Intraneural Granular Cell Tumor of a Cervical Dorsal Nerve Root: A Case Report and Review of the Literature

Ron Ron Cheng[1], Jessica A. Forcucci[2], Stephen P. Kalhorn[1]

Key words
- Cervical spine
- Granular cell tumor
- Nerve sheath tumor
- Spinal nerve root

Abbreviations and Acronyms
GCT: Granular cell tumor
MRI: Magnetic resonance imaging

Departments of [1]Neurosciences and [2]Pathology and Laboratory Medicine, Medical University of South Carolina, Charleston, South Carolina, USA

To whom correspondence should be addressed:
Ron Ron Cheng. M.D.
[E-mail: cheng.ronron@gmail.com]

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BACKGROUND: **Granular cell tumor (GCT) is a relatively uncommon predominantly benign lesion that usually presents as a solitary, painless cutaneous or submucosal nodule. Most of these tumors are found in the tongue. Although GCT is believed to have a Schwann cell origin, reports of GCT in peripheral and spinal nerves are uncommon.**

CASE DESCRIPTION: **We report the case of a 43-year-old man with neck pain and hand numbness who was found to have a heterogeneously enhancing left-sided C2 nerve sheath tumor on magnetic resonance imaging. He underwent C2 decompression and resection of the left-sided C2 nerve sheath tumor with subsequent C1-C2 arthrodesis and instrumentation. Histopathologic review showed GCT. Review of the literature yielded 4 other reported cases of GCT within the vicinity of a spinal nerve root. Only one of these explicitly showed spinal nerve root involvement. This is a rare case of a GCT presenting as cervical nerve root mass, and what we believe is the first reported case of this in the literature.**

CONCLUSIONS: **The surgeon should be aware of GCT when encountering spinal nerve root tumors because it may alter the surgical approach necessary for adequate resection compared with more commonly encountered nerve sheath tumors.**

INTRODUCTION

Granular cell tumor (GCT) is a relatively uncommon predominantly benign lesion that usually presents as a solitary, painless cutaneous or submucosal nodule. GCT commonly occurs in the tongue, skin, larynx, breast, bronchi, and gastrointestinal submucosa and comprises polygonal cells with eosinophilic, granular cytoplasm.[1] Although most GCTs are benign, approximately 2% of these tumors are malignant. Malignant GCTs may show necrosis, atypia, infiltrative growth, and increased mitotic activity.[1] Both benign and malignant forms of GCT may show local recurrence,[2] whereas malignant forms may show metastases.[3]

Although GCT is considered by most to have a Schwann cell origin, reports of GCT in peripheral and spinal nerves are uncommon.[4] In this article, a very rare case of GCT presenting as a C2 dorsal root nerve sheath tumor is reported.

CASE REPORT

History and Physical Examination

A 43-year-old man was referred for neurosurgical evaluation because of a 4- to 5-month history of neck pain and a few weeks of bilateral hand numbness. His pain was primarily located at the base of his neck on the right side, as well as in his shoulder. He had also noted paresthesias in the medial palm and fourth and fifth fingers bilaterally. Symptoms arose primarily at night and while driving. He believed that his hands had been clumsy, but he denied any weakness. He denied radicular symptoms and reported no bowel or bladder dysfunction.

On physical examination, the patient had full strength in all muscle groups. He showed very brisk deep tendon reflexes throughout. In addition, he showed mild pathologic spread of reflexes into the hand flexor muscle groups when testing the biceps and brachioradialis tendons. This additional hand muscle response was suggestive of upper motor neuron dysfunction.

Diagnostic Testing

Nerve conduction studies showed prolonged distal peak latency in the right median sensory nerve. Otherwise, all remaining nerves were within normal limits. Electromyography was normal. Magnetic resonance imaging (MRI) of the cervical spine with and without contrast showed a T2 mildly heterogeneously hyperintense, T1 isointense, and T1 heterogeneously enhancing lesion with both extradural and intradural components within the left-sided C1-C2 foramen, consistent with nerve sheath tumor (Figure 1). The tumor was causing mass effect on the spinal cord without any intrinsic signal change.

Intervention, Operative Findings, and Pathologic Diagnosis

Because of a mass effect on the spinal cord and signs of early myelopathy, we

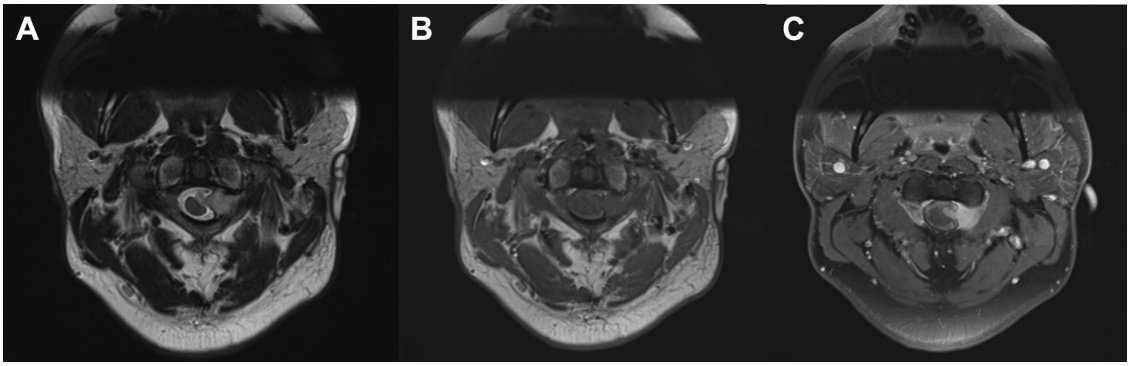

Figure 1. Preoperative axial magnetic resonance imaging of the cervical spine showing the left-sided C2 nerve root tumor with both extradural and intradural components as well as mass effect on the spinal cord. The tumor can be characterized as (**A**) T2 heterogeneously hyperintense, (**B**) T1 isointense before contrast, and (**C**) T1 heterogeneously enhancing after contrast.

discussed resection of the tumor for relief of the mass effect and tissue diagnosis. The patient was taken to the operating room, where he was positioned prone for a posterior cervical approach to the tumor. Neuromonitoring with somatosensory evoked potentials was used throughout the case. After making a midline incision and subsequently performing subperiosteal dissection of the upper cervical spine, we performed laminectomies at the C1 and C2 levels, slightly biased towards the left side according to tumor laterality. Once the initial decompression was complete, we noted a tan mass external to the dura with a component that appeared to extend into the dura. The dura was opened in a T-shaped fashion around the tumor, and the dural edges were retracted with sutures.

On visual inspection, the tumor involved the dorsal C2 nerve root (Figure 2). There did not appear to be tumor adhering to the dura. As work continued under the operating microscope, a plane was created around the mass with a combination of electrocautery and microdissector instruments. The left-sided C1-C2 articular joint was entered to achieve satisfactory circumferential exposure of the tumor. The tumor itself did not appear to be especially vascular. Also, there was no involvement of the left-sided vertebral artery. We were not able to separate tumor from the left-sided C2 nerve root. Because this nerve root is purely sensory in function and commonly sacrificed to facilitate placement of C1 instrumentation, we decided to define the medial and lateral border of the tumor and excise the tumor en bloc along with the C2 nerve root and its ganglion.

Despite the expected diagnosis of schwannoma, intraoperative pathology consultation showed a proliferation of round to polygonal cells with abundant, eosinophilic granular cytoplasm and small, hyperchromatic, eccentrically placed nuclei with interspersed ganglion cells (Figure 3). These findings were consistent with a diagnosis of GCT, which was confirmed by postoperative analysis of routine tissue samples.

Because of the generous bony decompression and violation of the C1-C2 joint on the side of the tumor, we were concerned about instability postoperatively. Therefore, we performed left-sided C1-C2 interarticular arthrodesis with placement of an interarticular allograft followed by C1 and C2 instrumentation. The large dural defect was repaired secondarily with muscle graft, collagen-matrix dural substitute onlay, gel foam, and fibrin glue. Along with subfascial and epifascial surgical drains, a lumbar drain was placed postoperatively for spinal fluid diversion. No neuromonitoring changes were noted during the procedure.

Postoperative Course

The patient remained in the surgical intensive care unit while his lumbar drain was in place. Postoperative radiography showed good alignment and positioning of spinal instrumentation. His surgical drains were removed on postoperative day 5 once the output had tapered. The lumbar drain was removed on postoperative day 7. The patient remained neurologically stable with full strength during his entire hospitalization. He reported new C2 distribution numbness on the left side after surgery. He was discharged home in stable condition.

DISCUSSION

Review of the literature yielded only 4 other cases of a GCT presenting in the vicinity of a spinal nerve root.[2,3,5,6] Vigier et al.[2] reported a 41-year-old man who discovered a painless lump on the right side of his neck. Imaging showed a mass located medial to the right sternocleidomastoid muscle and posterior to the internal jugular vein. The mass showed hyperintensity to adjacent tissues on T2-weighted MRI but no contrast enhancement. Surgical exploration showed a tumor involved with a segment of the cervical plexus, and this was confirmed to be a benign GCT.[2]

Nakamura et al.[3] described a 23-year-old man who presented with 6 months of right-sided back tenderness and a palpable 1-cm mass. In this case, the mass was imaged with ultrasonography and found to reside beneath the superficial fascial layer. On exploration, the mass was found to involve a nerve suspected to be a cutaneous branch of the T8 dorsal root. However, visualization of the nerve root itself was not reported. The investigators were unable to separate the tumor from