ORTHOSTATIC HYPOTENSION ASSOCIATED WITH CEREBELLAR HEMANGIOBLASTOMA

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SUMMARY

A case with a posterior fossa tumor presented because of orthostatic hypotension. Posterior fossa exploration and excision of the tumor revealed a hemangioblastoma.

INTRODUCTION

Adjustment of the circulation to changes of body posture from recumbent to sitting or upright posture depend on several mechanisms directed to preservation of arterial pressure (4). Postural changes provoke rapid reflex peripheral vasocostriction and an increase in heart rate and plasma norepinephrine content(3). The initial fall in blood pressure after assuming the upright position leads to a decrease in baroreceptor impulses from the carotid sinuses and aortic arch to the vasomotor centre which relays in the brainstem; an increase in tone of the adrenergic sympathetic sympathetic nerves should then occur, with consequent effects upon peripheral blood vessels and heart function(4).

Neurogenic orthostatic hypotension has been attributed to a variaety of diseases including idiopathic orthostatic hypotension, parkinsonian syndrome, cerebrovascular disease, diabetic neuropathy, familiar dysautonomia, the Holmes–Adie syndrome, the Guillaine–Barre syndrome, acute pandysautonomia, tabes dorsalis, Wernicke encephalopathy, transverse myelitis, spinal cord injury, porphyria, mitral valve prolapse and amyloidosis(1). In idiopathic orthostatic hypotension, functional studies imply a defective baroreceptor reflex arc and pathologic changes of diffuse neuronal degeneration of CNS included the sympathetic pathways (3,4). Pathophysio-
logy of orthostatic hypotension in other diseases is less clear, but it is now generally believed that the final factor in causing orthostatic hypotension is a failure of noradrenaline release from vasoconstrictor nerve endings (2, 3).

Focal brainstem lesions, including syringobulbia, vascular disease and tumor rarely cause orthostatic hypotension (3). A few tumors have been reported in the literature, including craniopharyngioma, prolactin secreting pituitary adenoma, brainstem neoplasms, cerebellar hemangio-blastoma, cerebellar gangliocytoma (Lhermitte–Duclos disease), cerebellar tuberculosis, fourth ventricle tumor (1, 2, 3, 5, 7, 10, 11). Also 2 cases with orthostatic hypotension have been reported, which appeared after having had a surgery on their brainstem neoplasms(6).

We present a patient with cerebellar hemangioblastoma in whom orthostatic hypotension was the initial major manifestation.

CASE REPORT

A 55 year old male patient (M. K) presented to the Neurosurgical Department of Trakya University Medical Faculty in March 1985 with a 4 year history of vertigo, postural hypotension, bitemporal headache, recent blurring of vision and a fall when getting out of his car one week previously. Blood pressure in the lying position was 120/70 mmHg, pulse 80/min, but in the sitting position it was 60/30 mmHg and pulse 110/min, with disabling vertigo. There were no other positive neurological findings and fundoscopy was also normal. CT scanning (30.03.85) showed a midline posterior fossa space occupying lesion, homogen enhancing after contrast injection, situated behind the 4. ventricle which could not be seen as it was possibly compressed by the tumor but no obvious obstructive hydrocephaly present (Fig: 1).

As the patient refused at first any surgical intervention, he was put on steroids for some time with no effect. Two months later, his complaints got worse and he developed cerebellar signs. Repeated CT scanning (30.05.85) showed that the posterior fossa tumor had become cystic, not enhancing and extending upto tentorial hiatus (Fig: 2). Posterior fossa exploration was carried out, with excision of the tumor by standart approach (8), histopathologic diagnosis was hemangioblastoma. Sadly, at the 10 the postoperative day he died as a consequence of pulmonary embolism being secondary to deep venous thrombosis in the right leg. Permission for autopsy was refused.
Figure 1. Initial CAT scanning shows a space occupying lesion in posterior fossa with homogene enhancement.

Figure 2. Repeated CAT scanning, (2 month after the initial CAT) shows the lesion became cystic and not enhancing with contrast.

DISCUSSION

Several important compensatory cardiovascular mechanisms come into play when one changes from the supine to the standing position, among
which are a reflex increase in heart rate and peripheral vascular resistance, constriction of the veins and the release of several vasoactive hormones. The purpose of these physiological changes is to ensure the constancy of blood pressure and, therefore, maintain an adequate blood supply to the brain, heart and other vital organs. The activation of these compensatory mechanisms requires the integrity of several afferent pathways, the low and high pressure baroreceptors and controller areas in the central nervous system. The lower brainstem plays a significant role in this sphere by receiving an afferent input of cardiovascular reflexes and interacting these inputs with the descending influences from other areas of the brain(6).

Tumors have been rarely observed associated with orthostatic hypotension. Orthostatic hypotension associated with tumors around the third ventricle was thought to be secondary to a disturbance of autonomic hypothalamic centres (7, 10). In patients with malignant tumors, orthostatic hypotension has been attributed to polyneuropathy, intra thoracic interruption of the low pressure afferent baroreceptor limb, a remote effect of cancer, hyponatremia, hypoaldosteronism (2, 3, 5).

Posterior fossa tumors have only rarely been associated with orthostatic hypotension (10). HSU et al, reported three patients with brainstem tumors and orthostatic hypotension. Two of them had a primary CNS tumor which involved the dorsal medulla, pons and rostral spinal cord and the third had a metastatic oat-cell carcinoma from the lung with subependymal spread to the medulla and pons(3). TELERMAN-TOPPET et al, reported a patient with longlasting orthostatic hypotension due to invasion of the medullary regulatory centers by a lower brainstem glioma (10). RIEDEL et al, reported two observations with orthostatic hypotension complicating posterior fossa neurosurgery. Although the anatomic site of the autonomic defect has not been confirmed, as the most likely area of dysfunction was thought to be the brainstem centers (6). In our case the mechanism of orthostatic hypotension was also thought to be secondary to the mass or destructive effect of the tumor on the medullary-regulatory centers.

Acknowledgement: The author (S.C) is extremely thankful to Prof. J. Douglas MILLER, Head of the Dept. of Clinical Neurosciences (Surgical Neurology), Edinburgh University, who kindly reviewed the article.

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