Management of Anteriorly Located C1-C2 Neurofibromata

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The authors discuss their recent experience with anteriorly located C1-C2 neurofibromata in five patients with cervical myelopathy and magnetic resonance scans consistent with intradural extramedullary masses in this region. Surgery was performed using a posterolateral approach with microscopic intradural exploration. Gross total intradural tumor removal was achieved in all cases. Improvement in cervical myelopathy occurred in all patients.

This report concludes that C1-C2 neurofibromata located anterior to the spinal cord can be totally and safely removed using a posterolateral approach. Improvement in neurologic dysfunction accompanies posterior decompression and gross total intradural tumor removal.

KEY WORDS: Cervical spine; Myelopathy; Neurofibromatosis

The diagnosis and management of neoplasms of the foramen magnum and upper cervical spinal cord have intrigued neurosurgeons for years. Frequently, these lesions resemble degenerative diseases of the central nervous system, resulting in diagnostic delay [3,4,6]. Surgical resection usually effects a cure, however, the optimal surgical approach to this region remains controversial [2,5,9,10,16].

Recent advances in neuroradiologic imaging allow for improved visualization of central nervous system abnormalities, thereby facilitating the rapid diagnosis of neoplasms in the region of the foramen magnum and upper cervical spine [1,13,14].

This report discusses the authors' recent experience with the diagnosis and management of anteriorly located C1-C2 neurofibromata.

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Material and Methods

Between 1984 and 1988, eight patients presented to the University of Michigan Hospitals with high cervical myelopathy and myelographic evidence for C1-C2 neurofibromata. Seven of these eight patients had neurofibromatosis. The most common symptoms included weakness, spasticity, and incoordination. Preoperative magnetic resonance (MR) scanning demonstrated anteriorly located intradural, extramedullary masses at the C1-C2 level in five of the eight cases (Figure 1).

Table 1 illustrates the clinical profile of these five patients. There were two women and three men and the average age was 41 years. All patients were ambulatory upon presentation. Patient GA was the only patient who did not have neurofibromatosis.

Surgery was performed with the patients placed in the prone position. A posterior midline incision was used and a wide C1-C2 laminectomy was performed. Additional lateral bony decompression was performed to the lateralizing side of the tumor, if present. All patients had extradural tumor as well; however, the intradural portion was removed first.

Upon opening the dura, the operating table was rotated 15° to 30° to facilitate exposure (Figure 2). Sectioning of the dentate ligaments and at times the posterior cervical nerve roots provided increased access to the tumor and increased mobility of the cervical spinal cord. The ultrasonic surgical aspirator was used to resect tumor in all cases (Figure 3).

In cases of gross total extradural tumor removal, the dural attachment was resected and a patch graft placed. Otherwise, the dural attachment was cauterized and repaired primarily.

Results

Gross total intradural tumor removal was achieved in all five patients. Pathological diagnosis was benign neurofibroma in all cases. Gross total extradural tumor removal was possible in three of five cases. One patient (FS) developed a spinal accessory nerve palsy which partially resolved. There were no cases of swan-neck deformity

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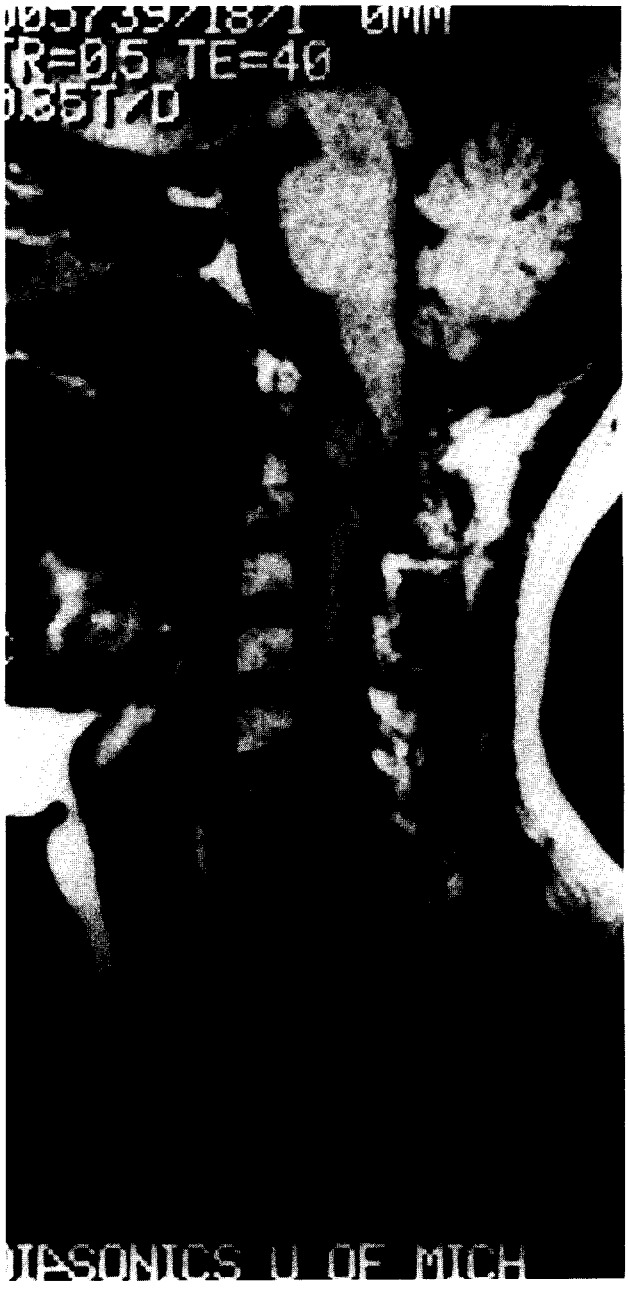

Figure 1. *Sagittal MR scan of patient GA demonstrating a mass located anterior to the spinal cord at C1-C2, with resultant anteroposterior compression.*

noted on postoperative x-rays. Improvement in myelopathy occurred in all five patients.

Discussion

One of the first reports regarding surgical management of upper cervical spinal cord tumors was by Elsberg in 1929 [4]. He described rotating the spinal cord on a divided slip of the dentate ligament to resect ventrally located tumor.

The Mayo Clinic has reported the largest series of patients, which has been divided into three consecutive reports. In 1956, Dodge et al [3] reported 30 benign tumors diagnosed between 1924 and 1956; 26 were meningiomas and 4 were neurofibromas. Yasuoka et al [16] reported the next series of 57 patients diagnosed between 1957 and 1976. This was followed by the comprehensive review of the Mayo Clinic experience of 102 total cases between 1924 and 1982 by Meyer et al [9]. There were 78 meningiomas, 23 neurofibromas, and 1 teratoma. The most frequent complaints were neck pain, ataxia, numbness, and incoordination. All patients were operated on in the sitting position using a posterior approach. Myelography and computed tomography were used when available.

Howe and Taren [6] described the pitfalls in diagnosing these lesions in six patients. They felt that a uniform clinical syndrome did not exist to describe the clinical manifestations of these tumors. Factors that were suggestive included cervical pain, upper motor neuron signs, and mixed sensory deficits.

Recently, Wagle et al [14] described the value of MR scanning in the diagnosis and management of four cases of foramen magnum meningioma. Prior to MR scanning, the radiological diagnosis of foramen magnum lesions was best imaged by myelography [7,8].

The incidence of central nervous system neurofibromata is increased in neurofibromatosis [11]. These lesions rarely undergo malignant degeneration [12]. The increased association between spinal column abnormalities and neurofibromatosis has been previously described [15,17].

In this report, seven of the initial eight patients had neurofibromatosis (88%), while in the comprehensive Mayo Clinic review, less than 50% of their patients with C1-C2 neurofibromas had neurofibromatosis [9]. Although these lesions are frequently difficult to diagnose, the increased incidence of neurofibromatosis in this re-

Table 1. *Clinical Profile of Five Patients with Anteriorly Located C1-C2 Neurofibromata*

	Age/Sex	Signs and symptoms	Year
MB	24 F	Right-sided weakness, incoordination	1984
GA	45 F	Upper extremity weakness, spasticity	1986
MG	44 M	Incoordination, spasticity	1987
FS	67 M	Weakness, ataxia, spasticity	1988
RE	25 M	Upper extremity weakness, lower extremity spasticity	1988

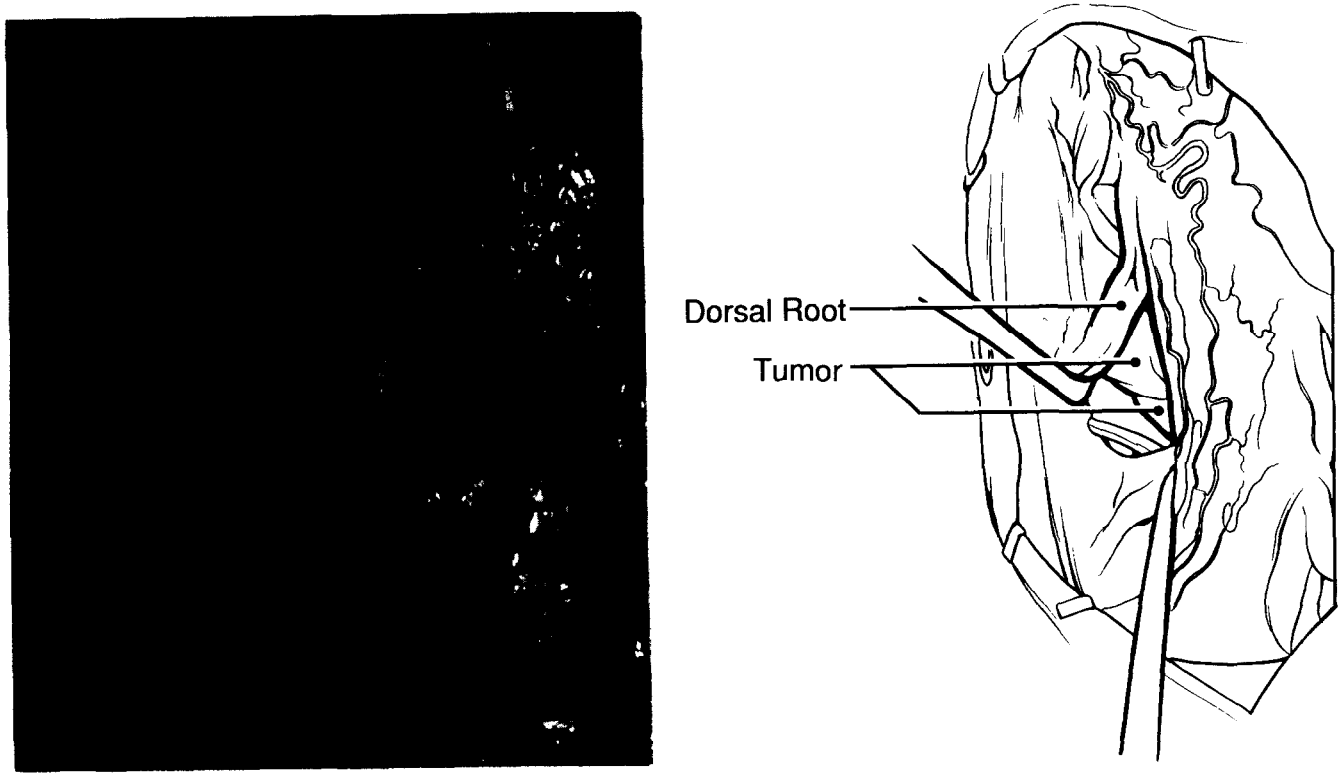

Figure 2. *Intraoperative view and diagram of patient FS demonstrating posterior and lateral exposure of an anteriorly located C1-C2 neurofibroma.*

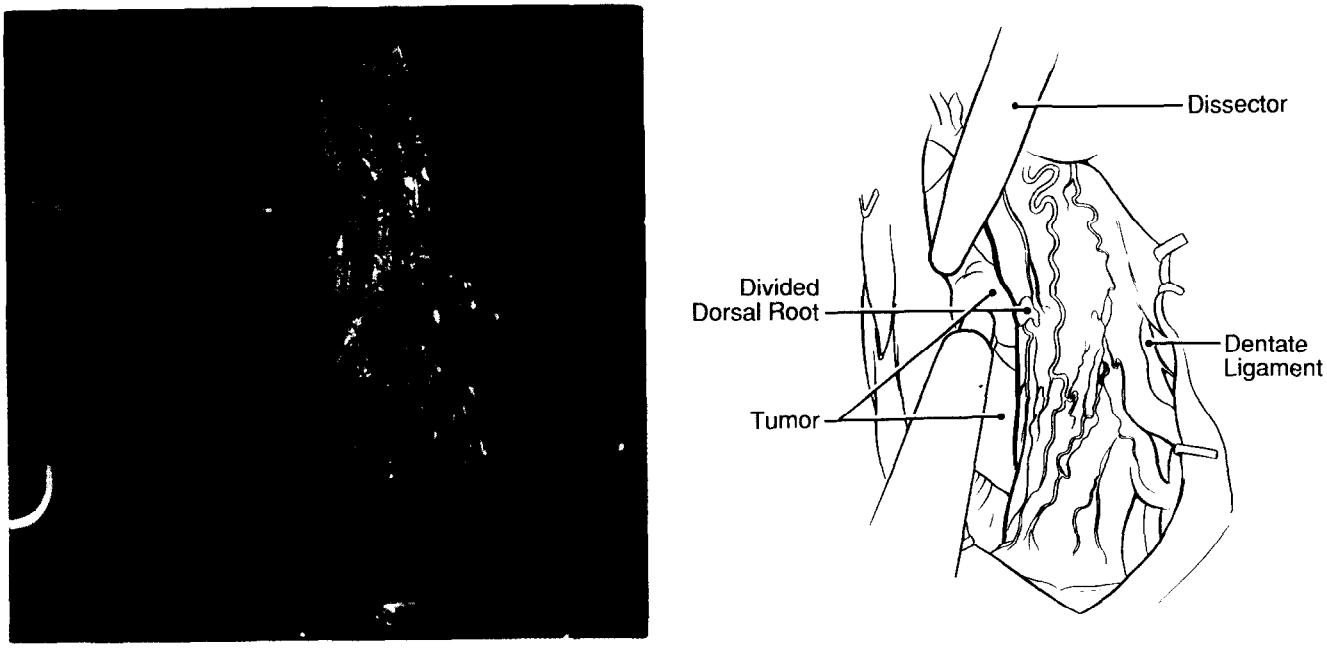

Figure 3. *Intraoperative view and diagram of patient FS demonstrating sectioning of the dorsal root of C2, gentle traction on the tumor, and ultrasonic surgical aspiration of the tumor.*

port resulted in early imaging with myelography and MR scanning, thereby facilitating diagnosis.

Gross total extradural tumor removal was possible in only three of five patients. In the remaining two patients, tumor was encasing the vertebral arteries bilaterally, thereby limiting total tumor removal. A thorough decompression was achieved in both patients, however.

Surgical resection of the anteriorly located tumor was facilitated through use of the ultrasonic surgical aspirator. Together with sectioning of the dentate ligaments, this instrument allowed for careful tumor debulking and removal in all five patients.

There were no postoperative spinal cord complications, and cervical myelopathy improved in all five patients. One patient developed a spinal accessory nerve palsy. This nerve was draped over tumor and presumably was injured due to manipulation. A more lateral exposure and approach may have avoided excessive manipulation, allowing for tumor debulking anterior to the nerve.

Despite the recent enthusiasm for anterior trans-oral approaches to this region of the spine and foramen magnum, this report demonstrates that certain anterior lesions can be safely approached and resected posteriorly [10].

Although MR scanning was invaluable in the initial work-up of this group of patients, it also proved useful in the follow-up management in three of five patients, including the patient who developed a spinal accessory nerve palsy.

This report concludes that (1) MR scanning is a sensitive and noninvasive means of imaging the upper cervical spine in patients with neurofibromatosis and cervical myelopathy, (2) C1-C2 neurofibromata located anterior to the spinal cord can be totally and safely removed using a posterolateral approach, and (3) improvement in neurologic dysfunction accompanies posterior decompression and gross total intradural tumor removal.

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