idiopathic granulomatous inflammation of the cavernous sinus/superior orbital fissure has been termed Tolosa-Hunt syndrome. 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 phenomenon and antinuclear factor 9 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.

Maher 10 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 extraocular muscles without cranial nerve involvement. In our case, the cranial dura was thinned 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 plexus 11 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.

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References


Intramédullary 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. 12 Sites of predilection include the lumbo-sacral 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

Fig Myelogram, antero-posterior projection, showing an enlarged cervical spinal cord.
A case of recurrent idiopathic ophthalmoplegic neuropathy (Miller Fisher syndrome)

Sir: In 1956 Miller Fisher described three cases of an acute neurological illness characterised by total external ophthalmoplegia, severe ataxia and tendon areflexia. This illness is now referred to as the Miller Fisher syndrome and is generally considered to be part of the spectrum of the idiopathic inflammatory polyneuropathies of which the Guillain-Barré syndrome is the most familiar example. Cases of recurrent Guillain-Barré syndrome have been well documented but this is not so for the Miller Fisher syndrome: such a case is reported here.

One week after an upper respiratory tract infection, a 33-year-old solicitor developed mild tingling in his hands and feet together with unsteadiness of gait. Within 24 hours his unsteadiness had deteriorated to such a degree that he was unable to walk, although to him the power in his limbs seemed normal. He also began to experience diplopia on forward gaze, unaccompanied by vertigo, or dysarthria. There was no past medical or family history of relevance. The patient was taking no drugs and had not been exposed to neurotoxic substances. Examination at this time revealed normal cardiovascular and respiratory function; his vital capacity was 2·8 l. On neurological examination he was fully alert. There was weakness of the right lateral rectus muscle but no facial weakness. Tone was reduced in all four limbs and there was mild generalised muscle weakness, most prominent proximally, and ataxia. All tendon reflexes were absent except for the biceps jerks which were both retained. There was no sensory deficit for any modality.

Investigation showed a normal full blood count, plasma electrolytes, serum and urine electrophoresis, liver and thyroid function. A porphyrin screen, VDRL and monospot test were all negative. Lumbar puncture produced clear cerebrospinal fluid (CSF) under normal pressure with protein content of 0·4 g/l; the CSF: blood glucose ratio was normal. No oligoclonal IgG bands were seen on CSF electrophoresis. Upper and lower limb motor and sensory nerve conduction studies were normal, including a median nerve F wave latency of 26 ms (wrist stimulation), although F waves were undetectable in extensor digitorum brevis on stimulation of the deep peroneal nerve at the ankle.

Within 1 week of his initial presentation, the patient’s signs evolved to a complete internal and external ophthalmoplegia, a right lower motor neuron facial weakness and marked ataxia affecting upper and lower limbs. He also developed complete urinary retention requiring bladder catheterisation for 5 days. A repeat CSF sample 3 weeks after the onset of symptoms contained 4·5 lymphocytes/mm³ with no red blood cells and a protein content of 2·1 g/l. Repeat nerve conduction studies again showed normal upper and lower limb motor velocities, including an F wave latency in extensor digitorum brevis of 49 ms with ankle stimulation. Sensory conduction studies revealed minor abnormalities with a right index finger/wrist median sensory action potential of reduced amplitude (4·4 µV) and a latency to peak at the upper limit of the normal range (3·6 ms). A right antidromic sural nerve action potential obtained with needle electrodes was of reduced amplitude (3 µV) with a reduced inflexion velocity of 24 m/s.

The patient began to improve 3 to 4 weeks after the onset of symptoms. He was discharged from hospital 5 weeks after admission and was able to return to work within 3 months. When reviewed in the clinic 6 months after discharge his symptoms had fully recovered but he remained areflexic.

One year later, 5 days following another respiratory tract infection, the patient once again began to experience tingling in his extremities and unsteadiness of gait. The development of a partial internal and external ophthalmoplegia, a slurring dysarthria, hypesthesia and minimal generalised limb weakness, again more marked proximally, followed within 4 days. There was no muscle wasting but the patient remained areflexic.

There was reduction of pin prick sensation in a glove distribution to the wrists but no loss in the lower limbs. Joint position sense was normal in the fingers but significantly impaired in the toes. There was marked ataxia in the upper and lower limbs. Again, the patient remained fully alert throughout his illness.

Investigation on this occasion showed a normal full blood count and biochemical screen. The CSF contained no white cells, with protein and glucose levels of 0·3 g/l and 3·2 mmol/l respectively. Nerve conduction studies revealed normal motor velocities and median and peroneal F wave latencies of 28 ms and 46 ms with stimulation at the wrist and ankle respectively. There were minor abnormalities in sensory conduction similar to those seen during the patient’s first illness. Somatosensory evoked potentials (SSEPs) from the upper limbs showed both a slight peripheral and central (N20) delay in latency. A repeat CSF specimen obtained 4 weeks after the onset of the relapse had a...