Carotid-ophthalmic artery aneurysm masquerading as optic neuritis

Sir: Acute monocular visual loss secondary to optic nerve compression is unusual, but has been associated with aneurysms, tumours, fibrous dysplasia, and sphenoid sinus mucocoeles and pyocoeles.¹⁻⁴ Generally, compressive optic nerve and chiasmal injuries are associated with fluctuating or progressive monocular visual loss, concomitant central scotomas, and contralateral visual field defects.⁵⁻⁹ Significant, spontaneous visual recovery has not been recognised. This case illustrates that compressive optic nerve lesions may clinically mimic optic neuritis.

A 48-year-old woman presented to her ophthalmologist with a complaint of blurred vision of the left eye. Additional symptoms included left orbito-temporal pain that increased with eye movement and brief "flashing light" phenomena. The initial examination revealed an afferent pupillary defect and mildly constricted visual fields by perimetry in the left eye. In the ensuing 24 hours the patient's visual acuity decreased to counting fingers concomitant with the development of hyperemic disc oedema and a large central scotoma by tangent fields. A pattern reversal VEP revealed a prolonged latency of 124 ms (control 110 ms). The visual acuity spontaneously and rapidly improved over one month, was associated with the disappearance of the afferent pupillary defect, and was 6/9 by 6 weeks. The right eye remained normal. Two months after the development of eye symptoms the patient began to complain of left face, arm, and leg numbness. No additional subjective or ob-

jective clinical findings were noted, and the contralateral eye showed no visual field defects. A CT scan revealed a left supraclinoid enhancing lesion suggestive of an aneurysm. Angiography subsequently demonstrated left carotid-ophthalmic and incidental right intracavernous carotid artery aneurysms (fig).

Operative exposure revealed a left carotid-ophthalmic artery aneurysm that arose immediately distal to the origin of the ophthalmic artery, and was compressing and indenting the optic nerve only. The aneurysm was clipped successfully. Post-operative visual field testing revealed a small, superior, relative scotoma. Visual acuity has remained essentially unchanged.

Recovery of visual function after optic nerve decompression is a frequently observed phenomenon. However, recovery during chronic compression is generally incomplete, and is usually fluctuating or protracted.^{5,10-13} Spontaneous visual recovery without optic nerve decompression probably represents remyelination and restoration of central conduction. Partial remyelination of optic nerves has been observed after 5 weeks of chronic compression.¹⁴ Recovery of visual acuity probably parallels remyelination, and coincides with the restoration of central conduction as observed in the demyelinated lesions of cat spinal cords.¹⁵ By the pattern of acute monocular visual loss, remyelination, and rapid, spontaneous recovery of visual acuity, optic nerve compressive lesions masquerade as optic neuritis.

ROBERT A RINGEL
JOHN F BRICK

Department of Neurology,
West Virginia University Medical Center,
Morgantown,
West Virginia

References

- Stern WE, Earnest JT. Intracranial ophthalmic artery aneurysm. *Am J Ophthalmol* 1975;**80**: 203-6.
- Senelick RC, Van Dyk HJL. Chromophobe adenoma masquerading as a corticosteroid-responsive optic neuritis. *Am J Ophthalmol* 1974;**78**:485-8.
- Melen O, Weinberg PE, Kim KS, et al. Fibrous dysplasia of bone with acute visual loss. *Ann Ophthalmol* 1980;**12**:734-9.
- Nugent GR, Sprinkle P, Bloor BM. Sphenoid sinus mucocoeles. *J Neurosurg* 1970;**32**:443-51.
- Meadows SP. Optic nerve compression and its differential diagnosis. *Proc R Soc Med* 1949;**42**:33-50.
- Jefferson G. Compression of the chiasm, optic nerves and optic tracts by intracranial aneurysms. *Brain* 1937;**60**:444-97.
- Jefferson G. Further concerning compression of the optic pathways by intracranial aneurysms. *Clin Neurosurg* 1955;**1**:55-103.

- Peiris JB, Ross Russell RW. Giant aneurysms of the carotid system presenting as a visual field defect. *J Neurol Neurosurg Psychiatry* 1980;**43**:1053-64.
- Walsh FB. Visual field defects due to aneurysms at the circle of Willis. *Arch Ophthalmol* 1964;**71**:49-61.
- Kayan A, Earl CJ. Compressive lesions of the optic nerves and chiasm. *Brain* 1975;**98**:13-28.
- Ferguson GG, Drake CG. Carotid-ophthalmic aneurysms: visual abnormalities in 32 patients and results of treatment. *Surg Neurol* 1981;**16**:1-8.
- Ferguson GG, Drake CG. Carotid-Ophthalmic aneurysms: the surgical management of those cases presenting with compression of the optic nerves and chiasm alone. *Clin Neurosurg* 1980;**27**:263-308.
- Hauser MJ, Gass GH. Optic nerve pressure by aneurysm relieved by decompression of optic nerve. *Arch Ophthalmol* 1952;**48**:627-32.
- Clifford-Jones RE, Landon DN, McDonald WI. Remyelination during optic nerve compression. *J Neurol Sci* 1980;**46**:239-243.
- Smith KJ, Blakemore WF, McDonald WI. The restoration of conduction by central remyelination. *Brain* 1981;**104**:383-404.

Accepted 10 September 1985

Orbital apex syndrome caused by rheumatoid nodules

Sir: Common clinical features of orbital apex syndrome are ptosis, painful ophthalmoplegia, sensory disturbance of distribution of the first division of the trigeminal nerve and visual disturbance with or without exophthalmus in one eye. These symptoms are caused by involvement of the vessels and nerves which pass through the superior orbital fissure and the optic foramen.^{1,2} Various kinds of pathological conditions¹⁻⁵ (trauma, tumours, syphilis, tuberculosis, non-specific local inflammatory processes, infections or mucocoeles spreading from neighbouring structures) manifest as the orbital apex syndrome, or similar syndromes such as the superior orbital fissure syndrome and the Tolosa-Hunt syndrome. In this report, we describe a case of rheumatoid arthritis which showed the orbital apex syndrome during exacerbation of general symptoms of rheumatoid arthritis. This rare neurological complication of rheumatoid arthritis, which was confirmed pathologically, has not previously been reported.

A 58-year-old female suffered from repeated arthralgia and swellings of joints in the feet, hands and extremities for 10 years. She showed typical rheumatoid deformities of joints in the feet and hands on admission to our hospital in September 1975 at the age

Fig Left internal carotid injection demonstrating left carotid-ophthalmic artery aneurysm, (arrow).

of 53. She had received a small dose of corticosteroid (Rinderon; 1 mg daily) for one year before worsening of general joint symptoms caused by rheumatoid arthritis in December 1980, at the age of 58. In January 1981, subcutaneous rheumatoid nodules were noticed in the left forearm and on the occipital scalp. In the middle of January, about one month after the beginning of the exacerbation of general joint symptoms of rheumatoid arthritis, she noticed hypaesthesia of the left upper part of the face and head. On 3 February, she started to complain of diplopia, and ptosis and progressive visual disturbance of the left eye associated with ipsilateral deep eye pain. Neurological examination in mid February revealed involvements of multiple cranial nerves which were restricted to her left side. Her left visual acuity had decreased from 1/2 to 0/1 and she had mild exophthalmus and swelling of conjunctiva of the left eye. Ptosis and total ophthalmoplegia were observed with mydriatic pupil which poorly reacted to light. The area of her sensory disturbance corresponded to the distribution of left ophthalmic branch of the trigeminal nerve. Although her left corneal reflex was preserved, she felt a stimulus on the left cornea as more painful than on the other side. Optic fundi of both eyes were normal. Other cranial nerves and peripheral nerves were spared and no other neurological abnormalities were detected. Although she was in a mildly depressed mood, her mental status was normal during the course of her illness. General physical examination revealed an increased intensity of systolic ejection heart murmur which had been audible before the onset of the exacerbation of general symptoms of rheumatoid arthritis.

The laboratory findings during the exacerbation of general symptoms of rheumatoid arthritis showed positive latex test for rheumatoid arthritis, high titre of rheumatoid factor ($\times 10\,240$), increased erythrocyte sedimentation rate (122 mm/hour), leucocytosis ($11\,500/\text{mm}^3$) and thrombocytosis ($56 \times 10^4/\text{mm}^3$). C-reactive protein was highly positive (5 mm). A serological test for syphilis was negative. Cerebrospinal fluid, which was obtained on 16 February, showed normal pressure and the content of sugar, protein and cell count was normal. Radiographs of skull and orbits were normal, including optic foramen. Repeated CT scans of the head revealed no abnormalities around the orbital apices. At the beginning of March, left cerebral angiography and transjugular cavernous sinography were performed. The left carotid angiogram showed segmental narrowing of the left oph-

thalmic artery in the vicinity of the orbital apex and normal configuration of the carotid artery at the siphon. The cavernous sinus was normal. General symptoms of the joints and the left total ophthalmoplegia began to improve slightly from the end of February but visual disturbance of the right eye with deep eye pain developed about one week before her sudden death on 16 March, 1981.

General findings at necropsy showed ankylosing changes of joints caused by rheumatoid arthritis and multiple rheumatoid nodules at mitral and aortic valves and papillary tendinous tissues in the heart. Partial detachment of the papillary tendons in the left heart ventricle was observed. These cardiac changes, especially the rupture of the papillary tendons in the left heart ventricle, suggested that the cause of her sudden death was due to acute cardiac failure. Microscopic examinations of other organs showed mild pulmonary oedema with pleural effusion and mild congestion of the spleen, liver, kidneys and the alimentary tracts.

Macroscopically, at the base of the skull, the dura covering the left orbital apex was thickened to a few millimetres. No abnormalities were observed in other parts of the dura and around the cavernous sinus. Intracranial portions of cranial nerves were intact. When the sphenoid bones covering both orbital apices were removed, the left orbital apex was apparently swollen compared to the right side owing to underlying pathological changes. Specimens for microscopic examination were taken *en bloc* from both orbital apices. Microscopically, the dura covering the left orbital apex was reactively thickened without forming rheumatoid nodules. Pathological changes were observed in bilateral orbital apices which are shown in microscopic photographs of sections from bilateral orbital apices in a lower-power magnification (fig). In both sections, multiple rheumatoid nodules(*), which fused with each other and were similar to those in the heart, located in the connective tissues between cranial nerves and optic nerve. The total size of these nodules and the degree of cell infiltrations were more prominent in the left orbital apex than those in the right. Most infiltrated cells in the nodules and surrounding connective tissues were small lymphocytes with scattered plasma cells. The centre of the nodules was occupied by necrotic tissue with scattered polymorphic leucocytes. Special stainings for bacilli and fungi in these lesions revealed no organisms. Pale staining of myelin of adjacent cranial nerves (N) to the rheumatoid

nodules indicated a loss of myelin in those nerves. A mild loss of nerve fibres in those nerve fascicles was also observed. The wall of both the ophthalmic arteries (OA), especially the adventitia, was thickened due to increased amount of surrounding connective tissues which narrowed the lumen of both the ophthalmic arteries. The medial and internal walls of these arteries were spared and there were no appearances of primary arteritis. The left optic nerve (ON) was more tightly compacted compared with the right side by surrounding thickened sheath.

No pathological features such as vasculitis were found in the meninges, falx and brain including choroid plexus.

In our case, rheumatoid nodules located in both orbital apices caused the orbital apex syndrome on the left side and visual disturbance on the right side. Although other cranial nerves in the right orbital apex showed similar but mild pathological changes to those observed in the left side, we could not detect signs and symptoms except the second cranial nerve on the right side.

Among various pathological processes described in the literature which involved the superior orbital fissure and the second cranial nerve, inflammatory processes have been the most common.⁴ Corticosteroid responsive painful ophthalmoplegia due to

Fig Sections from the left orbital apex (a) and right orbital apex (b) viewed posteriorly. (Luxsol Fast Blue and Periodic Acid-Schiff. Bar: 1 mm.)

idiopathic granulomatous inflammation of the cavernous sinus/superior orbital fissure has been termed Tolosa-Hunt syndrome.^{5,6} Though the Tolosa-Hunt syndrome is one of the clinical entities caused by cryptogenic granuloma which presents as painful ophthalmoplegia,⁷ the diagnosis of this syndrome is made only after other disease processes (such as that in our case) have been excluded. In this syndrome, only a few cases have been reported which showed positive rheumatoid factor,⁸ LE cell phenomenon⁸ and antinuclear factor⁹ but none of them disclosed symptoms of systemic collagen diseases such as rheumatoid arthritis. Only one of the cases had surgical exploration (case 3 in ref 8) but pathological changes were not detected around and outside the cavernous sinus.

Maier¹⁰ has first reported a case with rheumatoid arthritis in which rheumatoid nodules were found in the cranial dura in addition to the ciliary nerve and extra ocular muscles without cranial nerve involvement. In our case, the cranial dura was thickened near the left orbital apex in the absence of rheumatoid nodules. Lesions due to the rheumatoid nodules were restricted to the bilateral orbital apices without extension into intradural structures such as choroid plexus¹¹ and leptomeninges.¹²

Although we could not find similar cases in the literature, we wish to call attention to the possibility of rheumatoid nodules as a cause of the orbital apex syndrome.

We thank Dr J Nakagawa, Dr H Haibara and Dr H Higashi for providing clinical and pathological investigations.

TETSURO KONISHI,
TAKAHIKO SAIDA,
HIROSHI NISHITANI,
*Department of Neurology,
Utano National Hospital,
8 Ondoyama-cho, Narutaki,
Ukyo-ku Kyoto 616, Japan*

References

- ¹ Lakke JPWF. Superior orbital fissure syndrome: report of a case caused by local pachymeningitis. *Arch Neurol* 1962;7:289-300.
- ² Kander PL, Leaver PK. Orbital apex syndrome complicating frontal pyocoele. *J Laryngol Otol* 1974;88:551-7.
- ³ Hedges TR, Leung LSE. Parasellar and orbital apex syndrome caused by aspergillosis. *Neurology (Minneapolis)* 1976;26:117-20.
- ⁴ Lenzi GL, Fieschi C. Superior orbital fissure syndrome: review of 130 cases. *Eur Neurol* 1977;16:23-30.
- ⁵ Kline LB. The Tolosa-Hunt syndrome. *Surv Ophthalmol* 1982;27:79-95.
- ⁶ Smith JC, Taxdal DSR. Painful ophthalmoplegia: the Tolosa-Hunt syndrome. *Am J*

Ophthalmol 1966;61:1466-72.

⁷ Hunt WE. Tolosa-Hunt syndrome: one case of painful ophthalmoplegia. *J Neurosurg* 1976;44:544-9.

⁸ Mathew NT, Chandy J. Painful ophthalmoplegia. *J Neurol Sci* 1970;11:243-56.

⁹ Lesser R, Jampol LH. Tolosa-Hunt syndrome and antinuclear factor. *Am J Ophthalmol* 1974;77:732-4.

¹⁰ Maier JA. Dural nodules in rheumatoid arthritis: report of a case. *Arch Pathol* 1954;58:354-9.

¹¹ Kim RC. Rheumatoid disease with encephalopathy. *Ann Neurol* 1980;7:86-91.

¹² Steiner JW, Gelbloom AJ. Intracranial manifestations in two cases of systemic rheumatoid disease. *Arthritis Rheum* 1959;2:537-45.

Intradural dermoid of the cervical spinal cord in a child

Sir: Congenital dermoid cysts are relatively rare tumours, the origin of which is believed to result from retention of elements of cutaneous ectoderm beneath the surface during neural groove closure.^{1,2} Sites of predilection include the lumbosacral spinal region and the posterior fossa.³ We report an infant with a dermoid cyst in the cervical spinal cord. The cervical location is extremely uncommon for this kind of tumour.^{2,4}

A 13-month-old female infant was admitted with a history of delayed motor development and irritability. Pregnancy, delivery and neonatal period were unremarkable. The infant was able to hold her head erect at 3 months, and sat without assistance at 6 months. Subsequently, the parents observed a decrease in the motility of the lower limbs. At 13 months of age the infant could hardly move her legs, being unable to stand. The parents also referred to episodes of irritability together with abnormal twisting movements of the neck, during which she repetitively moved her hands to her neck. On examination, she was in good general condition. A dermal sinus surrounded by a hemangioma was found at the level of the 7th cervical vertebra. The infant showed a mild resistance to neck flexion. Lower limbs were spastic, and a mild amyotrophy was present in both shoulders. Electromyography showed signs of chronic partial denervation bilaterally in the C5 and C6 territories. Median and ulnar nerve conduction studies were normal in both arms. Radiographs of the cervical spine showed laminar defects from C4 to C6, and a widened spinal canal. Myelography showed an enlargement of the cervical spinal cord between C2 and C7 levels, suggestive of an intramedullary expansive lesion (fig). At operation a sinus tract was found to extend from the skin to the spinal cord, terminating in a 1 cm by

1 cm diameter yellowish intramedullary mass. Both, the sinus tract and the mass were dissected and completely removed. The tumour was, histologically, a dermoid cyst. The evolution of the patient has been satisfactory. A year after the intervention, lower limb motility and strength are normal and mild muscle atrophy still remains in both shoulders.

In a wide review of 1234 previously reported spinal cord tumours in children, Dreyer⁵ and Lorenzo *et al*⁶ found 197 tumours situated in the cervical region, but none of them was a dermoid. Reviewing the literature, we have found only three previous reports concerning cervical spinal dermoids in children.⁶⁻⁸ Koos⁶ in a review of spinal cord tumours in children and adolescents, mentioned one dermoid among 78 cervical neoplasms. Takeuchi *et al*,⁷ reviewing 84 reported dermoids, found the case of a 3-year-old child with a dermal sinus and a subdural dermoid situated at C7 level. The child presented with spastic gait and pain, and surgical ex-

Fig Myelogram, antero-posterior projection, showing an enlarged cervical spinal cord.