Intracerebral hemorrhages in Vogt-Koyanagi-Harada disease

Sir,

Vogt-Koyanagi-Harada (VKH) disease is a multisystem disorder characterized by granulomatous panuveitis with exudative retinal detachments often associated with neurologic, acoustic and cutaneous manifestations.[1] The neurological features of VKH disease include headache, meningismus, cranial nerve palsies, hemiparesis, transverse myelitis and ciliary ganglionitis.[2] Intracerebral hemorrhage has not been described earlier. We describe a male patient with VKH disease who presented with ataxia secondary to intracerebral hemorrhage.

A 26-year-old gentleman presented with acute onset headache, vomiting, imbalance while walking and incoordination of the upper limbs. There was no history of weakness, sensory impairment or any cranial nerve deficit. He gave a history of recurrent nontraumatic uveitis involving the right eye for the last 2 years. His first evaluation 2 weeks into the illness showed reduced visual acuity (20/200) in the right eye, the left eye being normal (20/20). The anterior chamber showed flare with hazy media. Evaluation for sarcoid, tuberculosis, syphilis and vasculitis was negative. He was initiated on steroids, with total improvement over 4 weeks. He later discontinued steroids due to weight gain. One month after stopping steroids, he developed redness of the same eye with visual deterioration. Evaluation showed exudative retinal detachment. Eventually, he developed complicated cataract and prephthisical changes. He also had two episodes of low-grade fever with headache and terminal neck stiffness without altered sensorium or focal deficits and was treated symptomatically. Examination at the present admission revealed patches of vitiligo on his back. Visual acuity was 20/20 in the left eye and no perception of light in the right eye. Ophthalmic evaluation showed phthisis bulbi and complicated cataract in the right eye [Figure 1]. The left eye showed optic disc edema, tortuous veins and choroidal folds [Figure 2]. Neurological examination showed gaze-
all patients with uveitis. VKH is related to an aberrant T cell-mediated immune response directed against self-antigens found on melanocytes. Yamaki and coworkers have shown that the tyrosinase-related proteins, TRP1 and TRP2, can induce disease in Lewis rats that is similar to VKH disease in humans. Thus, any organ containing melanocytes or tyrosinase can be affected in VKH disease. The International Committee on Nomenclature has established revised criteria for the diagnosis of VKH disease. Our patient satisfied the complete diagnostic criteria for VKH. A literature search reveals infrequent case reports of neurological complications. An early case report of VKH with vestibular and cerebellar ataxia and another with multiple cranial nerve palsies has been reported, which improved with steroid therapy, but imaging was not performed. Osaki et al. have reported the case of a 57-year-old Japanese male with VKH who presented with truncal ataxia in whom MRI revealed cerebellar and basifrontal region enhancement.

Cerebrospinal fluid (CSF) showed pleocytosis 90 cells/μl (100% lymphocytes) with normal sugar (CSF: Plasma glucose 0.65) and mildly elevated protein (64 mg/dl). Audiometry showed bilateral mild sensorineural hearing loss. Ultrasonography B-mode scan of the right eye showed cone-shaped retinal detachment with subretinal hemorrhage. Biopsy from the eye was withheld because of risk of sympathetic ophthalmitis. Magnetic resonance imaging (MRI) of the brain [Figure 3] showed bilateral cerebellar, vermian and basifrontal bleed with mild leptomeningeal enhancement on postcontrast T1-weighted images. Four-vessel digital subtraction angiogram was normal. He responded excellently to a course of intravenous steroids and was continued on oral steroids. On discharge, he could walk normally and was asymptomatic at the 3-month follow-up.

VKH syndrome is a rare systemic disease involving melanocyte-containing organs. VKH occurs more commonly in patients with a genetic predisposition to the disease, including Asian, Middle Eastern, Hispanic and Native American populations. The highest prevalence is in Japan, where VKH disease represents 6.8-9.2% of all patients with uveitis. VKH is related to an aberrant T cell-mediated immune response directed against self-antigens found on melanocytes. Yamaki and coworkers have shown that the tyrosinase-related proteins, TRP1 and TRP2, can induce disease in Lewis rats that is similar to VKH disease in humans. Thus, any organ containing melanocytes or tyrosinase can be affected in VKH disease. The International Committee on Nomenclature has established revised criteria for the diagnosis of VKH disease. Our patient satisfied the complete diagnostic criteria for VKH. A literature search reveals infrequent case reports of neurological complications. An early case report of VKH with vestibular and cerebellar ataxia and another with multiple cranial nerve palsies has been reported, which improved with steroid therapy, but imaging was not performed. Osaki et al. have reported the case of a 57-year-old Japanese male with VKH who presented with truncal ataxia in whom MRI revealed cerebellar vermian contrast enhancement.

Our patient had multiple areas of parenchymal bleed confirmed by SWI and leptomeningeal enhancement on postcontrast MRI. Intracerebral bleed in our case might have resulted from T cell-mediated cytotoxicity along with humoral autoimmune response directed against tyrosinase-related proteins found in the neurons. Immune-mediated damage breaks the blood

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**Figure 3:** 1.5 Tesla magnetic resonance imaging of the brain. Axial flair images (a-d) show hyperintensities involving bilateral cerebellar hemispheres, vermian, basifrontal region and genu of corpus callosum (black arrows). SWI axial images (b-e) show susceptibility effects in the right cerebellar hemispheres, vermian and basifrontal region (white arrows). Postcontrast T1-weighted axial images (c-f) show leptomeningeal enhancement of the right cerebellar and basifrontal region (black arrowheads).
brain barrier, which manifested as leptomeningeal enhancement. The fact that he made excellent recovery to immunomodulatory therapy highlights the need for prompt recognition and management to avoid morbidity.

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References


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Intravascular lymphomatosis with recurrent cerebral hemorrhages

Sir,

Intravascular lymphomatosis (IL) is a rare lymphoproliferative disorder characterized by proliferation of neoplastic lymphocytes within vessel lumen with no or minimal parenchymal involvement. The central nervous system and skin are the most common sites, however, rarely other organ involvement has also been described.[1-6] We report a patient with IL presenting with focal cerebral mass lesion and recurrent cerebral hemorrhage.

A 47-year-old male presented with two episodes of focal onset tonic-clonic seizures with secondary generalization in the one week before admission. On admission, general physical and neurological examination were normal, except for moderate anterograde amnesia. Laboratory investigation showed: Elevated erythrocyte sedimentation rate (58 mm1h); elevated serum lactic dehydrogenase level (1821 U/L); elevated C reactive protein level (15.3 mg/dl); low thrombocyte count (89-94 10^9/L); normal total and differential leukocyte counts; and normal prothrombin and partial thromboplastin time. Serological test for HIV was negative. Tumor markers including CA199, CA125, NSE, CEA, AFP were all within the normal range. Cerebrospinal fluid (CSF) opening pressure was 80 mmH^2O, cell count was 450 cells 10^6/L and 10^6 leukocytes 10^6/L, protein was 0.628 g/L and sugar was normal. Cytology for malignant cells was negative. Ultrasound of the abdomen was normal and computed tomography (CT) scan of chest and abdomen were normal. Non-contrast magnetic resonance imaging (MRI) scan demonstrated an abnormal low-signal area of 1.5 3 3 cm in the right frontal lobe. Contrast study showed nodular enhancement of the lesion with mild surrounding edema [Figure 1]. He had recurrence of seizures on day nine of admission and non-contrast MRI done on the day of third seizure revealed hemorrhage in the previous right frontal lobe and also a new hemorrhagic lesion in the left temporal lobe [Figure 2]. Patient was treated with antiepileptic medication and corticosteroids. He continued to have recurrent...