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Giant-Cell Glioblastoma of the Thoracic Cord

By

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With 3 Figures

Summary

A case of giant-cell glioblastoma occurring in the upper thoracic cord of a male aged 76 without evidence of intracranial lesion is reported. Partial tumour resection with decompression and radiotherapy did not improve the patient's neurological deficit.

Keywords: Intramedullary tumour; giant-cell glioblastoma.

Malignant astrocytomas (grades III and IV)^{10, 25} arising in the spinal cord are rare^{9, 17-19}. In 1971 Fortuna and Giuffrè⁶ collected 22 cases from the literature and added four personal observations. Among the available case reports, only two^{16, 17} were classified as monstro-cellular glioblastoma. A further example of primary anaplastic glioma of the thoracic cord with the features of giant-cell glioblastoma is described in the present report.

Case Report

Case History. This 76-year-old white male had been well until 9 months prior to hospital admission, when he noted slowly progressive weakness of the legs with hypaesthesia below the sternum. Neurological examination showed paraparesis of the legs and a sensory level at T8. A myelogram revealed an incomplete block at T3 (Fig. 1), and subsequent laminectomy from T2 to 4 showed the spinal cord segments T2-T4 swollen by a bluish, ill-delimited intramedullary tumour, which was partly resected by microsurgery.

Pathological Examination (N 275-80). All tumour specimens, altogether approximately 3 mm in diameter, were embedded in paraffin, and stained by routine methods. Histologically, the highly cellular and pleomorphic tumour was mainly composed of large protoplasmic astrocytes with large numbers of giant and multinucleated cells (Fig. 2). Their eosinophilic cytoplasm showed coarse thick processes. The nuclei were huge with variable chromatin distribution, and there were occasional cytoplasmic invaginations or "intranuclear inclusions"

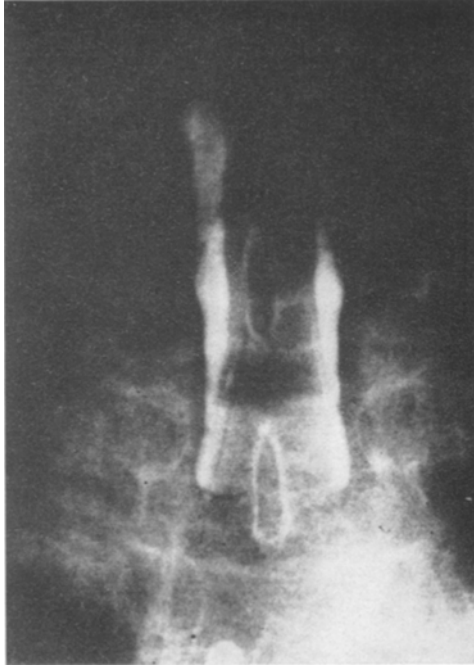


Fig. 1. Myelography showing incomplete block at T 3

(Fig. 3), while the number of mitotic figures was small. There were some small areas of necrosis and obvious vascular proliferation. No intact spinal cord tissue was seen. The diagnosis of giant-cell or monstrocellular glioblastoma grade IV was made and confirmed by the Brain Tumour Reference Centre of the German Association of Neuropathology.

Clinical Course. Postoperatively the condition of the patient was not changed. He received irradiation by betatron (4950 cGy) over four weeks directed locally at the tumour site. Despite the decompression and radiotherapy the patient's symptoms progressed over the subsequent two months to the point where there was almost complete motor and sensory deficit below the T 8 spinal cord level. No control myelogram was performed, and spinal CT scan was not rewarding. No systemic chemotherapy was given. At the time of writing there is slight improvement in the patient's motor and sensory deficit, and there is no evidence of intracerebral lesion.

Discussion

While glioblastoma multiforme accounts for more than 50 per cent of all primary intracranial gliomas^{9, 18, 19}, it accounts for only 0.1 to 5.8% of all spinal cord tumours^{9, 11, 17, 18, 20}, with an average of 1 to 2 per cent. In 1971 Fortuna and Giuffrè⁶ estimated the total number of intramedullary glioblastomas at about 60, but the subject

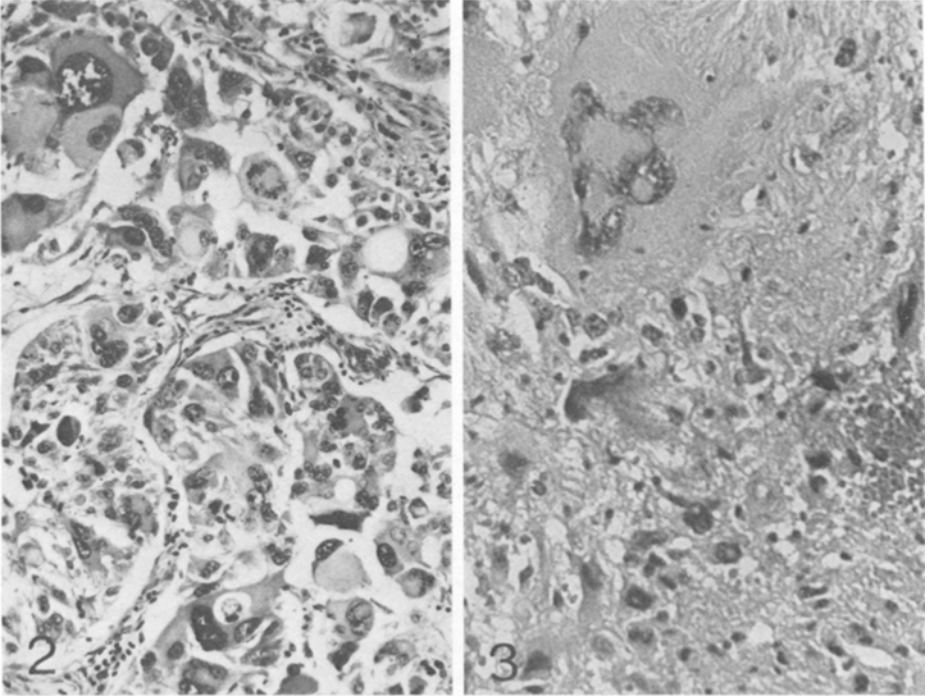


Fig. 2. Histology of intramedullary biopsy specimen showing monstrocellular glioblastoma with multinucleated giant cells and occasional mitoses. C. V. $\times 140$

Fig. 3. Detail of tumour showing bizarre multinucleated giant cell with cytoplasmic invaginations. H. and E. $\times 540$

is difficult to demarcate precisely because of the diversity of the nomenclature used for high grade gliomas. Some additional instances of intramedullary glioblastoma have been reported^{13, 16}. In a recent autopsy series of 288 glioblastomas among 1,491 CNS tumours there were 8 spinal medullary instances¹⁹. Intramedullary glioblastoma occurs at any age and not rarely affects younger subjects⁶; however, among 900 spinal tumours in infancy and childhood there were only 4 glioblastomas, or 0.5%¹¹. They may affect all levels of the

cord with slight preference for the cervical and thoracic segments^{6, 18}. Their common clinical features are: a short history with occurrence of pronounced neurological symptoms frequently progressing to complete motor and sensory deficit, often markedly increased CSF protein, negative plain X-rays of the spine but positive myelography, and a poor prognosis, irrespective of the extent of surgical removal and attempts at radiation and chemotherapy^{6, 16}.

The histological appearance of intramedullary glioblastoma corresponds to that of the glioblastoma described in the cerebral hemispheres, but the monstrocellular variant appears to be rare in the spinal cord. Recent ultrastructural and immunocytochemical studies have supported the astrocytic origin of this type of tumour^{3, 7}, which should be called a giant-cell glioblastoma. The irregular and most grotesque multinucleated giant cells, which were shown to have a slow rate of proliferation⁸, may occur spontaneously or as a result of some degeneration showing less GFAP (glial fibrillary acidic protein) than well differentiated glial cells, suggesting an inverse relationship between GFAP content and the degree of malignancy³. The morphogenesis of intramedullary glioblastoma is unknown. While dissemination from primary intracranial glioblastoma²³ can be excluded in most reported cases of intramedullary glioblastoma, it has also been argued that it may represent a spinal manifestation of multifocal gliomas⁶. The overall incidence of multifocal glioblastoma ranges from 2.5 to 6 per cent^{9, 18}, and their occurrence in both the cerebral and cerebellar hemispheres is well established^{9, 18, 21}. Multiple gliomas have been observed in neurofibromatosis¹². Some intramedullary glioblastomas, mainly in children, were associated with other intraspinal tumours⁶ or with intramedullary cavities, syringomyelia, and other malformations of the spine and cord^{2, 5, 22}. While invasion of the meninges and dissemination into the cerebral CSF pathways is rarely seen in isomorphic gliomas of the spinal cord^{4, 14, 15, 20, 24}, intramedullary glioblastoma, like its cerebral counterpart, often spreads into the CSF pathways with occasional intracranial seeding^{1, 4, 6, 18, 20}. In our patient, there was no evidence of intracranial lesion at the time of writing. While recent results of multimodality treatment of malignant cerebral gliomas give some encouraging results, the prognosis of intramedullary glioblastoma is extremely poor, and apparently very little experience of the effects of irradiation and chemotherapy of these neoplasms is available.

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