Acute inflammatory demyelinating polyradiculoneuropathy presenting as complete heart block and Stoke-Adams attacks

Bashir Ahmad Wani, Mukul Misra, Mehrajuddin Shah and Showkat Mufti

Department of Cardiology, Sher-i-Kashmir Institute of Medical Sciences, P.B.27, Srinagar-190 011, (J & K), India.

Summary: A patient with acute inflammatory demyelinating polyradiculoneuropathy (AIDP, Guillain-Barré syndrome) whose presenting complaints were related to autonomic dysfunction in the form of parasympathetic and sympathetic overactivity is reported. Parasympathetic overactivity was severe enough to cause complete atroventricular block (atropine-responsive) and Stoke-Adams attacks, for which a demand pacemaker was required. Limb weakness was apparent 48 hours later. To our knowledge no such presentation of AIDP has been previously recorded.

Introduction

The usual presentation of acute inflammatory polyradiculoneuropathy (AIDP) is subacute muscular weakness with areflexia arising from the lower limbs and ascending gradually in more than 90% cases. Autonomic dysfunction occurs not uncommonly and is expressed in several ways. Exceptionally, symptoms due to autonomic dysfunction may be the presenting complaint of the disorder. Rarely, patients show sympathetic and parasympathetic overactivity simultaneously. We report a patient with AIDP who not only showed sympathetic overactivity but who presented with complete atroventricular block due to parasympathetic overactivity resulting in recurrent Stoke-Adams attacks.

Case report

A 55 year old male was admitted to the coronary care unit with a history of recurrent syncopal attacks of about 24 hours duration. He had had moderate pyrexia and generalized myalgia with a few loose motions for 2 days previously. The patient was in coma with spontaneous respiration at a rate of 16–20/minute. The pulse was regular at a rate of 45/minute. All the pulses were equally palpable and the blood pressure in the right arm was 150/110 mmHg. There were signs of complete heart block but no murmur, rub or gallop sound was present, neither was there any sign of congestive heart failure. A neurological examination at this stage did not show any focal deficit and the pupils were equal and normally reacting. The deep tendon reflexes were present though sluggish and the plantar responses were normal.

Electrocardiogram showed complete heart block with a narrow QRS escape rhythm at a rate of 40–45/minute. There was no evidence of myocardial ischaemia or infarction. Intravenous atropine increased the heart rate to 88 beats/min with intermittent runs of sinus capture; the blood pressure did not change. Since the response lasted only 20–30 minutes, an isoprenaline drip was started which increased the heart rate to 60–65 beats/min, and blood pressure changed to 170/104 mmHg. However, to obtain a stable rhythm a temporary transvenous pacemaker was inserted. Gradually the patient regained his own rhythm after 48 hours. After the insertion of the pacemaker, there was no further syncopal attack and the patient regained consciousness within 30–40 minutes. Routine investigations, including the cardiac enzymes, were within normal limits. The serum sodium and potassium levels were 139 and 3.9 mmol/l, respectively, at the time of admission. Urine examination for porphobilinogen was negative. Chest X-ray was within normal limits. Antihypertensive therapy with alphamethyldopa 250 mg twice daily and a diuretic along with other supportive drugs was started.

However, on the third day of hospitalization, he developed flaccid paralysis with areflexia of all the four extremities with grade 0–1/5 power in the upper limb, and grade 1–2/5 power in the lower limb muscle groups. There was no involvement of the cranial nerves and respiratory muscles. Diagnosis of AIDP was suspected and nerve conduction velocity studies were done. In general, the distal latencies and conduction velocities were delayed in the nerves of lower limbs, suggestive of demyelination. The F waves were absent suggesting radiculopathy. Electromyograms

Correspondence: Professor B.A. Wani, M.D., D.M.
Accepted: 11 August 1988

© The Fellowship of Postgraduate Medicine, 1989
recorded from the right quadriceps muscle group and right and left abductor pollicis brevis muscles were of neurogenic type. The study of right median and ulnar sensory segments and both sural nerves suggested sensory impairment as well. Thus, from the electrophysiological studies, the clinical diagnosis of AIDP was confirmed. Cerebrospinal fluid examination was normal on two occasions at an interval of 2 weeks.

The patient showed a good neurological recovery with the power returning to grade 4/5 over a period of 6 weeks and was discharged with physiotherapy advice. His blood pressure was 120/70 mmHg without any antihypertensive drugs at the time of discharge.

Before discharge, we performed echocardiographic examination which revealed normal heart chamber sizes and left ventricular functions. Cardiac electrophysiological study was also done which showed normal PA (27 ms), AH (102 ms) and HV (44 ms) intervals. The response to rapid atrial pacing was normal and atrioventricular block did not appear up to atrial pacing rate of 180/min.

**Discussion**

Autonomic dysfunction occurs in patients with AIDP and presents in several ways. Sinus tachycardia is common, occurring in about 50% of severe cases. Its cause is disputed. However, the absence of a response to carotid sinus stimulation suggests vagal damage to be the responsible factor. Postural hypotension due to loss of sympathetically mediated vascular reflexes with fainting attacks as a rare presenting manifestation of abnormal Valsalva response, abnormal pressor response to agents like phenylephrine, and hypertension or hypotension occurring with equal frequency are some other clinical manifestations of autonomic dysfunction. The causes of hypertension are varied and include possible denervation of neck vessels, high plasma catecholamine levels, and high plasma renin activity. The sympathetic or parasympathetic function may be deficient but at other times, overactivity of both has been described in the same patients.

Our case also demonstrated overactivity of the parasympathetic system in the form of complete atrioventricular block with narrow QRS escape rhythm (responsive to atropine) while at the same time, the patient had reversible hypertension as a manifestation of sympathetic overactivity. The major point of interest is not only the association of these two features but that the Stoke-Adams attacks due to parasympathetic overactivity were the presenting complaint in the patient. Such an occurrence has not been previously described in the plethora of literature available on AIDP.

The use of demand pacemakers has been described previously in AIDP but the indications were otherwise than reversible atrioventricular block. One could argue that the atrioventricular block could have been due to myocarditis associated with AIDP in this patient. However, the absence of a rise in myocardial enzyme levels and of left ventricular dysfunction makes this complication unlikely.

**References**