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Multiple Intraparenchymal Schwannomas in the Cerebellum, Brainstem, and Cervical Spinal Cord

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Introduction

The authors report a patient with multiple intraparenchymal schwannomas occurring in both the brain and the cervical spinal cord, and describe the radiological findings and histopathological examination with immunohistochemical staining.

Case Report

This 29-year-old woman was admitted with a three-year history of gradually worsening gait disturbance. Physical examination revealed no stigmata of neurofibromatosis. On neurological examination, there were nystagmus, right facial hypaesthesia, right hearing disturbance, decreased gag reflex, and right leg weakness. Magnetic resonance (MR) imaging showed multiple round intraparenchymal masses, located in the cerebellar hemisphere, the brainstem, and the cervical spinal cord (Fig. 1). After performing a retrosigmoid suboccipital craniectomy, a pinkish protruding mass from the brainstem was found in the right cerebellopontine angle below the 8th cranial nerve. The tumour was found to be in contact with the brain stem, and easily separated from the lower cranial nerves, which were displaced downward. Eight months after subtotal removal, she died of aspiration pneumonia. Histopathological findings were compatible with schwannoma. In immunohistochemical staining, the tumour cells were found to be positive for S-100 protein and negative for GFAP and Masson-Trichrome staining.

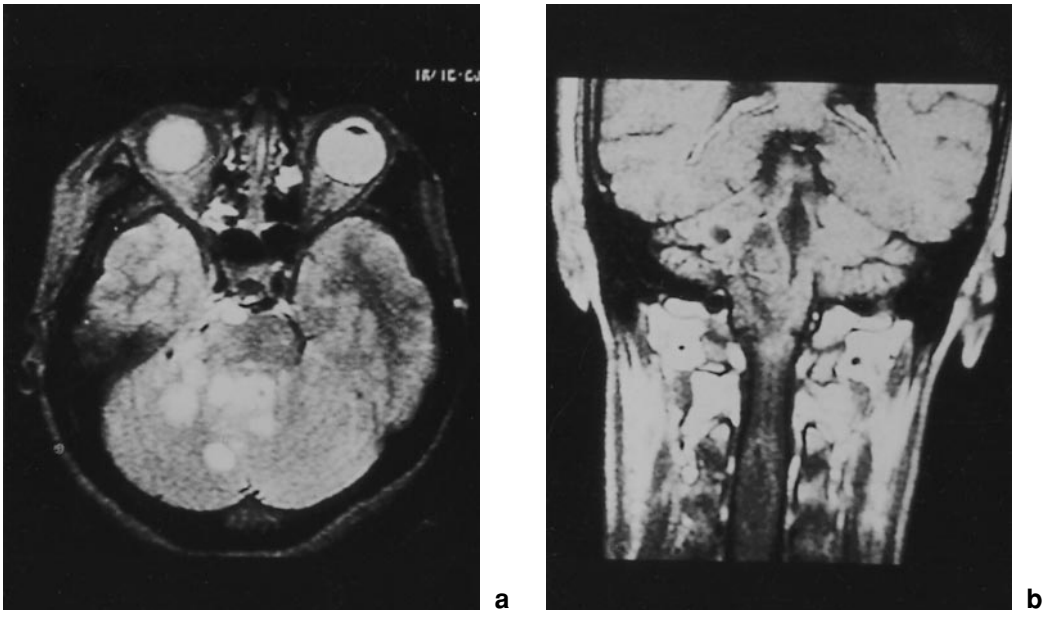

Fig. 1. Magnetic resonance imaging. (a) T2-weighted axial image showed multiple high signal intensity masses in the brain stem, cerebellar hemisphere and cerebellopontine angle. (b) T1-weighted sagittal and coronal image showed iso- or low signal intensity masses in the brain stem and cervical spinal cord

Discussion

After Gibson *et al.* published the first recorded case [2], 38 cases of intracerebral schwannomas have been reported. However, multiplicity has never been documented in intracerebral schwannomas. Ross, *et al.*, summarized the various theories proposed to explain the unexpected intramedullary location of these tumours [3]. These include: 1. hyperplasia of Schwann cells in the perivascular plexus penetrating the CNS, 2. neoplastic growth of Schwann cells at the nerve root entry zone, 3. the presence of aberrant intramedullary peripheral nerve fibers, 4. aberrant differentiation of primitive multipotential mesenchymal element, and 5. conversion of pial cells of neuro-ectodermal origin into Schwann cells. The authors have argued in favour of a developmental aetiology for intracerebral schwannoma occurring in younger patients versus a non-developmental pathogenesis of intraspinal tumors in older patients [1, 4]. Brainstem schwannomas, however, appear to be in a category intermediate between the two. In our case, however, the origin of schwannomas remains unclear because multiple intraparenchymal schwannomas occurred in both of the brain and the spinal cord.

There are no pathognomonic neuro-imaging features of intracerebral schwannomas. Immunohistochemical staining has been crucial in distinguishing these benign lesions from malignant glial tumours and metastatic lesions. Especially in the event of multiple lesions, metastasis and infectious disease should be considered in the differential diagnosis.

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