plasmic organelles between the various axons. In some axons the mitochondria were arranged peripherally around a core of filaments and microtubules, whereas in others the entire axonal profile was stuffed with mitochondria, dense bodies and filaments. These differences have been reported in lamellated sensory corpuscles in the oral mucosae of the adult cat and miniature pig, and have been correlated with terminal and ultraterminal segments of the central axes respectively. The aggregate was loosely encapsulated by fibroblast-like cells, similar to those that have been described forming a "pseudocapsule" around coiled simple corpuscles in primate skin.

This finding appeared to be incidental to the clinical presentation which was that of a multiple mononeuropathy associated with sarcoidosis. We have not encountered similar structures in any of over 100 other sural nerve biopsies. However, Pacinian corpuscles have been described within the connective tissue associated with human peripheral nerve fibres (Hall, Hughes and Atkinson, unpublished observations). Timofeev's corpuscles, encapsulated sensory receptors similar to, but smaller than, Pacinian corpuscles, have been described as transient structures which occur in close relation to pelvic autonomic nerves and ganglia in late fetal and early post-natal life; their function is unknown. Presumably the corpuscles that we have described are mechanosensitive: their situation within a relatively mobile section of peripheral nerve may therefore be significant.

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Permanent oculomotor palsy with occlusion of the internal carotid artery

Transient palsies of the third, fourth and sixth cranial nerves, as well as retinal ischaemia, have recently been described ipsilateral to occlusion of the internal carotid artery. We report a case of permanent oculomotor palsy occurring in this situation.

A 77 year old right handed white woman was admitted having developed sudden left sided weakness two weeks previously. Two days before admission she experienced supraorbital pain with visual loss in the right eye and abnormal eye movements.

Four months earlier she had developed left hemianesthesia, weakness and dysarthria in a stepwise manner over two weeks. There was occasional jerking of the left hand, she had a homonymous left hemianopia and left sided pyramidal signs. The fundi were normal. Carotid doppler ultrasound was normal. The CT brain scan showed patchy gyral enhancement in the right frontotemporal and parietal lobes, and a diagnosis of ischaemia in the right internal carotid artery territory was made. Over the next three months her neurological disability improved.

There was also a past history of ischaemic heart disease (treated with verapamil, aspirin and dipyridamole) and a five year history of chronic myelomonocytic leukaemia. Occasional myelosuppression was treated with etoposide the last treatment being given two months previously. The antineural factor was known to be weakly positive (speckled) with negative DNA and ENA antibodies. Cryoglobulins and circulating immune complexes. Serum complement levels were normal.

On examination the blood pressure was 150/90, and both carotid arteries were palpable with no bruits. The mental state was normal. Vision was NPL on the right and 6/6 on the left with a left hemianopia. The right globe was mildly injected, there was a complete ptosis on the right, and adduction, up- and downgaze were absent, with intortion on attempted downgaze. Abduction, and movements of the left eye, were normal. The right pupil was dilated and fixed, the left pupil reacting briskly to direct light and to accommodation. The retina was normal on the left but on the right it had the appearance of a central retinal artery occlusion with diffuse retinal oedema sparing the macula, and attenuated vessels. No haemorrhages or emboli were visible and the optic disc was normal. There was also weakness of the lower face on the left, and in the limbs tone was normal but there was no movement of the left arm or leg, together with left hyperreflexia and an extensor plantar response. Finally, there was sensory inattention on the left.

As before, the blood count and film, erythrocyte sedimentation rate and routine biochemistry were normal. The CT scan (fig 1A) now showed a large infarct involving the whole of the right middle cerebral and part of the anterior cerebral territory, with swelling of the hemisphere but no abnormal enhancement. In a selective right carotid angiogram (fig B), the right internal carotid artery was occluded 2 cm from its origin. There was no filling of the ophthalmic artery, nor any intracranial filling from the external carotid circulation. Flow through the ophthalmic veins, and the cavernous sinus itself, were normal.

There was no recovery in the oculo findings during a further month in hospital, although motor function improved slowly.

Thus the patient presented with occlusion of the right internal carotid artery, ischaemic visual loss in the right eye, and contralateral hemiparesis. The close temporal association of a complete right oculomotor palsy with these latter deficits suggests that it too was caused by ischaemia following the carotid occlusion. Indeed, there were no signs of brainstem ischaemia, nor indications of active vasculitis clinically or in laboratory tests. Her chronic leukaemia remained in remission, and the CT and angiographic studies showed no cavernous sinus lesion. Finally, the conscious level was not depressed at any stage, there were no ipsilateral motor deficits, and the vital signs remained stable, arguing
Penicillamine treatment of Wilson’s disease and optic neuropathy

We report a case of optic neuropathy associated with penicillamine treatment of Wilson’s disease.

A twenty six year old woman presented with a one year history of progressive shaking of her hands and four months of shaking of her head. As a result she had to give up her waitress line. There was no relevant family history of neurological or liver disease. She had a history of tachycardia and had taken disopyramide 100 mg three times a day for three years. A mitral regurgitation murmur had been heard in the past.

On examination her pulse was 80/min and regular, and blood pressure was 115/60. There was a mid and late systolic murmur loudest at the left sternal edge. Higher frequencies were intact, but her manner was disinhibited. Visual acuity was 6/9 on the right and 6/12 on the left eye. Bilateral optic disc swelling was noted. Slit lamp examination showed Kayser-Fleischer rings. The other cranial nerves were normal. She had titubation, tremor of the upper limbs, worse on the right and aggragation of normal and increased rigidity of both wrists. Reflexes, power and sensation were normal.

Penicillamine was stopped and pyridoxine 50 mg twice a day was started. Nine days later near vision was 6/36 on both sides. Trientine dihydrochloride 600 mg three times a day was started. Three weeks after stopping penicillamine the patient reported with failing vision. Nine weeks after starting treatment she had developed a “red light” in the centre of both visual fields and then progressive blurring of vision. On examination both optic discs were pale and both pupils reacted sluggishly to light. Corrected visual acuity was 6/24 on the right and 6/18 on the left. Near vision was N18 bilaterally. Visual evoked potentials (VEP) showed latency of 102 ms on the right and 112 ms on the left (Normal <115 ms). Brainstem auditory (BAEP) and somatosensory evoked potentials (SSEP) were bilaterally delayed. The wave form of the BAEPs was small. Electretroretinogram and autoimmun profile were normal.

Penicillamine was stopped and pyridoxine 50 mg twice a day was started. Nine days later near vision was 6/36 on both sides. Penicillamine treatment was continued. On September 29, 2022 by guest. Protected by copyright. http://jnnp.bmj.com/ J Neurol Neurosurg Psychiatry: first published as 10.1136/jnnp.54.8.745 on 1 August 1991. Downloaded from http://jnnp.bmj.com/ on September 28, 2022 by guest. Protected by copyright.

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