A CASE OF SARCOIDOSIS PRESENTING AS CRANIAL POLYNEURITIS

S H Lim, K Puvan, E S T Cheah

ABSTRACT

A Chinese female patient presented with cranial polyneuritis of unknown aetiology. Three years later, the diagnosis became obvious when she developed other features of sarcoidosis. This is the first reported local case of sarcoidosis presenting initially with nervous system involvement. It also highlighted sarcoidosis as a possible aetiology in cases of idiopathic cranial polyneuritis.

Key Words: Sarcoidosis, Neurologic Manifestation, Mutiple Cranial Nerve Palsies, Idiopathic Cranial Polyneuritis.

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INTRODUCTION

The syndrome of multiple cranial neuropathy is periodically encountered in neurological practice. It may be a manifestation of several known disorders, yet many cases remain undiagnosed after thorough investigation. Such cases are lumped together as a entity called "idiopathic cranial polyneuritis" (1). In some only after many years of follow up is an aetiology, such as sarcoidosis, clear.

Sarcoidosis is a multi-system, granulomatous disorder of unknown aetiology, which commonly affects young adults and presents most frequently with bilateral hilar lymphadenopathy, pulmonary infiltrates, skin and eye lesions (2). A diagnosis of sarcoidosis becomes difficult when patients first present with neurological features. Brain biopsy is seldom feasible, and the diagnosis requires a repeated search for systemic disease, exclusion of other causes and long-term follow-up.

Sarcoidosis is rare in Asia with the exception of Japan (3). So far 15 cases of pulmonary sarcoidosis have been reported in Singapore in the last 15 years (4-8), and among them three had concomitant neurological involvement mainly in the form of cranial nerve palsy (ies). This is the first case of sarcoidosis presenting as cranial polyneuritis reported locally.

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CASE REPORT

Mdm YOW, a 50 year old Chinese, presented in July 1984 with slowly progressive diplopia on looking to the left. Five months prior to this, she had one episode of twitching of the right hand, face and leg lasting about 30 seconds without loss of consciousness. There were no fever, joint pain, weight loss, weakness and numbness of all four limbs, visual impairment or other symptoms of brainstem dysfunction. The only significant past medical history was that of a thyroidectomy in 1978. Clinical examination then revealed a well nourished middle aged lady who was afebrile. She had a left abducens nerve palsy. The rest of the neurological examination was normal. General physical examination did not reveal any lymphadenopathy, hepato-slenomegaly, lung , skin or joint lesions.

A CT scan of the brain, with contrast study, was normal. Opening pressure during lumbar puncture was 20 cm of water. Cerebrospinal fluid contained no cells; glucose was 65mg/dl, chloride was 731mmol, protein was 30mg/dl and globulin was negative; microscopic examination for acid-fast bacilli, torula and malignant cells were negative; culture yielded no growth of mycobacteria or other micro-organism and serology for viruses was negative. X-rays of the petrous temporal bone, electroencephalogram, tensilon test and post-nasal biopsy were normal. The oral glucose tolerance test showed mild diabetic changes. Serum total thyroxine was 7.6 ug/dl. Chest X-ray was normal. Blood haemoglobin, platelet, total white and differential counts were normal. She was discharged to be observed as an out-patient. As for the diabetes, she was given dietary advice.

She was readmitted in August 1984 for worsening diplopia and numbness on the left forehead. Significant clinical findings then were that of bilateral abducens nerve palsies (Figure 1, 2) and diminished left corneal reflex. While in the ward, she developed a right lower motor neurone facial nerve palsy and subsequently the left was involved over a period of four days. At the same time, she developed a mild bulbar palsy with dysphagia and dysarthria. Corneal reflexes were diminished bilaterally but the sensation over the face was intact. The left ankle jerk, which was present before, was lost. The rest of the neurological and general physical examination including the lungs, joints, skin and eyes did not reveal any abnormality.

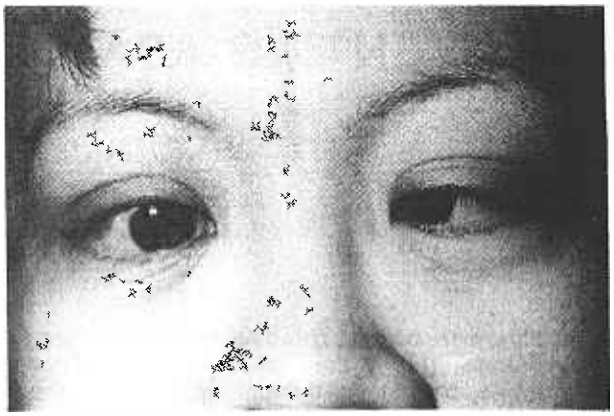

Figure 1:
Right abducens nerve palsy. Note that the patient was unable to abduct the right eye on looking to the right.

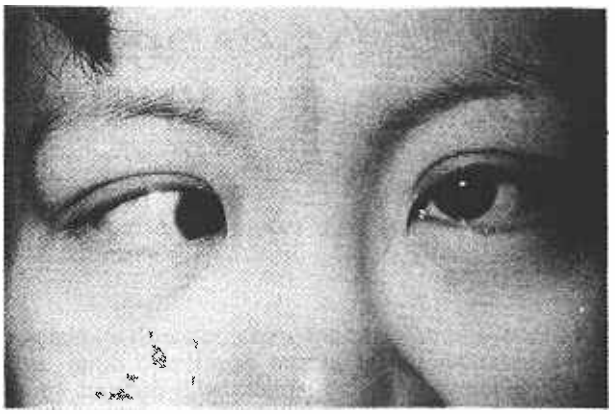

Figure 2:
Left abducens nerve palsy. Note the inability of the left eye to abduct on looking to the left.

A repeat CT scan of high resolution with pontine views was again normal. A second lumbar puncture yielded a CSF that was also normal on microscopy; the sugar, chloride and protein levels were normal; cultures for acid-fast bacilli and torula cultures were negative; malignant cells were absent; IgG was 15 mg/dl but IgA and IgM were not detectable. A repeat tensilon test, brainstem auditory evoked potentials, syphilitic serology and chest X-rays were all normal. The erytrocyte sedimentation rate was 45 mm/hr.

Her symptoms worsened over a month in hospital. The ankle jerks were lost bilaterally. Plantar responses became extensor. However, there were no other long tract signs. Further investigation including nerve conduction studies with the H reflex were normal. A third lumbar puncture was again normal.

She was treated empirically with intramuscular injection of ACTH (40 units every morning). One week later, her dysphagia and dysarthria started to improve. The facial palsies and corneal sensation also improved. She was discharged after two weeks of treatment.

She was reviewed in December 1984 and was found to show minimal diplopia on extreme lateral gaze. The rest of the cranial nerves, the ankle jerks and plantar responses were normal. Serological markers for collagen vascular diseases, including LE cells, antineuclear antibody, rheumatoid factors and anti-ds-DNA were not detected.

She was asymptomatic till February 1987 when she presented to a surgeon with lumps on the left side of the neck. Clinically she had enlarged supra-clavicular lymph nodes. There was no other lymphadenopathy. Lymph node biopsy was performed and the histology showed non-caseating granuloma consistent with sarcoidosis (Fig 3). Fungal culture was negative. Acid fast bacilli were not seen.

She had no other symptoms then. Neurological examination was normal except for the residual diplopia on extreme lateral gaze. Systemic examination, especially the lungs, did not show any abnormality. Chest X-ray showed minimal left hilar prominence but no parenchymal lesion. Lumbar puncture was not repeated as she had no new neurological symptoms and signs. No treatment was initiated and she was followed up regularly.

By September 1987, the CXR showed obvious widening of mediastinum (Fig 4). The CT scan of the thorax confirmed the presence of multiple enlarged lymph nodes in the hilar and right paratracheal regions, Serum calcium was 8.5 mg/dl.

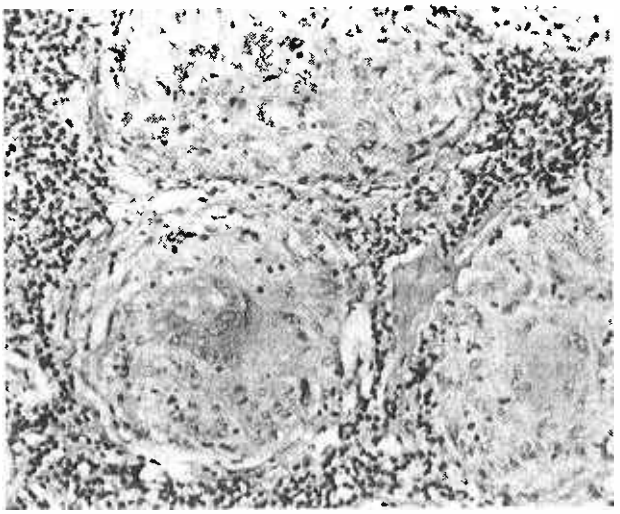

Figure 3:
The lymph node show well defined aggregates of epitheliod histiocytes which are separated from each other by lymphocytes. There are few giant cells. Necrosis is not apparent.

In November 1987, she was still free of respiratory symptoms. There were no rales in the lungs. The rest of the physical examination was normal. Neurological status remained unchanged. However, the CXR (Fig 5) showed worsening with reticular shadowing in both mid-and lower zones. Blood gas was normal. Lung function test showed a restrictive pattern with vital capacity of 1.66 l, functional residual capacity of 1.58 l and total lung capacity of 2.67 l (predicted normal value is 3.57 l). However, the carbon monocide transfer factor was 5.68 ml/min/mm Hg (Normal value is 4.22 ml/min/mm Hg). Transbronchial biopsy was attempted but failed. Nevertheless, she was treated as for pulmonary sarcoidosis with a course of prednisolone.

She remained asymptomatic and free of physical abnormality. When last seen in July 1988, there was no recurrence of cranial nerve palsies or other neurological involvement. Chest X-ray showed much improvement with disappearance of hilar lymphadenopathy and minimal parenchymal infiltrates. The lung function too had improved.

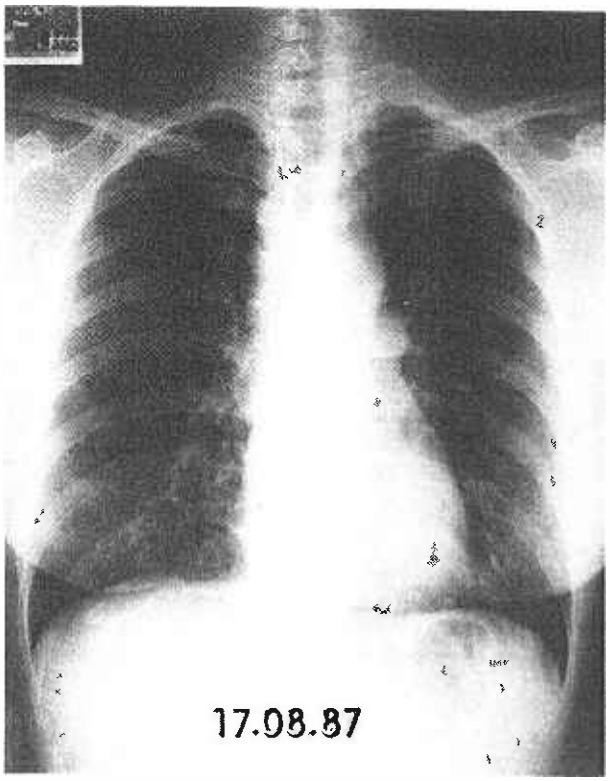

Figure 4:
Chest X ray showed widening of mediastinum.

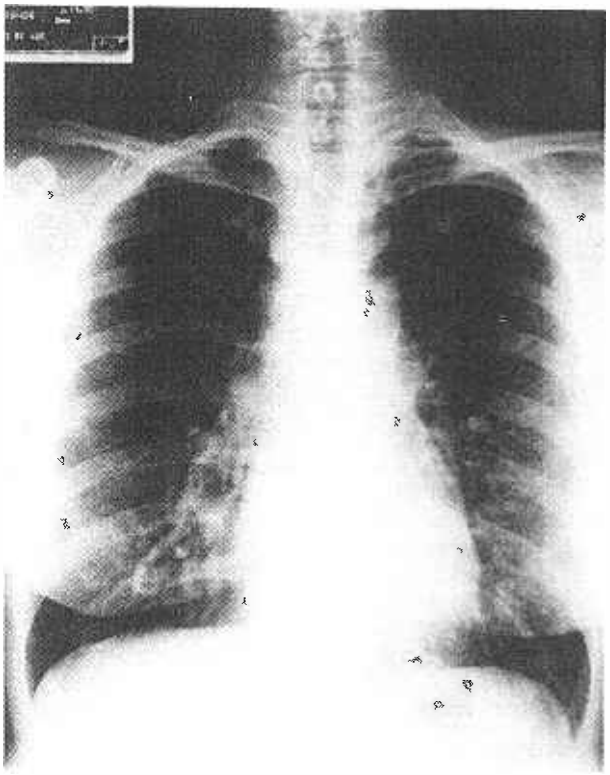

Figure 5:
Chest X ray in November 1987 showed reticulonodular shadowing in both middle and lower lung fields.

DISCUSSION

The syndrome of idiopathic, self limited cranial neuropathy was first recognised by Gower in his textbook as early as 1888. Many of the subsequent reported cases have been from Asia (9-10). Typically, patients present with subacute onset of facial pain preceding the onset of cranial nerve palsies. The cranial nerves most frequently involved are the third and sixth. Motor nerves are affected more than sensory, and the response to steroid treatment are usually dramatic (11). However, it is not known how many of idiopathic cranial polyneuritis could have shown up an etiology such as sarcoid.

Similarly, multiple cranial neuropathies is a remarkable and characteristic presentation of neurosarcoidosis. The onset is often with vague symptoms of ill health or with signs of systemic sarcoidosis. Cranial nerves that are commonly involved are the facial, optic, trigeminal and lower cranial nerves. In contrast, defects of ocular movement are relatively rare. The condition is also benign as most of the cranial nerve palsies recover spontaneously or with steroid (12-17).

Our patient's main presenting clinical manifestation was that of multiple cranial nerve palsies. These developed subacutely and appeared in succession over a period of one month. The involvement of facial, trigeminal, glossopharyngeal and vagus nerves were fairly typical of neurosarcoidosis. The unusual feature here was that of bilateral abducens nerve palsies. An abducens nerve palsy could occur in sarcoid due to raised intracranial pressure in the presence of cerebral sarcoid granuloma with mass effect (14-15, 18). Our patient did not have any evidence of raised intracranial pressure throughout the course of her illness.

In addition to the cranial polyneuropathy, she probably also had transient radiculopathy as evident by loss of the ankle jerks during the acute phase of her illness. Radicular involvement usually accompanied cranial neuropathy but symmetrical polyneuritis is relatively uncommon (14-15).

Our patient also had evidence of central nervous system involvement. The episode of involuntary twitching of right side of the body prior to development of cranial polyneuropathy could well be a focal fits. She later developed extensor plantar responses when her neurological deficits were at the peak. It is not surprising that repeated CT scans of the brain did not show any abnormality because cerebral granulomata are usually small and diffusely scattered. They may remain undetected even with the most sensitive diagnostic method, and yet be demonstrated at autopsy (19-20).

Routine CSF findings are non-specific and in nearly one third of cases are normal (17). Therefore it is not unexpected that our patient's CSF were normal on three occasions. However, serial CSF examination is mandatory to look for other causes of cranial polyneuropathy such as chronic meningitis which are much more common in our local population.

The brainstem auditory evoked potentials (BAEP) in our patient was normal. This did not necessary indicate that her brainstem was not involved. On the other hand, BAEP and visual evoked potientials (VEP) often reveal subclinical lesions in optic pathways and the brainstem of patients with neurosarcoidisis (21). Thus, it seems reasonable to examine BAEPs and VEPs in all patients with sarcoidosis compiaining of vague neurologic and/or ophthalmologic symptoms.

One interesting feature in this case was that she did not have any signs of systemic sarcoidosis at the time of first presentation. It was only 3 years later when other features of sarcoidosis appeared. This is unusual because, almost all patients with neurosarcoidosis had features of systemic sarcoidosis (15-17). Half of these patients had initial neurological manifestation but found to have subclinical systemic involvement. It is more common for neurosarcoidosis to develop after systemic involvement has appeared.

This case emphasises not only the importance of long term follow up in patients with the so-called idiopathic cranial polyneuritis but also a need to consider sarcoidosis in the diagnostic workup in patients presenting with multiple cranial nerve palsies.

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