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Cerebellar Glioblastoma Presenting Clinically as Wallenberg's Syndrome

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Summary. The clinico-neuropathological report is given of a glioblastoma multiforme with primary site in the cerebellum. The patient presented clinically with Wallenberg's syndrome; morphological investigations revealed the tumor partly invading the dorsolateral region of medulla oblongata. There are some reports of Wallenberg's syndrome not caused by vascular stenosis or occlusion, but by metastatic tumors in the lateral medullary region. The present report is the first of a cerebellar glioblastoma causing the peculiar brainstem syndrome.

Key words: Glioblastoma multiforme – Cerebellar tumor – Lateral medullary syndrome

Introduction

Glioblastomas arising primarily in the cerebellum are very rare (Russell and Rubinstein 1971; Rubinstein 1972; Miller et al. 1976; Jänisch et al. 1976). Until now about ten well-documented cases have been reported (Tibbs et al. 1980). We present a case of a glioblastoma in an unusual site showing clinical signs and symptoms of Wallenberg's (1895) syndrome. Although there have been reports of a lateral medullary syndrome caused by processes other than stenosis or occlusion of the posterior inferior cerebellar or vertebral arteries, a glioblastoma causing this syndrome has not so far been described.

Case Report

The patient was a 54-year-old woman with no relevant previous illness. Three weeks before admission she suddenly experienced a numb feeling in the left side of her face. Two days before admission she had hiccup and vomiting. Neurological examination (3 months before death)

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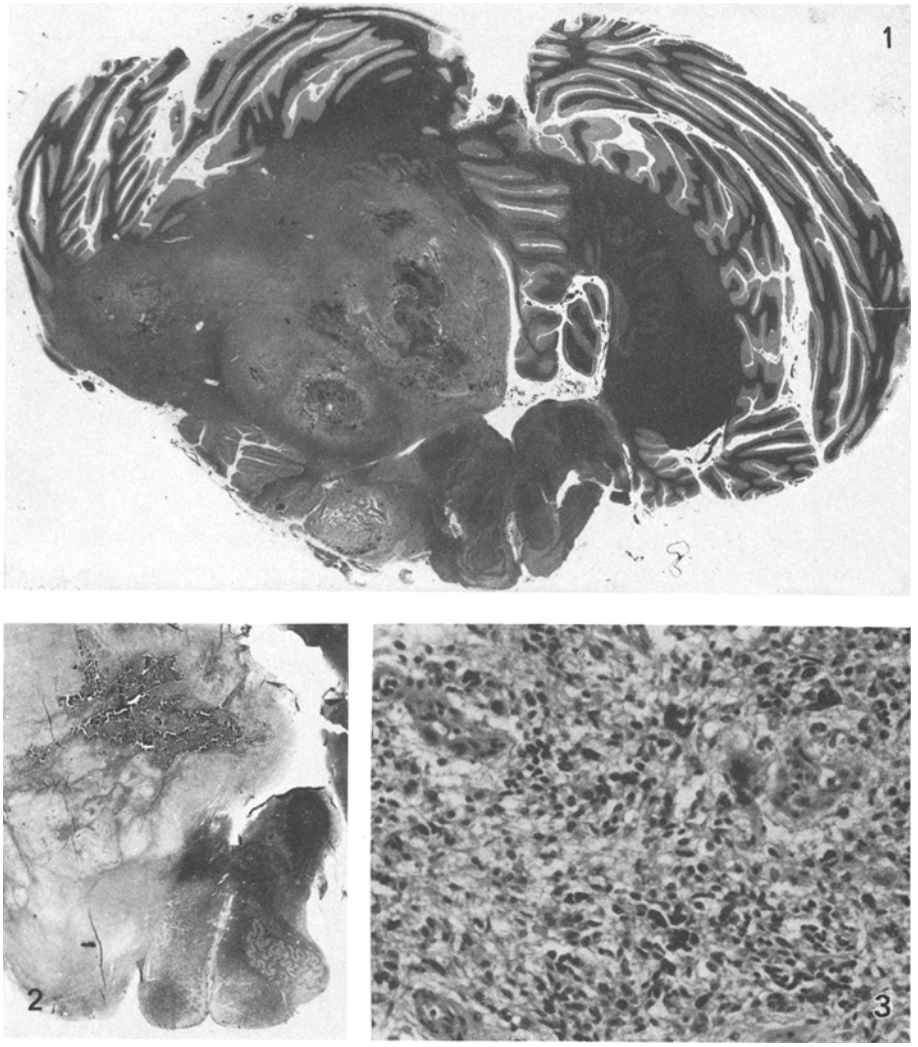

Figs. 1–3. Cerebellar glioblastoma

Fig. 1. Macroscopical appearance. Tumor in the left cerebellar hemisphere but not invading the dorsolateral part of the medulla oblongata. Paraffin, Heidenhain

Fig. 2. Tumor invading the left dorsolateral part of the medulla oblongata. Paraffin, HE

Fig. 3. Histological features showing poor differentiation, high cellularity with mitoses and endothelial proliferation. HE × 190

showed incomplete closing of the left eye and a left-sided ptosis without pupillary difference. Mild spontaneous nystagmus to the left was found bilaterally, the convergence reaction was missing on the left side. Corneal reflex was diminished on the left. There was hypo-aesthesia on the left side of the face and tongue and thermohypo-aesthesia of the left frontal region. Sense of pain and temperature sense was diminished on the right side of trunk and extremities. Speech was slightly aphonic but otherwise normal. Finger-nose test revealed slight ataxia of the left arm. Gait and station could not be examined because of severe vertigo.

EEG showed diffuse abnormalities with a left temporal theta-delta focus and activation by hyperventilation. X-ray examination of the skull and isotope brain scan were normal. Cranial computed tomography was inconclusive due to artefacts.

Course. After slight initial improvement symptoms worsened progressively. Three weeks before death bilateral retrograde brachial and right carotid arteriographies gave no definite pathological results. The patient's mental state and degree of consciousness then rapidly deteriorated.

Autopsy showed pneumonia and bilateral dilatation of the heart. A large tumor occupied most of the left cerebellar hemisphere (Fig. 1) and partially invaded the medulla oblongata so that the left dorsolateral area was completely destroyed in one plane (Fig. 2). The pons had shifted to the right side and showed invasion of the lateral areas. The tumor had the typical macroscopical and histological appearance of a *glioblastoma,* including high cellularity, poor differentiation, marked pleomorphism, frequent mitotic figures, necroses with pseudopalisading and vascular proliferation. Tumorous destruction of the ventral half of the dentate nucleus caused hypertrophy of the dorsal parts of the contralateral inferior olive.

Discussion

The reason for the very rare occurrence of glioblastomas in the cerebellum is unknown and cannot be explained by its smaller size compared with the cerebral hemispheres (Jänisch et al. 1976). Dedifferentiating astrocytomas of the cerebellum are somewhat more common, but usually have a longer clinical history and higher incidence in younger people. The age and short clinical history of our patient are in accordance with previous reports of glioblastomas both in the cerebellum and cerebral hemispheres.

The "lateral medullary syndrome" was first clinically delineated by Wallenberg in 1895 and ascribed to occlusion of the posterior inferior cerebellar artery (1901). Later reports found occlusion of the vertebral artery to be more frequent. Recent angiographic studies state that Wallenberg's syndrome is not specific to any particular site of vascular occlusion (Janzen et al. 1979).

Only a few cases of Wallenberg's syndrome are found, at autopsy, to be caused by processes other than vascular lesions: metastatic tumors, especially carcinoma of the lung and melanoma were found (Davison and Spiegel 1945; Ogawa et al. 1977; Ho 1980; McFarland and Truscott 1961); Volk et al. (1973) reported 'late' radiation necrosis after radiation therapy given against chemodectoma of the middle ear; localized inflammatory changes interpreted as metastatic bacterial encephalitis were found by Quast and Liebegott (1975); Barnett and Hyland (1952) dscribed a case of Wallenberg's syndrome which proved to be caused by a congenital varix at autopsy.

Wallenberg's syndrome consists of numerous neurological signs and symptoms varying slightly from case to case (Currier 1969; Walton 1977). The given ensemble of these signs and symptoms depends more or less on the variable site and size of the brainstem lesion as pointed out by Fisher et al. (1961), analyzing 16 cases of

Wallenberg's syndrome, and by Currier et al. (1961), comparing the clinical symptomatology of 39 cases. If there are additional symptoms and signs pointing to a more rostral involvement of the brainstem, the lesion is suspected not to be of vascular origin. Sudden onset of symptoms does not exclude a tumorous lesion, as seen in our case and in the majority of those cited above.

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