regardless of the cause. Upper spinal manipulation, usually administered in the course of chiropractic treatment, can cause stroke, and abducens paresis may develop if there is brainstem infarction or neck vessel dissection. Since our patient had no signs of brainstem infarction and her neck vessels appeared pristine on MRI, her abducens paresis undoubtedly resulted from the intracranial hypotension per se.

Ophthalmologists evaluating patients with orthostatic headache and ophthalmoplegia of unknown aetiology should consider the diagnosis of intracranial hypotension and inquire about recent chiropractic neck manipulation.

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REFERENCES

Unremitting sympathetic ophthalmia associated with homozygous interleukin-10-1082A single nucleotide polymorphism

Sympathetic ophthalmia (SO) is a rare, bilateral, granulomatous panuveitis following injury to one eye. Inflammation develops after the relative immune privilege of the eye is compromised, causing sensitisation to previously sequestered uveal antigens. Although potentially blinding, the outcome may be favourable if aggressive systemic immunosuppression is initiated early.

While certain genetic variants of the human leucocyte antigen (HLA) genes influence susceptibility to SO, single nucleotide polymorphisms (SNPs) in cytokine genes influence severity of disease. We report a case of SO who progressively deteriorated despite early immunosuppression. She was later found to be homozygous for the interleukin-10 (IL-10)-1082A SNP.

CASE REPORT
A 48-year-old woman presented with a 2-week history of blurred vision and floaters in her left eye, 9 weeks after a right ruptured globe repair. Visual acuities (VAs) were 6/9 left and no perception of light right. Examination of the left eye revealed 3+ anterior chamber cells and fine keratic precipitates, and 3+ vitreous cells. Intraocular pressure was 18 mmHg. Widespread choroiditis, disc swelling and cystoid macular oedema (CMO) were observed on fundoscopy, consistent with SO (fig 1).

She received intravenous methylprednisolone 1 g/day for three days followed by 60 mg oral prednisolone and 200 mg (3.5 mg/kg) cyclosporin A (CSA) per day. Over the next week, her VA improved to 6/6. Prednisolone was reduced to 20 mg and CSA to 150 mg per day. The right eye was enucleated for severe pain in a blind eye: histology showed a choroidal lymphocytic infiltrate, multinucleated giant cells and Dalen–Fuchs nodules, characteristic of SO.

Three weeks later her vision deteriorated to 6/12 with recurrent inflammatory activity and CMO. Fluorescein and indocyanine green angiography showed extensive confluent patches of choroidal non-perfusion with ischaemia throughout the macular region (fig 2A, B). She was admitted for a second course of methylprednisolone 1 g/day for 3 days, her CSA was increased to 200 mg and 1.5 g mycophenolate mofetil daily (MMF) was introduced. Despite remaining on 30 mg prednisolone, her VA continued to deteriorate to counting fingers (CF) over the following 4 weeks.

In the 9 months following this episode, she has remained on triple therapy with a minimum steroid maintenance dose of 15 mg/day, below which she relapses. Her VA has never recovered due to the formation of subretinal macular fibrosis.

Genetic testing at this time revealed that she was negative for HLA DRB1*14 and DQA1*03, but that she was homozygous for the IL-10-1082A SNP and ACC promoter haplotype (IL-10-1082A, −819C, −929C).

An application for biological therapy has been made and is still pending.

COMMENT
HLA alleles, DRB1*14 and DQA1*03, are markers of increased susceptibility to SO. Once established, cytokine gene polymorphisms influence the severity of SO, independent of HLA haplotype. The IL-10-1082A allele and ACC/ATA promoter haplotypes are linked to downregulation of IL-10. Since IL-10 is an anti-inflammatory cytokine, this would be expected to create a pro-inflammatory ocular environment. Furthermore, the IL-10-1082A allele is associated disease recurrence from previously stable disease.
Despite immunosuppressive therapy and the requirement for a higher level of steroid maintenance therapy in SO, this patient, homozygous for IL-10-1082A, continued to deteriorate despite high-dose intravenous methylprednisolone, high doses of oral steroid maintenance therapy and adjunctive CSA and MMF. Although there was room to increase the doses of CSA and MMF, these drugs are known to have perceivable beneficial effects even at relatively low doses which may be titrated to patient response. However, neither drug demonstrated any steroid-sparing effect or additive immunosuppressive effect such that this patient required long-term high doses of maintenance steroid. She clearly illustrates how the determination of genetic risk factors could have assisted in her management, since individuals possessing the IL-10-1082A SNP are more likely to require aggressive immunosuppression from the outset. This has been achieved with high-dose combination therapy, her outcome may have been more favourable.

References

**CASE REPORT**

A 49-year-old woman with a relapse of liver metastases was seen in consultation because of loss of vision in her left eye. She had a history of an excised gastrointestinal mesenteric tumour diagnosed as leiomyosarcoma 3 years previously. One year after surgery, she developed multiple hepatic metastases, which were treated with chemotherapy. Fundus examination showed a circumscribed non-pigmented superotemporal choroidal mass in her left eye. The right eye was normal. Ultrasonography showed a 2.4 mm height solid choroidal mass. A liver biopsy was performed. Histopathological examination revealed a CD117-positive GIST. The patient began therapy with imatinib (Gleevec) 400 mg/day. The choroidal mass remained unchanged until 4 months later, when fundus examination disclosed enlargement of choroidal mass surrounded by a flat retinal detachment (fig 1). Ultrasonography showed a medium to low reflectivity choroidal mass, measuring 4.2 mm in thickness (fig 2). A computed tomography scan of the brain revealed cerebral metastases. Abdominal imaging showed that the liver lesions had increased. She underwent maintenance chemotherapy combined with imatinib therapy. During the following months, eye examinations revealed no change in the tumour, and her clinical condition deteriorated. She died 9 months after initial ophthalmic examination.

**DISCUSSION**

Although uncommon, GIST are the most common mesenchymal tumours of the gastrointestinal tract. Morphologically, they are similar to other benign and malignant smooth muscle and neural stromal neoplasm. They occur in patients aged 40 to 70 years. Approximately 70% of GIST develop in the stomach, 20% in the small intestine, and less than 10% in the oesophagus, colon and rectum. The detection rate of GIST has increased due to the availability of the antibody directed against the CD117 antigen (KIT), a receptor tyrosine kinase encoded by protooncogene C-Kit (also known as KIT). Until the discovery of imatinib, which acts specifically on the growth factor receptor of this tumour, surgery was the only treatment available. Imatinib has improved the prognosis of these tumours. Most of GISTs recurred in the abdominal cavity or may develop metastases into the liver, and rarely to the lungs and bone. Hughes et al reported a patient with systemic and brain metastasis without response after imatinib therapy who also had bilateral intraocular metastasis. In the present case, the growth of the choroidal metastasis was temporally inhibited after imatinib therapy, followed by relapse and death.

**Choroidal metastasis from gastrointestinal stromal tumour: a case report**

Metastatic disease is the most frequent intraocular malignancy in adults. The most common sites of origin are breast and lung. The vast majority of metastatic intraocular tumours are carcinomas, and rarely neuroendocrine tumours and sarcomas. Gastrointestinal stromal tumours (GIST) constitute a distinct group of gastrointestinal tumours arising from the interstitial cells of Cajal, regulators of peristalsis. The development of imatinib mesylate, a receptor tyrosine kinase inhibitor, has made a major impact on the management of advanced GISTs. We report the case of a patient with choroidal and systemic disease from a GIST, who underwent therapy with imatinib. An accurate diagnosis is required since therapy may differ from the most common metastatic carcinomas.

**REFERENCES**


Figure 1 Elevated non-pigmented superotemporal mass with flat retinal detachment.

Figure 2 (A) B-scan ultrasonography showed a solid elevated choroidal mass without choroidal excavation. (B) Standardised A-scan ultrasonography of the tumour revealed medium to low reflectivity.
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