SYNDROMES OF OPHTHALMOPLEGIA, ATAXIA AND AREFLEXIA — CASE REPORT

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SUMMARY

Two cases of Syndromes of Ophthalmoplegia, Ataxia and Areflexia were reported. A brief discussion on its clinical feature and differential diagnosis was made.

INTRODUCTION

In 1956, Fisher, C.M. reported three patients who developed opthalmoplegia, ataxia and areflexia after minor respiratory tract infection. The cases were remarkable in that they have good prognosis in spite of their "alarming" appearance. The author suggested that it was a varient of the Guillain Barre Syndrome. Since then other reports of similar cases have appeared. The following are two patients seen in the University Hospital, Kuala Lumpur.

Case 1

P.T.H. was a 47 years old Chinese male construction worker from Malacca. He was admitted to the University Hospital on 9.1.81. His main problem was that two weeks before admission, he had an attack of upper respiratory tract infection with running nose, sore throat, cough and mild fever. These symptoms improved. Three days before admission, he developed giddiness which was more on erect posture, unsteadiness of gait, inability to open both his eyelids, regurgitation of fluid through the nose with swallowing, numbness of both hands and feet and progressive weakness of all four limbs more marked in the lower limbs. The patient had past history of bronchial asthma from youth until about ten years ago. He smoked one package of cigarettes daily for many years and was an occasional drinker. There was no other history of significance.

On physical examination, the patient's mental state was normal. He was dysarthric. There was bilateral ptosis and total opthalmoplegia. Bilateral lower motor neurone facial palsy was present which was more marked on the left side. The gag reflex was present. Power in all limbs were normal. The muscle tone was reduced with generalized hyporeflexia. Sensory testing was normal. The patient had past pointing to finger nose test and had dysdiadokokinesia. He was very ataxic on walking. The blood pressure was 130/80 mmHg. Examination of the other systems were normal.

Investigation showed that the tensilon test was negative; full blood count, blood urea, serum electrolyte, serum VDRL, chest x-ray, skull x-ray and nerve conduction studies were all normal. CSF examination showed protein of 26 mg%, there was no cell and organism seen. The patient improved gradually. On discharge six weeks later, he was able to walk normally with normal range of eye movement; but still had mild bilateral facial palsy and generalized hyporeflexia.

Case 2

C.K.C. was a 57 year old Chinese male hawker from Malacca. He was admitted to University Hospital on 11.1.80. His main problem was that ten days prior to admission, he had an attack of upper respiratory tract infection with running nose, sore throat and fever. Two days later, he began to develop giddiness, diplopia, regurgitation of fluid through nose on swallowing, numbness and weakness of both lower limbs and respiratory difficulties. The symptoms progressed over the next few days so that he sought treatment in the University Hospital. He had a past history of haemorrhoidectomy in 1970, peptic ulcer in 1975. He smoked 30-40 cigarettes per day. There was no other history of note.

Examination on admission showed that he was mentally clear. The speech was slurred. There was bilateral total ophthalmoplegia. The pupils were dilated at 10 mm both sides and were sluggishly reactive to light. The fundi were normal. There was bilateral lower motor neurone facial palsy. Other cranial nerves were normal.

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Examination of the limbs showed that the tone was reduced with grade four power of both hip flexors. Other muscles power were normal. There was areflexia of all the tendon jerks with the plantar responses flexor. Sensory examination was completely normal. There was dysdiadokokinesia and past pointing in finger-nose testing. He was confined to bed because of the weakness of the lower limbs. Systemic examination showed a supine blood pressure of 150/110 mmHg. The pulse was regular at 80/min. There was tachypnoea. Other examinations were normal.

Investigations done showed negative tensilon test; normal full blood count, ESR, blood urea and serum electrolyte, urine microscopy, chest x-ray and skull x-ray. The blood VDRL was negative. Serum protein and immunoglobulin electrophoresis were normal. CSF examination showed a protein level of 62 mg% RBC of 10/ul, 4 lymphocytes/ul. The blood gases showed PO_2 of 72 mmHg, PCO_2 of 32 mmHg. Nerve conduction study showed a prolonged motor and sensory distal latencies of the median nerve from the wrist. The patient was started on dexamethasone. Two days later when the power of the lower limbs improved, it was noted that there was marked trunkal ataxia.

Patient's general condition improved rapidly. A week later on discharge he was able to walk unaided. The blood pressure was normal and there was no dyspnoea. However, the ophthalmoplegia and the areflexia was still persistent. The patient did not come back for follow-up.

DISCUSSION

C.M. Fisher in 1956, reported three patients with ophthalmoplegia, hyporeflexia and ataxia following minor respiratory infections. The ophthalmoplegia was often complete, the ataxia was out of proportion to the motor and sensory loss. These patients had good prognosis. Their CSF protein showed delayed rise. Fisher suggested that they were a variant of the L Guallian Barre syndrome and called it the syndrome of ophthalmoplegia, ataxia and areflexia. Other features which were also described include drowsiness but with normal orientation, ptosis, sluggish pupillary reaction, absent bell's phenomenon, no response to caloric irrigation, bilateral facial palsy, trigerminal weakness, parathesia and weakness of the limbs. It was important to recognise this syndrome as the prognosis was good though the patient may appear "alarming." Since then, other reports of the same illness have appeared (Elizan *et al.*, 1971) including those seen in children (Ball, *et al*, 1970). Some of the cases described as Bickerstaff's Encephalitis (1957) were probably also examples of the same syndrome.

It should be noted that pathological changes within the cerebellar system had not been noted in cases of infective polyneuritis. Richter (1962) reported an autopsy of the case where the patient had the Fisher's syndrome four years prior to death. The dorsal roots were demyelinated and there was loss of myelin (but not neuron) and glosis of the Clarke's column as well as demyelination of the middle root zone of the posterior column. It was suggested, therefore that deafferentation of the Clarke's column was responsible for the ataxia.

Our patients with total ophthalmoplegia, facial weakness, areflexia and ataxia, fit into the cases of Fisher's syndrome very well. Respiratory complication and transient hypertension were well known to occur in infective polyneuritis. Thus our first patient with hypoxia and transient hypertension further support that Fisher's syndrome is a variant of the Infective Polyneuritis.

Clinically, the differential diagnosis to be considered include myasthenia gravis, botulism, poliomyelitis and Wernicke's encephalopathy. The negative edrophonum test and ataxia made the diagnosis of myasthenia gravis and botulism unlikely. Bulbar poliomyelitis has been said to cause external ophthalmoplegia only rarely and probably never outlasted the first few days of the disease. Pupillary paralysis never occurred (Price *et al*, 1978). The relatively normal CSF was also unusual for acute poliomyelitis. Werwicke's encephalopathy, when causing such severe ophthalmoplegia, would be expected to have marked impairment of the conscious state.

ACKNOWLEDGEMENT

I wish to express my gratitude to Puan Rohani who helped to type the manuscript.

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