Recurrence of cluster headaches presenting with a virtually painless Horner’s syndrome

Throughout every winter from the ages of 25 to 33 a 56 year old property developer had had monthly bouts of daily pain, each bout lasting one to two weeks. During these he would experience up to five extremely severe headaches each evening between 6.00 pm and 7.00 am, of which two or three would wake him from sleep. The pain would last 45 minutes, was situated above and around the right eye, and was associated with nasal stuffiness and watering of the eye. These pains had remained in remission for 23 years.

He was then referred urgently because of the development of a right Horner’s syndrome associated with some burning over and around the right eye. Nine days previously he had some influenza-like symptoms, waking the next morning with vomiting and vertigo. This soon settled, although he continued to experience transient vertigo when he lay with his left ear downwards. When first seen he had a right Horner’s syndrome with no demonstrable sensory loss or other neurological signs. A carotid Doppler study and CT and MRI were normal.

On the evening after his CT the pains restarted, following exactly the same pattern as before. The Horner’s syndrome had been resolving but his wife reported that it became more prominent in the attacks of pain. When seen three weeks later he was given prednisolone (40 mg daily) for a week, declining over the second week, and his headaches resolved within three days.

Miosis is a common feature of attacks of cluster headaches, and can often persist after the bout is over, and occasionally permanently. It is unusual for a patient to develop such an unequivocal Horner’s syndrome with only minimal facial burning discomfort. The vomiting and vertigo, both spontaneous and positional, were considered suggestive of a viral vestibular neuritis, particularly as no radiological evidence of a brain stem infarct could be obtained.
Optic neuritis in cerebral toxocariasis

Optic neuritis occurs in an isolated manner or in the presence of multiple sclerosis. Rarely optic neuritis has been described in association with a nematode infection.1 All of the documented lesions to date involve the optic nerve head, resulting from a direct intraocular infection. This article presents the first demonstration of retrobulbar optic neuritis verified by MRI in a patient with eosinophilic meningoencephalomyelitis due to *Toxocara canis*.

A 21-year-old woman was admitted to our hospital because of headache, low-grade fever, and convulsions. Her illness had begun 4 weeks before admission with a constriciting frontal headache and fever of 37.5°C, followed by several episodes of convulsions two weeks later. She had been exposed to a dog for 8 years, since it was a puppy. General physical examination on admission showed a temperature of 37°C but no evidence of skin rash or hepatomegaly. The mental state of the patient was slightly impaired. Positive findings on neuroanatomical examination included evidence of meningeal irritation such as neck stiffness and Kernig's sign. Cerebellar ataxia, with a predilection for the right side, was also noted. Complete ophthalmological examination, including examination of visual acuity, the anterior segment of the eye, and retinal, yielded normal results.

Laboratory studies showed a slight leukocytosis (9100/μl) with an increased number of eosinophils (23%). Her CSF contained 330 leucocytes/μl with 30% of eosinophils, and a protein concentration of 55 mg/dl. Synthesis of IgG was increased at 43.6 mg/day. Cultures of blood, urine, and CSF, and multiple examinations of stool for ova and parasites were all negative. Results of CT and MRI (1.5T unit) on admission were unremarkable. Indirect immunofluorescence tests, with embryonated *Toxocara canis* eggs, were positive for both serum and CSF. We also performed an immunoblotting assay (IBA) and an enzyme linked immunosorbent assay (ELISA) with the secretory products of second-stage larvae of *Toxocara canis* as an antigen according to methods described previously.7 Both the IBA and ELISA yielded positive results. The ELISA values for *Toxocara canis* in this patient were 1:687 in serum and 0:049 in CSF, whereas those in controls were 1:050 (0:340) (mean SD, n = 250, 0:025 (0:001) (n = 10), respectively. By contrast, ELISA tests for antibodies to *Angiostrongylus cantonensis*, *Anisakis*, *Dipylidium caninum*, *Spriorneta erinacei*, and *Trichinella spiralis* were negative.

Despite treatment with diethylcarbamazine (300 mg/day for 8 weeks) and prednisolone (40 mg/day), leg spasticity, sensory impairment below the level of C4, and Lhermitte's sign developed. MRI performed 4 weeks after admission showed lesions located mainly in cortical or subcortical layers of cerebrum, the cerebellum, and the upper cervical spinal cord. These lesions had a hyperintense appearance on T2-weighted images and were enhanced with gadolinium.

Ten weeks after admission, the patient began to complain of blurred vision in the upper visual field of the right eye and pain behind the affected eye with attempted eye movement. An ophthalmological examination showed a reduced visual acuity of light perception OD, a right relative afferent pupillary defect, and a normal optic disc, indicating retrobulbar optic neuritis. Repeat examination of her CSF showed 19 leucocytes/μl without eosinophils, 52 mg/dl protein, and IgG synthesis of 17.5 mg/day. In addition, the occurrence of the right frontal lobe lesion, CT and MRI disclosed swelling and a gadolinium enhanced lesion of the right optic nerve (figure) respectively. Brain biopsy of the frontal lobe lesion failed to find the worm but showed the accumulation of inflammatory cells around the vessels. Treatment with intravenous methylprednisolone (1000 mg for 3 days) and cyclosporin (4 mg/kg/twice daily) failed to lead to the recovery of her visual acuity. Two weeks later, mild oedema of the optic disc with minimal hyperaemia became evident in the right eye. Regardless of several sub-Tenon's betamethasone injections (5 mg) at this stage, the patient developed optic atrophy of the right eye and her visual acuity remained reduced at 20/60 OD.

Twelve weeks after the initial attack of optic neuritis, the patient developed retrobulbar pain and loss of vision in her opposite left eye. Swelling and a gadolinium enhanced lesion was again evident in the left optic nerve with MRI, whereas pronounced reduction was noted in size and number of the lesions in the other areas. After the intensive treatment with intravenous methylprednisolone and sub-Tenon's betamethasone, the ocular pain rapidly subsided, but her visual acuity was reduced to 20/200 OS. A repeat assay for anti *Toxocara canis* antibody showed a decrease in titre to 0-739 in serum and 0-032 in CSF. The immunosuppressive drugs were gradually discontinued. Although neurological sequelae, such as diminished visual acuity and cerebellar ataxia, have remained, her neuromuscular follow-up during the past year has been unre- markable, and she is not taking medication.

This is the first report of optic neuritis...