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CASE REPORT

BRAINSTEM ENCEPHALITIS WITH KIKUCHI-FUJIMOTO DISEASE

Saad Shafqat, Saima Bano Memon, Shiraz Hyder, Sheema H. Hasan* and Raymond A. Smego Jr**

ABSTRACT

A case of young woman is described who developed clinical and MRI features of brainstem encephalitis in the setting of fever and cervical lymphadenopathy. Lymph node biopsy revealed histiocytic necrotizing lymphadenitis (Kikuchi-Fujimoto disease), which may reflect host response to an unspecified immune insult.

KEY WORDS: Brainstem. Encephalitis. Lymphadenitis. Lymphadenopathy.

INTRODUCTION

Selective pathologic involvement of the brainstem produces an uncommon but distinct clinical syndrome characterized by a variable combination of ophthalmoplegia and ataxia, with or without long-tract signs. Originally considered an infective or inflammatory entity,¹ the syndrome is traditionally referred to as 'brainstem encephalitis' even though its true pathophysiological spectrum remains unclear. Accumulating data from neuroimaging studies, autoantibody serology, bacteriology and virology have identified a range of factors that may be etiologically related to brainstem encephalitis, including infectious, non-infectious (autoimmune), and post-infectious causes. Common to all these processes is focal affinity for a specific neural substrate, which may reflect targeted immune attack on brainstem structures. We report a patient who developed brainstem encephalitis concurrent with histiocytic necrotizing lymphadenitis (Kikuchi-Fujimoto disease).² While true incidence figures are unknown, both brainstem encephalitis and Kikuchi-Fujimoto disease are considered uncommon, although not rare.

CASE REPORT

A 23-year-old woman presented with a 35-day history of fever and cervical lymph node enlargement despite several courses of oral antibiotics, including 4-drug antituberculous therapy. The fever was noted daily and was mostly low-grade, with occasional spikes to 102°F. Lymphadenopathy had been noted concurrent with the fever, and comprised several soft, mobile and mildly tender nodes upto 2 cm in diameter, palpable in the posterior cervical region bilaterally. Lymph node biopsy revealed necrotizing lymphadenitis with histiocytic infiltration consistent with Kikuchi-Fujimoto disease (Figure 1A). All antimicrobials were discontinued and she was treated with antipyretics alone, but the fever persisted and the patient was admitted for further work-up.

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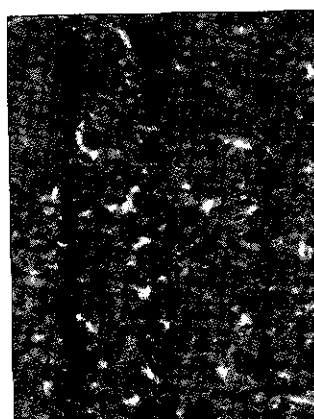


Figure 1: Histopathology and MRI findings of the patients.

A: Photomicrograph of lymph node biopsy showing a necrotic focus devoid of neutrophils but with karyorrhectic debris and infiltration of mononuclear histiocytic cells, consistent with Kikuchi-Fujimoto disease (H & E, 40X).



Figure 1B: Initial T2-weighted MRI scan revealing circumscribed hyperintense signals in the pons and diffusely hyperintense signal in the middle cerebellar peduncles bilaterally.

On the second day of admission she developed numbness on the right side of her body and became increasingly somnolent. On examination she was drowsy but could be easily roused to make appropriate conversation. There was no nystagmus and no cranial neuropathies, including normal pupillary responses and eye movements. There was a mild right-sided hemiparesis with an ipsilateral upgoing plantar response. Sensory examination was normal. The gait was broad-based with swaying to the right, and right-sided appendicular ataxia (upper limb greater than lower limb) was noted.

MRI brain revealed two relatively well-defined areas of increased FLAIR- and T2-signal intensity in the basis pontis, as well as similar ill-defined signal change in the pontine tegmentum that extended into the midbrain and the middle and superior cerebellar peduncles (Figure 1B). Mild hydrocephalus was also present. There were no abnormalities in the cerebral hemispheres. Lumbar puncture revealed white blood cells 2 /mm³, protein 96 mg/dL, and glucose 13 mg/dL. Gram stain, fungal and acid-fast smears and cultures were

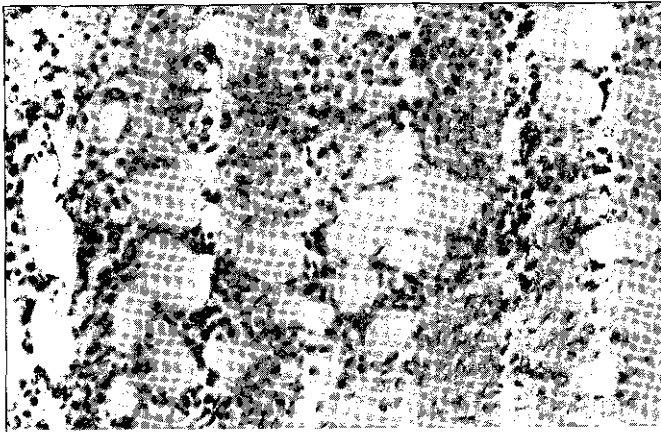


Figure 3: Histological section of mesenteric fat showing fibrosis and infiltration by chronic inflammatory cells. (Hematoxylin and eosin. Original magnification x 200).

bowel wall and mesenteric lymph nodes were unremarkable. His follow-up in outpatient, over the last 6 months, has been unremarkable.

DISCUSSION

Jura was first to describe a case of 'retractile mesenteritis' in 1942.^{2,3,5} Since that time the entity has been reported by various names,^{1,2} including mesenteric lipodystrophy, mesenteric fibromatosis, sclerosing mesenteritis, lipogranuloma of the mesentery, retroperitoneal xanthogranuloma and mesenteric Weber-Christian disease. Present knowledge suggests that these terms represent part of spectrum of pathological changes of the same disease process.^{1,5}

Remmele *et al.*⁶ have differentiated between primary and secondary MP, the latter having recognized associated conditions like pancreatitis. Primary MP has unknown etiology and pathogenesis. Previous abdominal surgery, infection, ischemia, autoimmune diseases have all been implicated as causative agents^{1,3,5} without any definite evidence.

Although it is common in the middle aged with male predominance (male-to-female ratio 1.8:1), cases in children have also been reported.^{1,2} Small bowel mesentery involvement is more common than sigmoid mesocolon involvement.^{1,5}

The histological endpoint is infiltration of mesenteric fat by lymphocytes, plasma cells, eosinophils and fibrous tissue.⁷ The process may extend into submucosa of intestine, obstructing mesenteric lymphatics with submucosal edema.⁷ The mucosa, however, remains intact. The pathogenesis of intestinal obstruction is thought to be kinking of bowel due to rigid mesentery and narrowing of lumen due to submucosal edema.⁷

The outcome depends upon the degree of inflammation and fibrosis. Cases with predominant inflammation usually resolve and recover completely while those with predominant fibrosis may get complicated by intestinal obstruction or venous thrombosis.³

Clinical presentation of MP varies. This includes incidental findings on radiologic investigation of unrelated pathology, abdominal pain,¹ weight loss, cachexia, ascites,⁴ fever of unknown origin, hemoperitoneum,² pulsatile abdominal mass¹ or intestinal obstruction.

Routine laboratory investigations are non-specific. Plain radiographs may show dystrophic calcification.³ Barium studies may show distortion of bowel loops or narrowing of adjacent bowel with no mucosal abnormality.⁵ Colonoscopy is helpful in cases of sigmoid mesocolon involvement and reveals normal mucosa and luminal narrowing, if present.⁵ CT scan and magnetic resonance imaging (MRI) are non-specific and show soft tissue masses.⁷ These two radiological investigations are important to rule out other diagnoses and in follow-up of cases that are managed conservatively. Color Doppler ultrasound demonstrates fine vessels and may help in performing a safe needle biopsy.⁸

The differential diagnosis of MP includes lymphoma, liposarcoma, desmoid, and metastatic neoplasm. Histological examination is required to differentiate these conditions.¹ Multiple biopsies are recommended. Some advocate CT-guided FNA.⁹ However, because of its rarity FNA may not be conclusive.¹⁰ Surgery, open or laparoscopic, plays an important role in establishing diagnosis.

Management of MP depends upon its clinical presentation. As it runs a self-limiting course and recurrence is rare, conservative approach should be taken, however, cases complicated by obstruction require resection or bypass. Cases presenting with abdominal pain and managed conservatively or by bypass of bowel usually continue to experience bothersome symptoms.^{3,7}

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