

## Superior orbital fissure syndrome in a latent type 2 diabetic patient

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### SUMMARY

**Although isolated cranial nerve palsies are common in diabetic patients, multiple, simultaneous cranial neuropathies are rare. We describe the second case of a complete superior orbital fissure syndrome including the optic nerve in a middle-aged Papuan man with newly diagnosed diabetes mellitus. The differential diagnosis included septic cavernous sinus thrombosis and Tolosa Hunt syndrome, and management was initially directed at excluding these serious, treatable conditions.**

### Case Report

A Papuan man in his 40s from a local urban village presented to Accident and Emergency at Port Moresby General Hospital with diplopia secondary to a complete left ophthalmoplegia.

He had suffered from a discharging left ear and left hemicranial headache for many months, but 4 days before presentation had complained of left eye pain and noticed increasing diplopia. He had not complained of sweating or vomiting.

He did not have any past history or family history of tuberculosis. He is from a village noted for its high incidence of type 2 diabetes, but did not have any known personal history of diabetes. He was not taking any medication at this time.

On examination, a complete left ophthalmoplegia was noted with normal pupillary reflexes and a partial ptosis. There was a moderate degree of proptosis of the left eye. Visual acuity was normal bilaterally at this time, and the right eye was normal to examination. Fundoscopy revealed some arteriolar narrowing bilaterally but normal discs with no evidence of haemorrhages or of ischaemic or proliferative diabetic retinopathy.



Figure 1. Complete ptosis, four weeks after discharge (photograph published with the permission of the patient).

He did not appear toxic and was afebrile.

Examination of his left ear revealed a chronic exudate with mild inflammation of the external auditory meatus. There was no evidence of surrounding cellulitis, and there was no cervical lymphadenopathy. There was no bruit heard over the orbits. No abnormalities

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were detected on examination of the other cranial nerves, nor was there evidence of a peripheral sensory neuropathy.

It was noted that his random blood glucose was 19.9 mmol/l on admission and he was commenced on glibenclamide at this time. His blood sugar was normal the following morning, and no further monitoring of his blood sugar was possible due to the lack of reagents. Inflammatory markers were within normal limits, including an erythrocyte sedimentation rate. Biochemical parameters of renal and hepatic function were normal, and HIV antibody screening tests were nonreactive.

The main concern on admission was that of either a septic or nonseptic cavernous sinus thrombosis and he was commenced on subcutaneous heparin 10,000 units tds as well as chloramphenicol 1 g qid IV and flucloxacillin 1 g qid IV.

Over the following day, his ptosis became complete and his left pupil was fixed and dilated (Figures 1 and 2). Visual acuity was reduced to counting fingers by the second day after admission and his diplopia resolved. On the fourth day after admission he began to complain of altered sensation in the distribution of the supraorbital nerve, but there were no objective findings. He remained afebrile during his admission.

Following discussion with the radiologists, an ultrasound of the left orbit was performed, which did not suggest any retro-orbital mass. Doppler studies of the orbital veins appeared normal. On the seventh day after admission, the patient raised the necessary funds for a CT scan of the orbits, which showed no evidence of retro-orbital masses or cavernous sinus thrombosis on either horizontal or coronal reconstructions with contrast.

In view of these investigations and given the lack of response to antibiotics and anticoagulation, these were stopped at this time. Cerebrospinal fluid analysis revealed normal protein and glucose concentrations and microscopy was normal. Cryptococcal antigen detection was negative on both serum and CSF. Orbital venography was attempted but cannulation of the left frontal vein was not possible. A carotid angiogram was decided



Figure 2. Complete left ophthalmoplegia with fixed, dilated pupil, four weeks after discharge (photograph published with the permission of the patient).

against in light of the previous normal imaging, the risk involved and the unavailability of the necessary equipment.

He was commenced on prednisolone 40 mg daily and his glibenclamide was changed to mixtard 30/70. However, his ptosis, ophthalmoplegia and visual acuity did not improve over the following week and he was discharged. On review four weeks later, there had been no clinical improvement.

## Discussion

Given the history of chronic ear discharge, our most immediate concern was to exclude a septic cavernous sinus thrombosis and initial management was directed to this diagnosis. His subsequent clinical course, the results of investigations and the lack of response to antibiotics are sufficient to exclude this possibility. A cranial arteritis would be unlikely with a normal erythrocyte sedimentation rate and in view of his age.

We feel that the course of the illness, its lack of response to treatment and the new diagnosis of diabetes mellitus supports the diagnosis of an ischaemic process. This is further supported by the examination on admission at which there was apparent pupillary sparing. Its subsequent progression also correlates with previous studies of the evolution of ischaemic oculomotor palsies, describing a mean duration between onset and maximal weakness of 3.3 days (1).

Isolated cranial nerve palsies, particularly of the oculomotor nerve, facial nerve and abducens nerve as well as posterior ischaemic optic neuropathy are all well described in diabetic patients (2). Simultaneous external optic neuropathy involving the third, fourth and sixth cranial nerves was first reported in 1950 (3) and has subsequently been described both with (4,5) and without (6) pupillary sparing. To our knowledge, this is the second case in the literature of a complete superior orbital fissure syndrome which has included the optic nerve in a latent diabetic patient. In the only previous case, Jabs et al. (7) performed an excisional biopsy of the optic nerve which confirmed the ischaemic aetiology of the lesion.

Treatment of extraocular palsies is usually expectant with 68% improving in 4 weeks, 96% in 8 weeks and all by 12 weeks (1). Failure of clinical improvement after this time should prompt a search for alternative diagnoses. Non-steroidal anti-inflammatory drugs are sometimes used but are of unproven benefit.

The clinicopathological entity of Tolosa Hunt syndrome is also a possibility in this patient. This syndrome, caused by granulomatous inflammation in the region of the anterior cavernous sinus and orbital apex, is characterized by painful palsies affecting the nerves traversing the cavernous sinus. This is most commonly the oculomotor, abducens and trochlear nerves, but more rarely may also involve the ophthalmic division of the trigeminal nerve or the optic nerve. Patients may have spontaneous remissions, but a relapsing, remitting course is most common. A further characteristic of this syndrome is a dramatic response to steroids, usually within 24-48 hours (8), which was not seen in our patient.

Of particular interest are case reports associating Tolosa Hunt syndrome with autoimmune diseases including type 1 diabetes (9,10). Ideally in this patient magnetic resonance imaging may have helped us differentiate between ischaemic and inflammatory aetiologies (11), but this imaging modality is not available in Papua New Guinea.

A final possibility is that of a retro-orbital or cavernous sinus mass or tumour. The limited series of investigations that were available to us were unable to exclude this possibility entirely, but we feel that normal cerebrospinal fluid analysis, CT scan and orbital ultrasound made a significant mass unlikely.

### Conclusion

Multiple, simultaneous ocular nerve palsies are rare and may be caused by local pathology involving the cavernous sinus or orbital apex or more systemic ischaemic processes as seen with diabetes. Clinicians should be aware of the possibility of unrecognized diabetes when considering a presentation with any unexplained cranial neuropathy.

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