Barr virus were negative. Blood cultures was also negative. A chest radiograph was normal. The lymph node biopsy showed enlargement of the node with areas of necrosis associated to fibrosis and significant histiocytic infiltrate and absence of neutrophils suggestive of KFD. The patient was treated with betametasone for several days and discharged from that hospital. One month later her general condition worsened and she was admitted into our hospital with confusion and intermittent periods of agitation. She also had one episode of generalized tonic-clonic seizure, fever, cervical lymphadenopathy, hemorrhagic conjunctivitis and hepatosplenomegaly. The neurological examination revealed a drowsy patient with impairment of attention and memory, bilateral papilloedema, and signs of meningeal irritation. There was no abnormality of language, cranial nerves or motor function. Laboratory investigations showed an elevated ERS of 90 mm/h, anemia and leukopenia. HIV, VDRL, hepatitis B and C, Anti-DNA assays and antinuclear antibody test were negative. Coombs test and C3 and C4 were also normal. Lumbar puncture revealed glucose of 47mg/dL, protein 140mg/dL and 38 white blood cells (75% monocytes) in the cerebrospinal fluid (CSF). EEG examination was normal. MRI Brain showed well defined areas of increased FLAIR-and T2-signal intensity in grey

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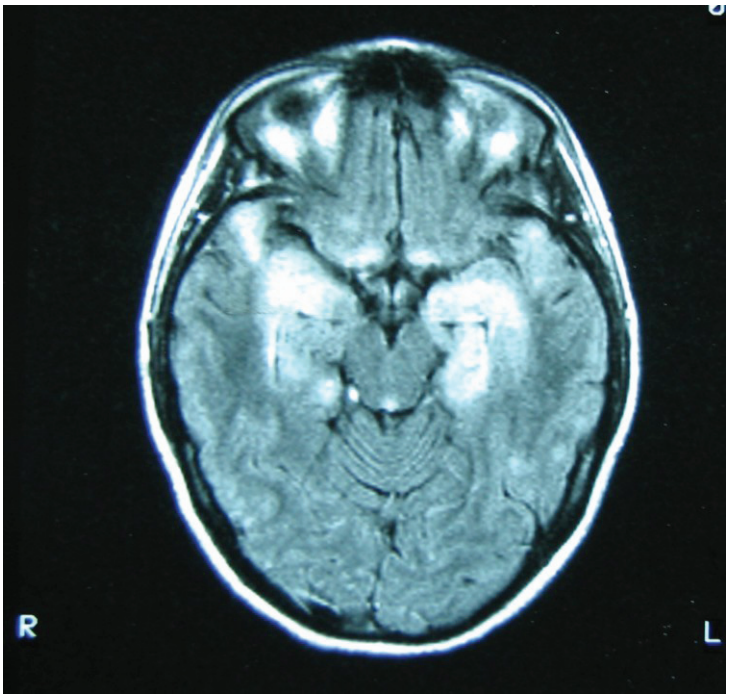

Figure 1: Initial MRI in FLAIR showed increased hyperintense signals in the medial part of both temporal lobe, insula and hippocampus.

and white cortex in the medial an anterior part of both temporal lobes, insula, hippocampus, and caudate nucleus, with enhancement after gadolinium contrast (Figure 1). As the diagnosis of herpetic encephalitis was suspected, a sample of CSF was sent for herpes simplex virus-1 and-2 test by polymerase chain reaction (PCR) study. She was also treated with acyclovir 500mg/TID for 10 days. There was no improvement clinically or in the CSF examination. After completion of acyclovir, the CSF showed glucose 0.71mg/dL, protein 0.70mg/dL and 65 white blood cells, 100% monocytes and herpes virus PCR was negative. She was then given intravenous pulse methylprednisolone 1g/d for 10 days. She improved after a few days, clinically as well as in CSF examination. The repeat CSF examination showed glucose of 0.54mg/dL, protein 0.54mg/dL and 4 white blood cells. Brain MRI repeated after 2 weeks demonstrated significant resolution of the previously noted abnormalities especially those in the temporal lobes. The cervical node biopsy was obtained from the local hospital for review and complementary study. Immunohistochemistry technique showed a polyclonal kappa and lambda pattern and positive alfa-1-antiquimiotripsina in numerous histiocytic cells associated with a high degree of immunoreactivity. The immunostain for Epstein-Barr virus was negative.

DISCUSSION

KFD has a benign course with remission spontaneously or when treated with steroids. Nevertheless, fatal outcome have been reported in a few patients.[5] The cause of KFD is unknown but the most favored theory is a viral infection followed by an autoimmune process in patients with a genetic predisposition.[6] The involvement of CNS is very rare. In a review of the literature the most reported neurological picture associated with KFD was aseptic meningitis, usually with good outcome.[7-9] Only, one case of encephalitis has been reported[10] in a young woman who developed clinical and MRI features of brainstem encephalitis 35 days after onset of fever and cervical lymph node enlargement. Our patient presented about 3 months after a febrile illness with encephalitis, predominantly affecting both temporal lobes, with mild disability on last follow up. The latency between the onset of cervical lymphadenitis and limbic encephalitis in our patient is consistent with a post-infectious or autoimmune process.

In conclusion, KFD is usually regarded as a benign or self-limiting disorder. Our patient, and a review of literature has demonstrated that it can involve the CNS, especially for patients with long-standing evolution.

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