Post radiation monomelic amyotrophy

Lamy et al recently reported three cases of post radiation lower motor neuron syndrome presenting as monomelic amyotrophy.1 I would like to report a further case.

A thirty six year old man presented in 1989 with weakness abducting his left hip. Nineteen years before he had been treated for a left testicular seminoma with an orchidectomy and radiotherapy. The abdominal radiotherapy field dispensed a prescribed tumour dose of 32.5 Gy, and the para-aortic fields increased the dose to 50 Gy in twenty fractions. The fields included the lower 6 cm of the spinal cord, the whole cauda equina and the lumbarosacral roots and plexuses. The initial weakness of the left leg has gradually progressed to weakness and wasting of all muscle groups with ankle dorsiflexion, hip flexion and abduction being more severely affected. He now walks with a stick. The limp is areflexic with a flexor plantar response. There is no sensory loss or sphincter involvement. The right leg is not involved. Six months of treatment with prednisolone (20 mg per day) was of no benefit. Electrophysiological examination showed advanced denervation confined to the muscles of the left leg, with large amplitude motor unit potentials. An abnormal axon reflex and absent F wave in extensor digitorum brevis suggests a lesion at the root or anterior horn cell level. Motor and sensory conduction velocities and distal latencies were normal. General examination, laboratory investigations including CSF analysis and a myelogram were all normal.

The 17 year latency in this case, and the nine and twelve year latencies reported by Lamy et al,1 as well as the previously mentioned electrophysiological data point to an anterior horn cell disorder reminiscent of the poliomyelitine syndrome.2 It is plausible that the radiotherapy damaged a critical number of motor neurons and that surviving neurons sprout to reinnervate more muscle fibres than normal. This process produces large motor units that may stress the cell body. After a number of years these hyperfunctioning motor units may not be able to maintain the metabolic demands of all their sprouts and a deterioration of individual terminals may result. Eventually enough nerve terminals are destroyed and enough reserves are diminshed for weakness to appear. This would be consistent with the fall in nature of post radiation lower motor neuron syndrome and its slow, stepwise and unpredictable progression.

As Lamy et al state, the cause of this disorder seems unpredictable. My patient continues to deteriorate. Maier et al report that three of their 15 patients had a monomelic amyotrophy, but give no information on clinical course. Vibeke Schioldt and Kristenson2 report that two of their patients had monomelic amyotrophy—one had “slight subjective weakness of the left leg” which had improved by six months; the other case had non-progressive right leg weakness.

Sellar tuberculoma

In Asian countries, before the advent of chemotherapy, tuberculomas accounted for about 30% of all intracranial lesions.3 They remain a major problem even though they are now less common due to antituberculosis drugs and improved living conditions. Intrasellar tuberculomas, not uncommonly, present as post mortem examinations, rarely present clinically.2,4 Only five surgically verified cases have been reported to date.

A 40 year old man, resident of an area where tuberculosi is endemic, presented in January 1988 complaining of an intermittent, dull, generalised headache of two years duration, and progressive diminution of vision in both eyes over a period of six months. He was in good general health with no clinical signs of endocrinopathy. Visual acuity was reduced (right eye—finger counting 3 m; left eye—hand movement perception at one meter). Perimetry showed a restricted field of vision in the right eye, the left was unascertainable. He had bilateral optic atrophy, but no other neurological abnormality. The clinical diagnosis was a sellar tumour. Haematological and biochemical investigations were unremarkable except for an erythrocyte sedimentation rate (ESR) of 90 mm in the first hour. Radiographic examination revealed a bony hypodense sellar mass with an area of hypodensity within it. CT scan (figure) showed a uniformly hyperdense enhancing sellar mass with a suprasellar extension. An operation was performed on 14 January 1988 using a transsphenoidal transnasal approach. The sellar floor and dura mater were intact. The dura mater was tough and thickened and when it was opened, a greyish white, tough intrasellar tumour was revealed. It was adequately decompressed under intraoperative pneumocephalography. Microscopic examination showed that pituitary tissue had been almost completely replaced by granulomas comprising of epitheloid cells, Langhan type of giant cells surrounded by lymphocytes, and plasma cells. Minimal casation was present in some granulomas. Tuberculoma of the pituitary gland was diagnosed. A postoperative Mantoux test was positive but sputum culture for acid fast bacilli was negative. Treatment was started with isonex, rifampicin and ethambutol. The patient developed hepatotoxicity to rifampicin, but isonex and ethambutol were continued for nine months. He was in good health, had no headache, and vision in the left eye had improved (finger counting-3 m). The right eye vision showed no improvement in the constricted field (finger counting—6 m).

A pituitary tuberculoma is extremely rare but usually presents as a chiasmal syndrome. In two reported cases the lesion was successfully removed subfrontally.5,6 An exclusively intrasellar tuberculoma was approached transtemporally and treated with isonex only for three months. In our patient the lesion was intrasellar with a suprasellar extension. It could be treated, however, by a transnasal transphenoidal approach. There was little reason to suspect a pituitary tuberculoma before the operation except for the raised ESR. The transnasal transphenoidal approach allowed a subtotal removal of the tuberculoma while avoiding CSF contamination by tuberculous material.

Tuberculomas meningitis occurs in the majority of surgically treated intracranial tuberculomas without antituberculosis chemotherapy.1-3 Chemotherapy should usually be given in a three drug combination for three months, followed by a two drug combination for a further 15 months.

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