Short report

Orthostatic hypotension with lower brain stem glioma

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SUMMARY A patient with long standing orthostatic hypotension with fixed pulse rate due to involvement of the medullary regulatory centres by a lower brain stem glioma is described.

Paroxysmal hypertension recently has been related to stimulation of the sympathetic vasomotor centre of the lateral medulla by posterior fossa lesions.¹ Orthostatic hypotension could be anticipated in destructive lesions of the same region. However, orthostatic hypotension is more frequently related to gradual failure of peripheral autonomic nerves than to central nervous system lesions. The purpose of this paper is to describe the characteristics of long standing orthostatic hypotension associated with a lower brain stem glioma.

Case report

This 30-year-old patient fell down when getting up one morning; he did not lose consciousness but was unable to stand up. These difficulties continued for a few days and then subsided but recurred four years later. At that time neurological examination disclosed orthostatic hypotension and left palatal paresis. The patient was able to work as a charcoal burner until 50 years of age, being only incapacitated two or three times a year by hypotensive attacks associated with sweating and occasional loss of consciousness. At the age of 50, transient dysphagia was noted and followed two years later by painful attacks of paraesthesiae and cold sensations starting at the right hand, progressing to the right shoulder and right side of the face and lasting a few minutes. Later, the right thoracic wall became also involved in this process. The patient was able to shorten those attacks by grasping something hard in his right hand. At times he complained about hoarseness of his voice. At 54, his general condition deteriorated progressively because of increasing frequency of orthostatic "fainting" and painful paraesthetic attacks recurred 5 to 10 times a day. At that time, loss of the left corneal reflex was noticed along with left facial

hypoesthesia, left palatal and laryngeal paralysis and broad-based gait. His blood pressure fell from 120/90 to 60/40 mmHg without cardiac acceleration when he attempted to stand up from the supine position. Vestibular testing demonstrated directional preponderance of the caloric response to the left. When 55 years of age, the patient became progressively bedridden. Paradoxically moderate hypertension was observed transiently during painful attacks of paraesthesiae in association with restlessness, salivation and pilo-erection. Several determinations of the urine catecholamine level were in the normal range. Neurological examination disclosed central vestibular nystagmus, left Horner's syndrome and on the same side, marked decrease of the corneal reflex, palatal vocal cord and sternocleidomastoid paralysis as well as slight atrophy and fasciculation of the tongue. There were no pyramidal signs. The resting bloodpressure varied from 120/80 to 70/50 mmHg from day to day but fell to almost zero each time the patient tried to sit up, causing loss of consciousness without pulse acceleration. 9-alpha-fluorohydrocortisone administration (0.1 mg three times a day) improved the orthostatic hypotension and the patient was able to walk with a somewhat ataxic gait. However, his condition deteriorated again, and he developed increasing peripheral left facial palsy, incoordination of both left limbs, worsening of the orthostatic hypotension and died a few months later at the age of 55.

General necropsy was unrevealing, except for hepatosplenorenal congestion. In the neuropathological examination no abnormality could be found in the spinal cord, anterior and posterior roots, cerebral hemispheres, basal ganglia and hypothalamic nuclei. Except for some binucleated ganglion cells, there were no morphological changes in the cervical sympathetic ganglion cells and fibres. No Lewy bodies could be seen. The pigmented brain stem nuclei were also normal. The only striking abnormality was a strictly delineated white firm left lower brain stem tumour extending from the obex to 5 mm below the emergence of the trigeminal nerve, in the left posterior part of upper medulla and pons, pushing back the surrounding structures and invading almost the entire lower part of

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Received 1 July 1982.
Accepted 3 September 1982.

Fig 1 A. Upper pons without tumoral infiltration. B. Middle pons with small tumoral infiltration in the left dorsal tegmentum. C. Lower pons with larger tumoral infiltration in the left tegmentum. D. Full development of the tumour in the left medulla with displacement of the midline. The bar is 5 mm. 10 μ m paraffin sections. Hematoxylin-eosin staining.

medulla. This tumour was 5 cm in length and 3 cm in width (fig 1 A, B, C, D).

Microscopically, the centre of the tumour was poorly cellular, disclosing bundles of elongated bipolar cells with gliofibrillar processes (fig 2A). Rosenthal fibers were also present as well as eosinophilic masses of varying shape. The periphery of the tumour showed advanced anaplastic changes with highly cellular infiltration of the surrounding structures (fig 2B) and with striking preservation of neurons together with polymorphism of nuclei and rare mitoses. The anaplastic changes extended from the initially benign tumour to the upper pons and right part of the brain stem.

Discussion

The histological features of this tumour are those of pilocytic astrocytoma. According to Russell and Rubinstein,² a unilateral pontine and medullar localisation characterises a subgroup of those tumours; although the 25 years evolution of our case seems rather exceptional, the maximal duration quoted by Russell and Rubinstein was 30 years in a temporal lobe pilocytic astrocytoma; anaplastic changes are a terminal event in 60% of the cases.

Orthostatic hypotension has been described in association with various central nervous disorders but pathological verifications are scanty and lesions too disseminated to correlate orthostatic hypotension with a definite localisation. For example, orthostatic hypotension is seen in the Shy-Drager³ variant, in elderly idiopathic orthostatic hypotension,⁴ in multiple sclerosis,⁵ cerebrovascular disease⁶ and Wernicke's encephalopathy.^{8,9} Spinal lesions

have been also postulated, but not always demonstrated, in tabes,^{10,11} syringomyelia,¹⁰ trauma,¹⁰ surgical chordotomy¹² and subacute combined degeneration of the spinal cord.^{13,14} Orthostatic hypotension has been observed with some intracranial tumours around the third ventricle suggesting a disturbance of autonomic hypothalamic centres.¹⁵ Posterior fossa tumours have been rarely associated with orthostatic hypotension. Wagner¹⁶ mentioned an elderly woman with multiple cranial nerve deficit due to a tumour on the floor of the fourth ventricle, without other details. Two similar cases were added by O'Malley *et al*¹⁷ in 1970 and Riedel *et al*¹⁸ reported two observations with orthostatic hypotension complicating posterior fossa neurosurgery. In our case, orthostatic hypotension was the only presenting symptom, the next clinical signs to appear being referable to the left medulla pilocytic astrocytoma. Consequently, destructive changes of the left lateral medulla vasomotor centre can be reasonably incriminated in the pathogenesis of this centrally-induced orthostatic hypotension with fixed pulse rate.

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Fig 2 A. Pilocytic astrocytoma showing elongated cells replacing longitudinal and transverse myelinated tracts. B. Anaplastic changes in the vicinity of the pilocytic astrocytoma. 10 μm paraffin sections. Hematoxylin-eosin staining.

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