Multiple Intramedullary Schwannomas
—Case Report—

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Abstract

A 15-year-old boy with von Recklinghausen's disease presented with two very rare symptomatic cervical intramedullary schwannomas manifesting as slowly progressive weakness. The tumors were totally resected and he made a complete recovery.

Key words: intramedullary schwannoma, neurofibromatosis, spinal cord neoplasm

Introduction

Von Recklinghausen's disease (neurofibromatosis type 1: NF1) affects approximately 1,000,000 people at any given time.25 Despite the frequent presence of peripheral neurofibromas in NF1, an intramedullary location is rare, with reports of only four solitary spinal intramedullary schwannomas in patients with NF1,4,10,13,14 and only one case of multiple intramedullary schwannomas in a patient without associated neurofibromatosis.5 The occurrence of nerve sheath tumors in an intramedullary location raises important questions regarding the histogenesis. Preoperative recognition of the possibility of a benign intramedullary schwannoma may have therapeutic implications, as radical resection could lead to a cure.

We describe a patient with von Recklinghausen's disease harboring two symptomatic cervical intramedullary schwannomas.

Case Report

A 15-year-old boy presented with progressive weakness and stiffness of all four extremities which had developed over 1.5 years. The disability had progressed to the extent that he was unable to walk without assistance and could not perform routine activities with his hands. There were no urinary or bowel dysfunctions. He had had a squint in the left eye since birth.

Neurological examination revealed a fixed and dilated left pupil (6 mm). The adduction movement of the left eye was absent. There was no ptosis. He manifested spastic quadriplegia. The left temporomandibular joint was ankylosed with hypoplastic mandibular ramus. There were multiple large café au lait spots all over the body. Magnetic resonance (MR) imaging with gadolinium revealed two discrete intramedullary masses located close to each other, the larger at the third and fourth cervical segments measuring 29 mm in maximum diameter, and the

Fig. 1 Sagittal T1-weighted MR image with gadolinium showing two distinct intramedullary tumors, the upper located at the third and fourth cervical levels and the lower at the fifth cervical level.
smaller at the fifth cervical segment measuring 9.8 mm in maximum diameter (Figs. 1 and 2). MR imaging of the brain revealed a small nodular lesion at the left temporal pole which was hypointense on T1 and hyperintense on T2-weighted images. Computed tomography showed the left mandibular hypoplasia.

A wide laminectomy was followed by midline myelotomy over the maximum bulges produced by the entirely intramedullary lesions. Two firm encapsulated tumors were excised after initial debulking. Both tumors were only mildly vascular and could be relatively easily dissected from the adjoining neural structures.

Histological examination of the tumor specimens revealed relatively uniform cells with ovoid nuclei arranged in palisading fashion with Antoni type A and B cellular patterns (Fig. 3). The histology of both tumors was exactly the same.

Postoperatively he showed progressive improvement and recovered almost completely over 2 months.

Discussion

Only 35 cases of intramedullary schwannomas have been reported, including only one case of multiple intramedullary schwannomas without evidence of NF1, and four cases of solitary intramedullary spinal schwannomas in patients with NF1. Multiple intramedullary ependymomas and astrocytomas in patients with NF1 have been described. Patients of neurofibromatosis have harbored many dysplastic lesions involving Schwann, meningeal, neuroglial, and ependymal cells. Intramedullary schwannosis containing foci of Schwann cells within the spinal cord in the Lissauer's zone, which encloses intrinsic nerve fibers, are more frequently encountered in patients with NF1. Such cells are the probable origin of these rare tumors. Another possible origin for intramedullary schwannoma is perivascular schwannosis or foci of Schwann cell proliferation occurring adjacent to the anterior spinal artery and its branches, which is also common in patients with NF1. Schwannosis is differentiated from intramedullary schwannoma on the basis of presence of abundant axons and myelin sheaths situated around the anterior spinal artery, absence of verocay bodies and nuclear palisading, microscopic dimensions, and the specific absence of clinical manifestations. Various theories have been proposed to explain the intraparenchymal presence of Schwann cell tumors, including distorted embryogenesis, misplaced myelinated fibers, displaced neural crest cells, and multipotency of mesenchymal cells.

Our patient had various atypical features including multiple stigmata of NF1, multiple intramedullary schwannomas, left temporomandibular joint pseudoarthrosis, hypoplasia of the left ramus of the mandible, and partial left third cranial nerve paresis. Association of all these features with NF1 is rare. The benign nature of this tumor indicated the need for radical tumor excision despite the location in a surgically difficult area. The long duration of the symptoms and minimal deficits at the time of presentation suggested slow tumor growth. Preoperative MR imaging with gadolinium accurately delineated the tumor, but assumption of the histological nature of the lesion is difficult. At surgery the presence of a well-defined, usually solid mass raised the suspicion of a benign tumor. Accurate interpretation of frozen sections during operation is important. Reoperation was required in a few reported...
cases after partial excision was achieved during the first surgery. Complete removal should be attempted in the first instance as these tumors tend to have good plane of cleavage. Histologically the tumor should be differentiated from microcystic meningiomas and pilocytic astrocytomas. The long-term prognosis after complete excision is good.

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