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Recurrent lobar pneumonia associated with idiopathic Eaton-Lambert syndrome

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ABSTRACT A patient with idiopathic Eaton-Lambert syndrome presenting with recurrent pneumonia as a consequence of the underlying muscle weakness is described.

Eaton-Lambert (myasthenic) syndrome with proximal muscle weakness and a characteristic electromyographic pattern was first described in 1957.[1,2] It differs from classical myasthenia gravis in that the underlying disorder is presynaptic as opposed to postsynaptic.[3] Diseases associated with muscle weakness are recognised as causes of pulmonary infections. We describe a case of Eaton-Lambert syndrome without underlying malignancy who presented with recurrent lobar pneumonia.

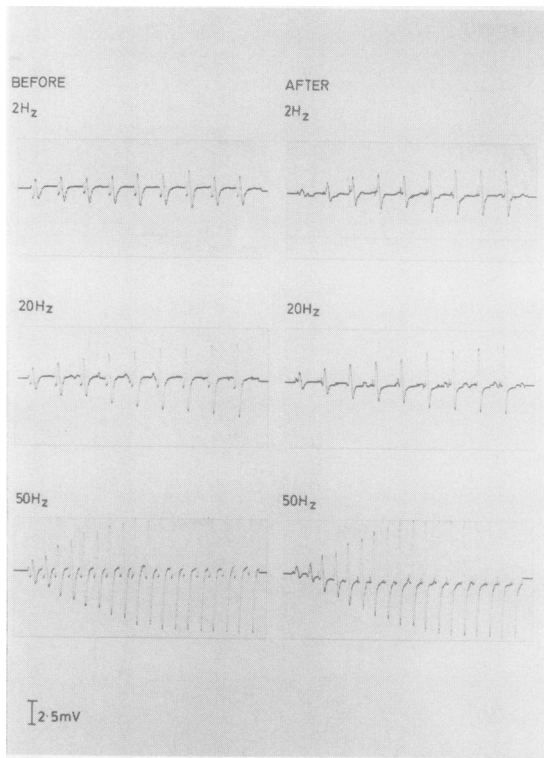
Case report

A 72 year old retired sherry importer presented with a one week history of an upper respiratory tract infection. He reported malaise and cough productive of discoloured sputum with minimal dyspnoea but no haemoptysis or chest pain. His medical practitioner described a rapid progression to a state of "myalgic immobility." There had been gradual impairment of walking over eight years with weakness of his knees and floppiness of his feet. He had no paraesthesiae, back pain, bladder symptoms, weakness, bulbar symptoms (dysarthria, dysphasia, dysphagia), or deterioration in vision or hearing. Systemic inquiry disclosed nothing remarkable.

Nine months previously he had had right upper lobe pneumonia, which slowly cleared with ampicillin treatment. He also had benign prostate enlargement, and a parapelvic cyst of the right kidney with mild renal impairment (plasma creatinine 199 μmol/l). He had smoked a pipe until 15 years previously but rarely drank alcohol. There was no family history of respiratory or neurological problems.

On examination he looked unwell and was febrile, drowsy, and so weak that he required assistance to sit up in bed. He was mildly cyanosed with inspiratory crackles and a pleural rub audible at the right base and crackles at the left base. After he had recovered from acute pneumonia neurological examination showed normal gait with no muscle tenderness or muscle wasting and normal muscle power. At rest the deep tendon reflexes were uniformly absent even with reinforcement but could be elicited for several minutes after forceful contraction of the specific muscle. Plantar responses were flexor. Cranial nerves and sensation were normal.

Chest radiography showed extensive right sided opacification in the mid and lower zones and a further small area above the left diaphragm. Sputum and bronchial washings, taken after he had started treatment with amoxycillin and clavulanic acid, showed moderate numbers of pus cells and *Escherichia coli* sensitive to co-trimoxazole and the amoxycillin-clavulanic acid combination. Culture of midstream urine and blood, sputum culture for *Mycobacterium tuberculosis* and the results of serological tests for atypical pneumonia organisms were negative. Bronchoscopy showed



Electromyogram showing the increase in amplitude of muscle contraction response to repetitive stimulation of the nerve at frequencies of over 2 Hz. After guanidine treatment this response occurred at lower stimulus frequencies.

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no underlying tumour. Other investigations showed a normal haemoglobin concentration and platelet count, a white cell count of 13·4 × 10^9/l, an erythrocyte sedimentation rate of 125 mm in one hour, a blood urea concentration of 24·9 mmol/l, and a serum creatinine concentration of 212 μmol/l. Values for arterial blood gases on admission, while the patient was breathing air, were: oxygen tension 7 kPa, carbon dioxide tension 5·4 kPa, hydrogen ion 44 nm/l. Protein electrophoresis showed a diffuse increase in globulins. Serum cobalamin and folate concentrations and the results of an autoantibody screen for thyroid disease and pernicious anaemia were normal; serological testing for syphilis gave a negative result.

Sensory and motor nerve conduction velocity studies gave normal results but muscle action potentials were of low amplitude after single stimuli, though they showed an incremental rise in response to repetitive stimulation at frequencies above 10 Hz, as is characteristic of Eaton-Lambert syndrome (fig).

The pneumonia responded clinically and radiologically to treatment. Guanidine hydrochloride 500 mg thrice daily was started, with a good symptomatic response and an increase in exercise tolerance and in FEV_1 (from 2·8 to 3·6 l). The dose was reduced to 300 mg thrice daily because of diarrhoea and he has remained well with this treatment for one year.

Discussion

Eaton-Lambert syndrome presenting with recurrent pneumonia in the absence of bronchial carcinoma has not, so far as we are aware, been described. Respiratory infections are a well recognised complication of several diseases associated with muscle weakness, including Guillain-Barré syndrome, congenital myopathies, motorneurone disease and classical myasthenia gravis.[4 5] There is evidence that intercostal muscles are affected in Eaton-Lambert syndrome,[3] which may lead to hypoventilation and predispose to pneumonia.

Eaton-Lambert syndrome was first described in 1957 in association with small cell carcinoma of the lung.[1] It differs from classical myasthenia gravis in the distribution of the weakness, which usually affects the proximal leg muscles and spares the bulbar muscles, and in affecting the presynaptic nerves by impeding the release of acetylcholine.[6 7] Although Eaton-Lambert syndrome occurs in 3% of patients with small cell bronchial carcinoma it is now recognised that many cases are not associated with an underlying carcinoma, and in such cases there is an increased incidence of autoimmune disease.[6] In view of the eight year history of weakness and the absence of any evidence of a bronchial or other carcinoma this case falls into the idiopathic group, recurrent respiratory infections presumably being caused by virtue of respiratory muscle weakness. As Eaton-Lambert syndrome is an eminently treatable condition the diagnosis is important, particularly for patients with the non-carcinomatous form of the disease.

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