Hemiballismus from cerebellar metastases

Sir: A 50 year old alcoholic woman with metastatic mucinous signet ring cell adenocarcinoma of the colon developed irregular movements of the proximal and, to a lesser extent, distal joints of her right arm and leg. The next day, the movements became more abrupt, of wide-amplitude and almost continuous. They worsened when she became anxious during the neurologic examination and disappeared when she slept. Her face and tongue were spared. Aside from generalised muscle weakness and hypoactive muscle stretch reflexes, her neurologic examination was normal. She had no ataxia or terminal tremors of her limbs, nystagmus, hypotonia, slurred speech or abnormal mental function. She died of cardiorespiratory arrest two days later. At necropsy, she had a large metastatic tumour in her right cerebellar hemisphere impinging on the dentate nucleus and stretching the superior cerebellar peduncle (fig) and a smaller metastasis in the folia of the left cerebellar hemisphere. No gross or microscopic tumour was found in the decussation of the superior cerebellar peduncle, red nucleus or subthalamic nucleus of Luys. The latter was sectioned throughout its rostrocaudal extent on both sides. Our patient and others with no lesions in their subthalamus at necropsy or on neuroimaging and the few patients with subthalamic lesions but no hemiballismus remind us that the aetiopathogenesis of hemiballismus is still unexplained. Also, we should no longer teach that hemiballismus necessarily results from a lesion in the subthalamus.

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References

Matters arising

A neuromuscular transmission disorder: combined myasthenia gravis and Lambert-Eaton syndrome in one patient

Sir: In a recent article, Neuromuscular transmission disorder: combined myasthenia gravis and Lambert-Eaton syndrome in one patient Taphoorn1 described a case with clinical and neurophysiological characteristics that suggested an association between the two diseases. The neurophysiological data were compatible with Lambert-Eaton myasthenic syndrome (LEMS) whereas some clinical observations indicated myasthenia gravis (MG). In another report antibodies against acetylcholine receptors and anti-striated muscle antibodies were present in the serum2 as well as antibodies against thyroid and gastric mucosa; however, this finding is not an absolute criterion in differential diagnosis, since in both LEMS and MG associations with other autoimmune diseases have been reported.3

We observed a patient who died of an unidentified cause after presenting electromyography (EMG) findings and a clinical picture suggestive of a mixed form of LEMS and MG. The patient was a 61 year old man who had a 2 month history of myalgia, weakness of the pelvic and shoulder girdles with muscular exhaustibility on effort, dysphonia and dysphagia, slight bilateral ptosis of the eyelids, dry mouth and constipation. The symptoms had a recurrent pattern, each lasting several days, with abrupt onset and slowly progressive resolution.

The main symptoms reported by the patient (bilateral ptosis of the eyelids, impairment of the bulbar musculature, fluctuations of the symptoms) suggested MG whereas two EMGs performed during relapses were compatible with LEMS. The latter examinations demonstrated signs of myopathy of the proximal muscles and the first compound muscle action potential (CMAP) evoked by ulnar nerve stimulation had a small amplitude with an about 300% increase of the CMAP at repetitive stimulation at 30 Hz. Administration of edrophonium chloride did not modify the clinical characteristics or EMG findings, nor was any benefit obtained from treatment with pyridostigmine bromide, performed also for diagnostic purposes. Screening for autoantibodies was negative, as were studies performed to demonstrate possible neoplastic disease (bone scintigraphy, computed tomography and magnetic resonance imaging of the mediastinum).

While the patient was still in hospital but in fairly good clinical condition and not receiving any treatment, on waking one morning he had a sudden dyspnoeic crisis, became cyanotic and died in a few minutes. Necropsy did not reveal any possible cause of death, although a small-cell bronchial carcinoma of small size was found with lymph node invasion.

The manner of death and the absence of a documented cause seemed to us strongly suggestive of a myasthenic crisis, which has been reported in the literature in only three patients with LEMS;4,5 in these cases there was a progressive weakness, most marked at the shoulder and pelvic girdles, and scanty involvement of the ocular and bulbar muscles. In these patients also death occurred suddenly and "unexpectedly".

We conclude that there may be a more frequent association between MG and LEMS than reported in the literature. The clinical and neurophysiological characteristics of the "overlap myasthenic syndrome" should be better defined with the aim of clarifying its pathogenesis and because of possible therapeutic implications.

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