Extraspinal-Interdural Surgical Approach for C2 Neurinomas—Report of an Experience with 50 Cases

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OBJECTIVE: **We report our experience with surgery in 50 patients with C2 neurinomas. The anatomic subtleties of these discrete forms of tumors and their surgical implications are analyzed.**

METHODS: **During the period 2006–2016, we operated on 50 patients with 55 C2 neurinomas. Type A tumors were located within the spinal canal, type B tumors were located in the lateral gutter, and type C tumors had a paraspinal extension. By working within the dural confines of the tumor and appropriately angulating the microscope, the entire tumor bulk extending into 1 or all 3 compartments was resected. Follow-up duration ranged from 3 months to 10 years (mean 68 months).**

RESULTS: **This series included 36 male and 14 female patients. Age range of patients was 14–70 years (mean age 36 years). Progressive symptoms of myelopathy were present in 41 patients. There were 16 type A + B tumors, 27 type B tumors, 10 type B + C tumors, and 2 type A + B + C tumors. All patients experienced symptom improvement after surgery and were able to resume their normal lifestyle.**

CONCLUSIONS: **C2 neurinomas arise in the region of the C2 ganglion, and despite the fact that some achieve a large size, they remain confined within the dura. Radical tumor resection can be achieved by working within the layers of the dural cover. Bone removal and opening of spinal dura for tumor exposure and resection can be avoided.**

INTRODUCTION

In 2008, we evaluated our experience with 60 patients with C2 neurinomas treated between the years 1992 and 2006.[1] We presented a classification scheme of these tumors according to their anatomic location and nature of their relationship with the dural envelope.[1] On the basis of this experience, we speculated, for the first time in the literature, that if the anatomic and dural subtleties are appropriately understood, it is possible to resect these tumors without any bone removal.[1] In the present study we evaluate our subsequent experience, which provided newer insights regarding the dural relationships of these tumors. All tumors were resected by working within the dural confines of the tumor without any bone removal and without resorting to opening of the midline spinal dural tube. The clinical profile, anatomic peculiarities, and outcome of these discrete forms of benign spinal tumor are evaluated.

MATERIALS AND METHODS

We reviewed the clinical details of 50 patients with 55 C2 neurinomas from our database. These patients were surgically treated consecutively between 2006 and 2016. The clinical profiles were retrospectively analyzed.

RESULTS

Clinical Presentation

Of 50 patients with 55 C2 neurinomas, 45 had unilateral and 5 had bilateral C2 neurinomas. Six patients had evidence of neurofibromatosis 2 (NF2), including all 5 patients with bilateral C2 neurinomas. All patients with NF2 had multiple cranial, spinal, and extraspinal tumors.

The mean age of the patients was 36 years (range, 14–70 years). There were 36 male and 14 female patients. The clinical details are summarized in **Table 1**. The mean duration of symptoms at the time of presentation was 27 months (range, 1 month to 12 years). Mild to moderate myelopathy was present in 39 patients; they could walk unaided. Two patients had severe myelopathy and were bedridden. The remaining patients had either local symptoms, such as neck pain, or symptoms essentially unrelated to C2 neurinomas.

Key words
- C2 neurinoma
- Dura
- Neurofibromatosis

Abbreviations and Acronyms
NF2: Neurofibromatosis 2

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Table 1. Clinical Features of Patients with C2 Neurinomas

Clinical Features	Number of Patients (%)
Age range, years	
0–10	
11–20	2 (4%)
21–30	16 (32%)
31–40	17 (34%)
41–50	10 (20%)
>50	5 (10%)
Sex	
Male	36 (72%)
Female	14 (28%)
Duration of symptoms	
<1 month	6 (12%)
1–6 months	9 (18%)
6 months to 1 year	7 (14%)
1–2 years	12 (24%)
>2 years	16 (32%)
Presenting symptoms	
Neck pain	20 (40%)
Radicular pain	16 (32%)
Neck mass	3 (6%)
Myelopathy	41 (82%)
Sensory disturbances	25 (50%)
NF	6 (12%)

NF, neurofibromatosis.

Radiologic Findings

Erosion of the C2 pedicle and adjoining lamina and arch of the atlas (C1) were seen uniformly. On magnetic resonance imaging, the tumors were predominantly isointense on T1-weighted and hyperintense on T2-weighted sequences. The lesions generally had smooth contours, had a homogeneous appearance, enhanced uniformly, and were well delineated after contrast agent administration. None of the tumors had imaging evidence of hypervascularity. In none of the cases was the vertebral artery directly encased by the tumor mass.

As per our previously published classification, the tumors were divided into 3 types (Figures 1–6).[1] Type A neurinomas were located in the spinal canal. Type B lesions were located in the region of the C2 ganglion, posterior to the atlantoaxial articulation. Type C lesions extended in the paraspinal region. A vertical line joining the tips of transverse processes of C1 and C2 formed the arbitrary line that demarcated type C lesions from type B lesions. Larger lesions extended from 1 compartment to the other. All tumors had a component of type B. Table 2 shows the extensions of the tumor as per this classification scheme. All tumors had a dural covering as seen in the line drawing

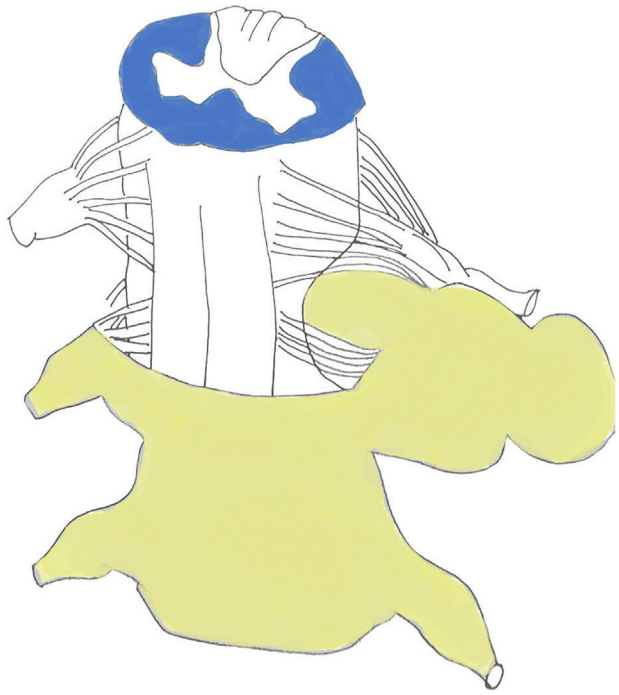

Figure 1. Line drawing showing the concept of dural configuration of tumors. As can be appreciated, the intraspinal component of the tumor is also interdural.

(Figure 1). The tumors ranged in size from 5 to 72 mm (mean 35 mm). In the spinal canal, all tumors extended anterior or anterolateral to the cord. Posterior extension of the tumor in relationship to the cord was never encountered. All types of tumors were located essentially within the dural confines. Even type A tumors had a well-defined sheath of dura that covered the entire dome of the tumor. The pedicle of C2, facets of C1 and C2, and adjoining posterior arch of the atlas and lamina of C2 were uniformly eroded. The adjoining tissues in the region, such as the venous sinuses, and the vertebral artery were displaced but were never encased by the tumor bulk.

Surgery

A midline surgical approach was undertaken. The spinous process of the C2 vertebra was the primary landmark of surgery. The exposure included suboccipital bone, arch of atlas, C2-C3-spinous process, and lamina on the side of the tumor. Sectioning of the muscles attached to the C2 spinous process and exposure of the contralateral side laminae were avoided. The exposure was then extended laterally on the side of the tumor. By working underneath the lateral aspect of the C1 arch and in the lateral gutter, the tumor bulge was first identified and then widely exposed.

A well-defined layer of membrane that was continuous with the spinal dura covered the tumor bulge. The tumors were generally firm and moderately vascular. The second cervical nerve root and ganglion were inseparably merged into the tumor confines. No attempt was made to save the root or the ganglion. The dural

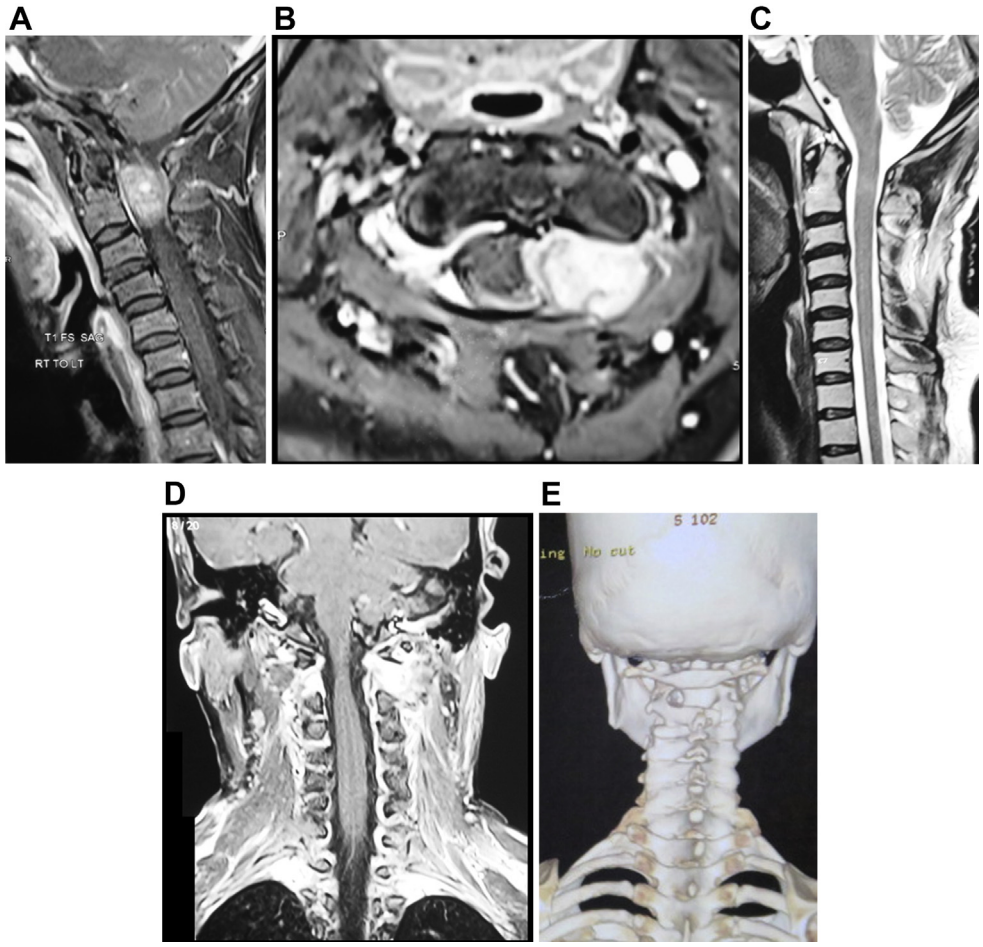

Figure 2. Magnetic resonance imaging and computed tomography of a 46-year-old woman. (**A**) T1-weighted sagittal contrast image showing enhancing tumor at C2 level. (**B**) T1-weighted axial contrast image showing type A + B tumor. (**C**) Postoperative T2-weighted sagittal image showing excision of tumor without any bone removal. (**D**) Postoperative coronal contrast image showing total excision of tumor. (**E**) Postoperative reconstructed computed tomography image showing preservation of the bone elements.

membrane cover provided a firm and reliable dissection plane between the tumor and the adjoining vertebral artery on the lateral aspect and spinal cord on the medial aspect. The prominent venous plexus in the region of the lateral gutter was displaced outside the confines of the tumor bulge. By progressive debulking of the tumor and by remaining confined to "interdural" space, the entire tumor in all the compartments was resected (Video 1).

In 3 cases with type A tumor, the midline spinal dural tube was opened in the process of tumor removal, which resulted in cerebrospinal fluid egress into surgical field. However, there was no need to make a separate incision in the spinal dural tube to resect the spinal canal component of the tumor. A shredded muscle piece was packed and stabilized with Gelfoam (Pfizer, New York, New York, USA) at the site of cerebrospinal fluid egress during closure in these cases. No dural suturing was necessary during closure in any case.

Video

Video available at WORLDNEUROSURGERY.org

Postoperative outcome was uneventful in all cases, and clinical symptoms improved in all patients after surgery. The tumor was completely resected in all cases. During the follow-up period (range, 3–120 months; average 68 months), there have been no cases of tumor recurrence.

DISCUSSION

The C2 ganglion is the largest spinal ganglion.[1,2] Among all ganglia (cranial and spinal included), the C2 ganglion is next in size only to the Gasserian ganglion.[3,4] Although other spinal ganglia are located in the region of spinal and root canal, the C2 ganglion has exterior exposure and is located outside the spinal canal and posterior to the atlantoaxial articulation. Although all other spinal neurinomas are located within the confines of bone, C2 neurinomas are exposed posteriorly.

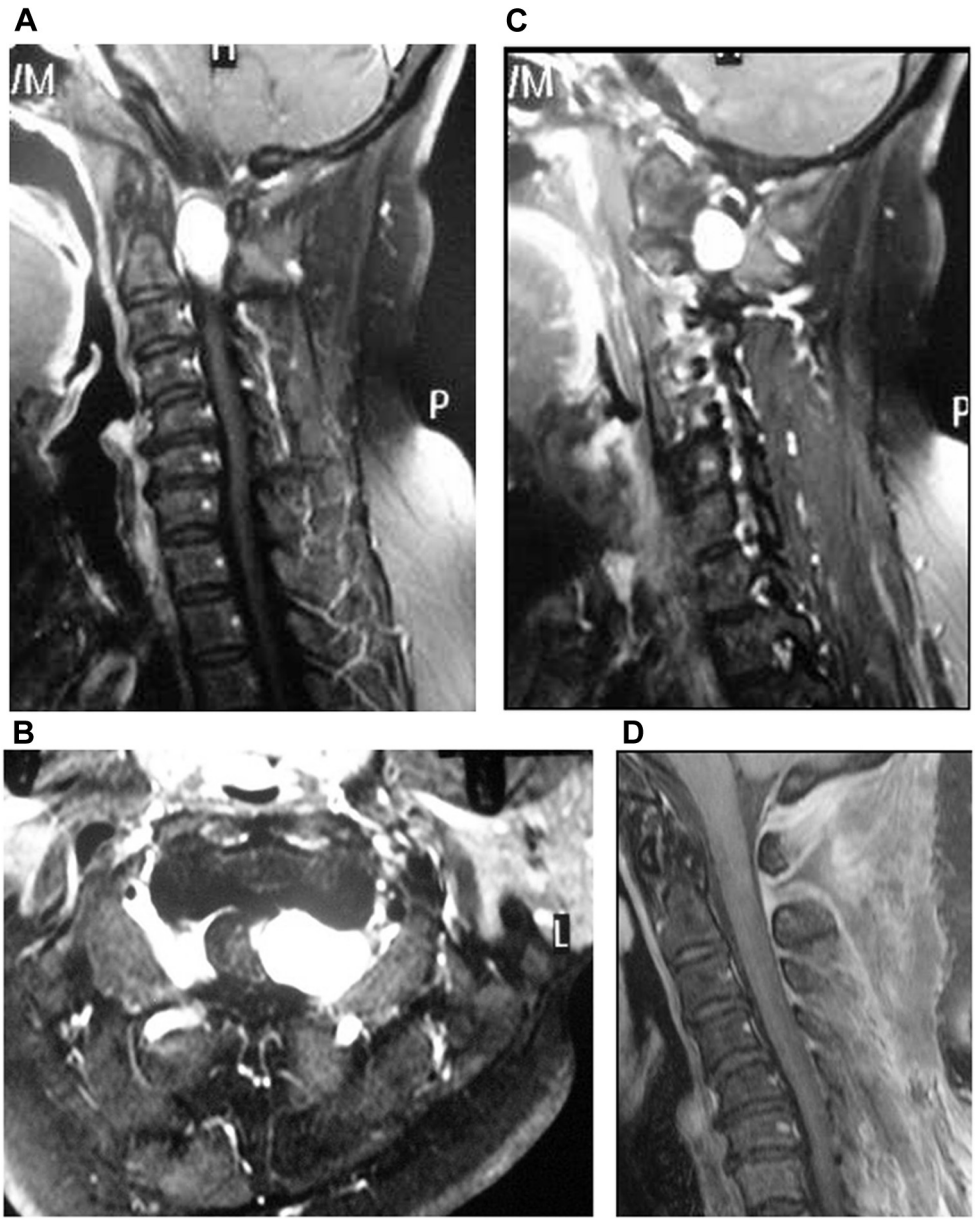

Figure 3. Magnetic resonance imaging of a 50-year-old woman. (**A**) Sagittal contrast-enhanced image showing C2 neurinoma. (**B**) T1-weighted axial contrast image showing type A + B tumor. (**C**) T1-weighted sagittal contrast image showing location of tumor behind the C2 facet. (**D**) Postoperative contrast-enhanced sagittal image showing excision of tumor.

C2 neurinomas are relatively common benign surgical lesions. C2 neurinomas are the most common spinal neurinomas.[1,5] Frequently, C2 neurinomas are associated with NF2.[6,7] Bilateral C2 neurinomas are common in cases associated with NF2. In cases with NF2, C2 neurinomas may be associated with several other cranial and spinal neurinomas. Their location, anatomic relationships, and dural configuration are characteristic making preoperative diagnosis possible and treatment of an otherwise formidable appearing and anatomically located lesion a rather straightforward surgical procedure with gratifying results. We recently identified the dural relationship of C2 neurinomas and evaluated its surgical implications.[1] We first reported the possibility of resection of these tumors without resorting to any bone removal for tumor exposure and resection.

The tumors are divided into 3 types. Type A neurinomas are located in the spinal canal, type B lesions are located in the region of the C2 ganglion, and type C lesions are located outside the spinal canal. A vertical line joining the transverse processes of C1

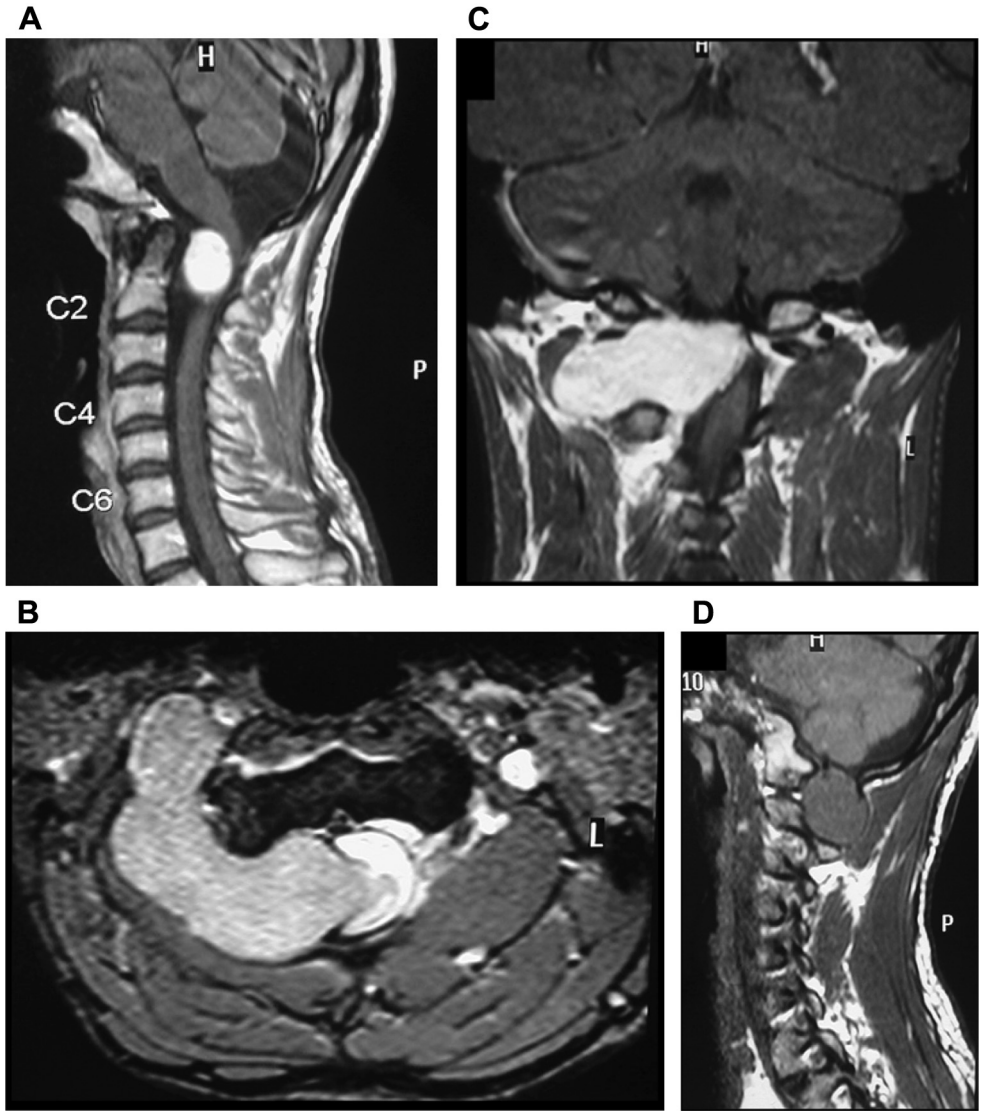

Figure 4. Magnetic resonance imaging of a 28-year-old man. (**A**) T1-weighted sagittal contrast image showing enhancing tumor at C2 level. (**B**) Axial contrast image showing type A + B + C tumor. (**C**) Coronal image showing extensions of tumor. (**D**) T1-weighted sagittal image showing location of tumor behind C2 facet.

and C2 formed the arbitrary line that demarcated type C lesions from type B lesions. Larger lesions involve the spinal canal and the lateral gutter of C1-2 (type A + B) or involve the lateral gutter and the extraspinal space (type B + C). Tumors can also be a combination of all 3 types (A + B + C). All tumors had their main bulk in the region of the ganglion or had type B anatomic location. From our experience in the present series and our earlier reported experience,[1] it appears that the site of origin of C2 neurinomas is from the region of the C2 ganglion. After its origin from the ganglion, the tumor extends medially (type A) and/or laterally (type C). Although the final judgment of dural classification can be made only by direct operative inspection, our experience in dealing with such tumors suggests that if clinical and radiologic parameters are suggestive of a lesion being C2 neurinoma, interdural location of the tumors can be taken for granted, and surgical planning can accordingly be done.

Figure 1 shows the nature of dural coverings of these tumors. This pattern of dural relationship is in variance with the pattern observed by us earlier. In our previous report, we identified 5 type A C2 neurinomas (of 60 cases). In the present series, we have identified that all type A tumors had an extension toward the lateral masses of atlas and axis and posterior to the atlantoaxial articulation, or were type A + B or type A + B + C. We had observed that type A tumors or type A extension of

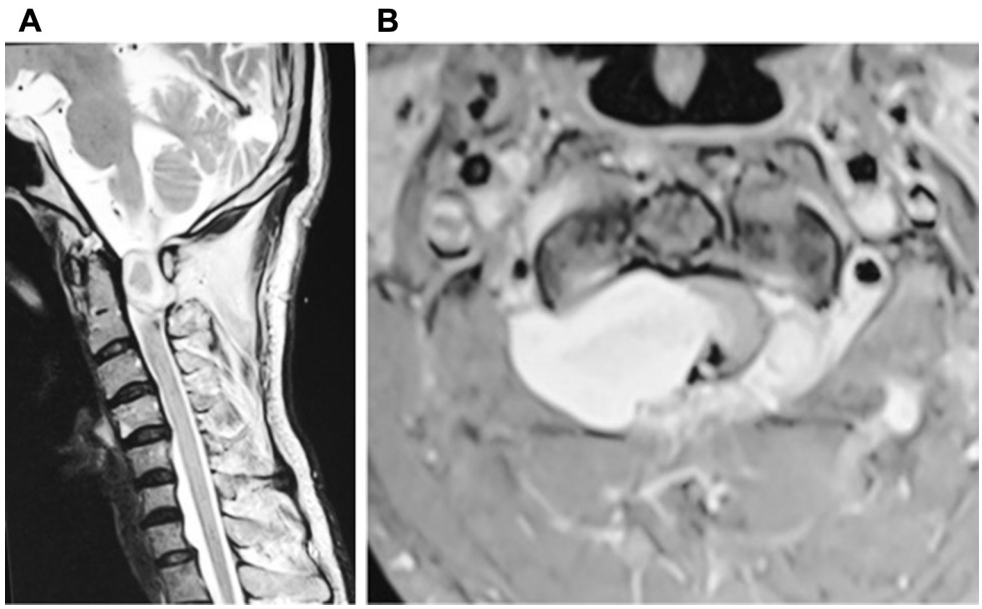

Figure 5. Magnetic resonance imaging of a 33-year-old man. (**A**) T2-weighted sagittal image showing C2 neurinoma. (**B**) T1-weighted axial contrast image showing type A + B tumor.

larger tumors (type A + B or type A + B + C) were essentially intradural in nature or were located in the subarachnoid spinal compartment.[1] However, we now have identified that even type A tumors or type A extension of larger tumors had a dural cover that was continuous with the spinal dura and with the dural cover of the extraspinal component of the tumor. Essentially, with this understanding, the entire tumor could be resected by working in the interdural compartment without resorting to opening of the spinal dura and by avoiding working in the intradural or subarachnoid compartment. Such dural relationships are remarkably similar to relationships seen in trigeminal neurinomas that arise from the Gasserian ganglion, the largest ganglion of the body. We had identified interdural location of posterior cranial fossa, middle cranial fossa, and extracranial compartmental trigeminal neurinomas and accordingly tailored the surgical strategy.[3,8] It was speculated that the origin of trigeminal neurinomas was from the Gasserian ganglion.[3,8]

No special attempt was made to save the uninvolved fibers of the C2 ganglion or the nerve roots. This was based on our experience with sectioning of the C2 ganglion for exposure of atlantoaxial articulation for stabilization procedures and the observation that except for moderate disability related to hypoesthesia in the suboccipital region, such a procedure does not result in any significant neurologic sequelae.[2,9,10]

Although several lateral, far lateral, and extreme lateral approaches are possible and have been recommended for these anterolaterally located foramen magnum tumors,[11-15] understanding of the dural configuration can make a posterior midline approach most suitable, effective, and remarkably quick.[1,16] Understanding of the fact that C2 neurinomas are interdural in their location has surgical implications, as dissection is facilitated by the presence of dura, and direct handling of critical spinal cord and vertebral artery can be avoided by limiting the dissection by the plane provided by the dura. Moreover, the surgical exposure can be reduced. The tumors can be resected by working posterior to the lateral masses of atlas and axis without removal of any bone of the spinal canal. No control of the vertebral artery is required during surgery, and tumor resection can be done within the dural plane that separates it from the vertebral artery. The tumor bulge in the region compresses the large adjoining venous plexuses, a fact that results in minimum venous bleeding during the tumor resection within the dura. It is impossible to decipher on imaging if a type A tumor is entirely or partially intradural or entirely interdural in nature. Although not done in the present series, if the tumor in the intraspinal canal is significantly large, partial laminectomy of C2 and resection of the lateral half of the arch of the atlas can be done to achieve an enhanced exposure. Dural opening may be necessary when intradural extension of the tumor either is suspected or is confirmed during surgery. Wound closure is quick, as dural suturing is not necessary.

The articular pillars of the atlas and axis were eroded to a varying extent in all tumors, but the joints appeared functional and intact. Because the facet joints were anterior to the tumor, they were not exposed, manipulated, or resected. Immediate or delayed spinal instability was not encountered in any case. Although not done in this series, if there is any concern regarding postoperative spinal instability, lateral mass screw and plate fixation may be performed after tumor resection, as the facets are already widely exposed after tumor resection.[9]

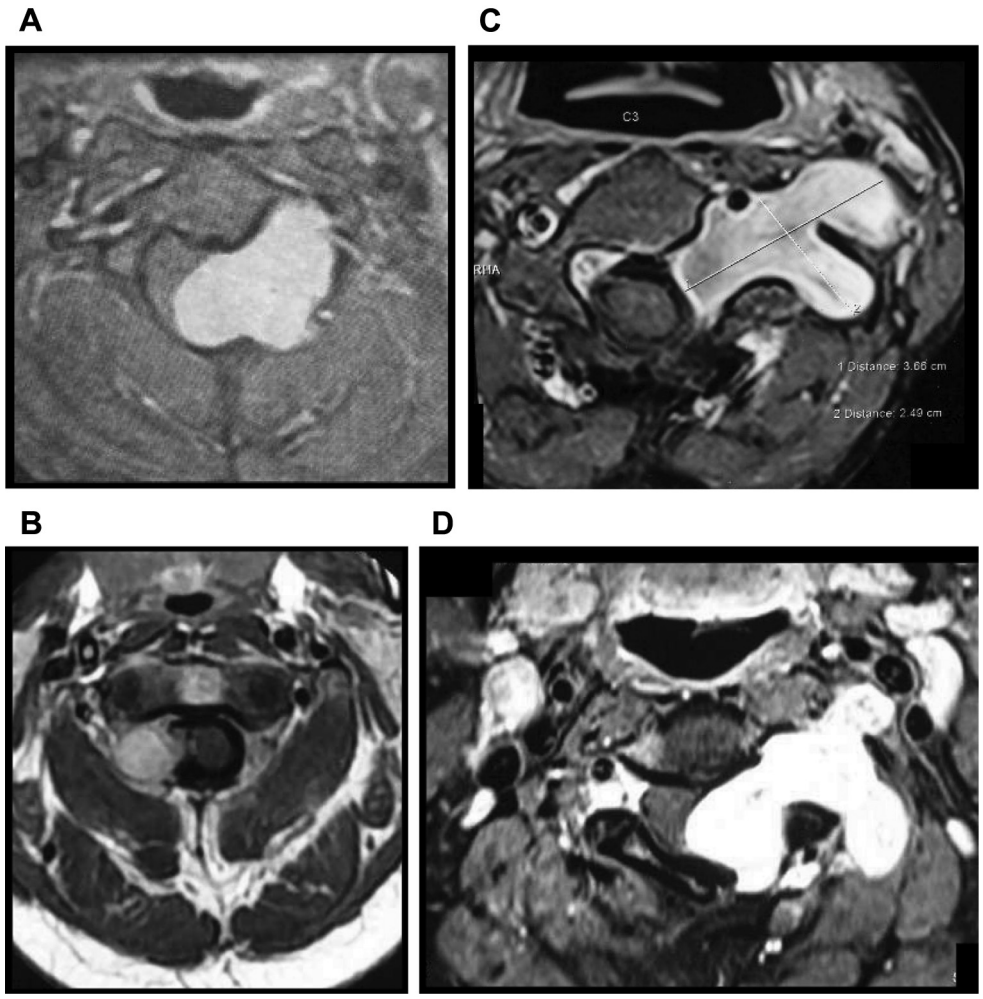

Figure 6. Axial magnetic resonance imaging showing the various tumor types according to the classification. (**A**) Contrast-enhanced axial image showing tumor extending into the spinal canal and in the lateral gutter in the region of C2 ganglion (type A + B tumor). (**B**) Axial image showing tumor confined to the lateral gutter in the region of the C2 ganglion (type B tumor). (**C**) Axial image showing tumor in the region of the C2 ganglion and extending laterally in the paraspinal region (type B + C tumor). (**D**) Axial image showing extensions of tumor in the spinal canal, in the lateral gutter, and in the paraspinal region (type A + B + C tumor).

Table 2. Radiologic/Surgical Classification Showing Extensions of Tumor

Type of Tumor	Number of Patients
A	—
A + B	16
B	27
B + C	10
C	—
A + B + C	2

The clinical neurologic outcome of surgery of these essentially benign tumors is gratifying. On the basis of our current experience and earlier reported experience, it appears that tumor recurrence rates of C2 neurinomas are extremely low, even after a partial or subtotal resection.

CONCLUSIONS

With understanding of the dural anatomic subtleties, the surgical exposure of C2 neurinomas can be minimized, and bone removal and spinal dural incision can be avoided. The vertebral artery and the venous sinuses in relationship with the tumor bulk are displaced, and the dural membrane forms a reliable surgical plane of dissection.

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