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Letter to the Editor

Behçet's Disease Long Time after Sulfur Mustard Poisoning

Dear Editor,

A 35-year-old man presented with severe headache and left sided hemiparesis since one day prior to admission. He reported frequent bouts of oral and genital aphthous ulcers, an attack of deep venous thrombosis of left lower extremity, one episode of right knee arthritis, and several attacks of headache, neck rigidity, and fever. He had been a victim of mustard-gas poisoning at the time of Iran-Iraq war (14 years before). The symptoms of poisoning were tearing, photophobia, dyspnea, nausea, vomiting, anorexia, and papules and pustules on legs that were the sites of exposure to the gas. These symptoms had improved after about 2 months but he experienced reactivation of skin symptoms in the same parts of the body in the following years.

In physical examination, he was afebrile. He had multiple oral aphthous ulcers and two scrotal ulcers. In neurological examination, he was drowsy and confused. He had neck rigidity and weakness of muscles in the left side of face, arm, and leg associated with Hoffman and Babinski signs in the same side. Deep tendon reflexes were ½ in both sides.

Routine laboratories and vasculitic profiles were all negative but pathergy skin reaction was positive. The brain magnetic resonance imaging (MRI) showed an area of increased signal intensity in the right basal ganglia and right cerebral peduncle in T_2-weighted and fluid-attenuated inversion recovery (FLAIR) images. Smaller similar changes were also seen in the left basal ganglia. Some patches of enhancement were demonstrated after gadolinium administration in the right basal ganglia. Cerebrospinal fluid analysis showed mononuclear pleocytosis but normal protein and glucose.

The patient revealed a dramatic response to pulse dose (1000 mg) of intravenous methylprednisolone and discharged with oral prednisolone, azathioprine, colchicines and aspirin. Ten months later, he presented with confusion, spastic quadriparesis and urinary incontinence. New MRI revealed new large confluent hyper-signal lesions in thalami, basal ganglia, and midbrain of both sides. Intravenous and oral corticosteroids and monthly intravenous cyclophosphamide were prescribed with unsatisfactory response. Two months later, he passed away with a fatal pneumonia.

In autopsy, major microscopic pathologic findings in cerebral structures were moderate brain edema and margination and permeation of capillaries and venules by mononuclear inflammatory cells (mainly mature lymphocytes) most prominently in basal ganglia, midbrain, and thalamic regions (figure). Informed consent was taken from his family for this report. The patient fulfilled ISG criteria for Behçet's disease and his histopathological findings were consistent with main pathological finding of neuro-Behçet's disease.[1]

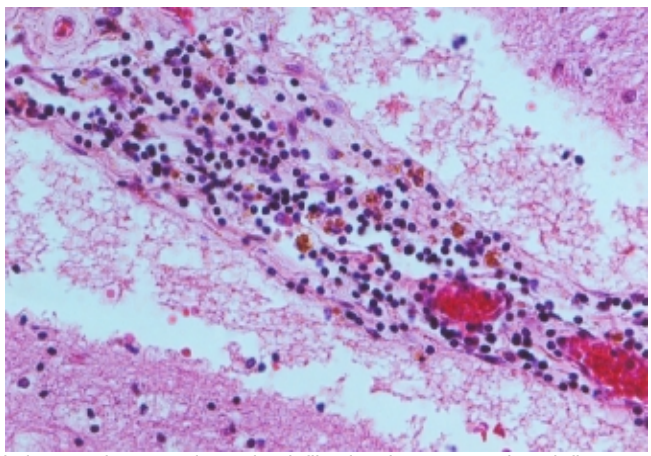

Figure: Section from right thalamus shows perivascular infiltration by mononuclear inflammatory cells (mainly mature lymphocytes) (H&E ×250).

Immunological effects of mustard poisoning long time after exposure have been previously reported in several studies.[2] Diminishment of natural killer cells after exposure to sulphur mustard has

been reported in two Iranian studies.[3,4] It was also shown that exposure to sulphur mustard resulted in impairment of natural killer cells function in Japanese sulphur mustard gas workers.[5] Meanwhile, it was demonstrated that natural killer cell activity in the clinically active stage of Behçet's disease was significantly lower than that of healthy controls and patients in the convalescent stage.[6]

In summary, although we cannot consider this case with pathologically confirmed Behçet's disease and legally proved sulphur mustard poisoning more than a coincidence but we can justify this association with the role of natural killer cells.

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