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CASE REPORT

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Incidental craniovertebral junction schwannoma: Surgical or radiosurgical management?

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Key Clinical Message

Craniovertebral junction (CVJ) schwannomas are rare tumors, showing direct involvement of the atlanto-occipital and atlanto-axial joints. Microsurgical removal is the standard of care to improve symptoms and local control, but stereotactic radiosurgery (SRS) is an option. Both, surgery, and SRS, may show risks of severe complications. A 41-year-old male was referred to our department after incidental finding of a right-sided C1 tumor. A CT angiogram with 3D reconstructions showed the close relationship between the tumor and the right vertebral artery (VA). A post-contrast enhancement MRI revealed the presence of an extradural mass, sited at the level of the CVJ, mainly at the level of the right articular mass of C1. After multidisciplinary assessment, involving the gamma-knife and neurosurgical teams, we performed a microsurgical resection of the tumor. Histology confirmed the diagnosis of schwannoma. At 1 year follow-up the patient is stable, with no recurrence of the tumor. CVJ schwannoma's current standard of care is surgical resection, but longitudinal studies are required, and should promoted promptly since the recent introduction of the new version of GKSRS that allow the treatment of CVJ's lesions.

KEYWORDS

craniovertebral junction, gamma-knife, radiosurgery, schwannoma

1 | INTRODUCTION

Craniovertebral junction (CVJ) schwannomas are rare tumors located at the jugular foramen, hypoglossal canal, or C1-C2 foramina, showing direct involvement of the atlanto-occipital and atlanto-axial joints.[1,2] Symptoms are often misleading, and include neuropathies and neurological dysfunction, while radiological investigation typically document contrast-enhancing dumbbell-shaped lesions.[3] Microsurgical removal is the standard of care to improve symptoms and local control, but stereotactic radiosurgery (SRS) may be considered in patients not eligible to undergo surgery or that refuse it and for treating postsurgical tumor remnants or recurrences.[5] Both, surgery, and SRS, may show risks of severe complications. We present a case illustration on this debated topic.

2 | CASE PRESENTATION

A 41-year-old male was referred to our department after incidental finding of a right sided C1 tumor, during a head MRI without contrast performed for tinnitus to the right ear. A previous ENT evaluation was negative for any cause of tinnitus. The preoperative planning included CT angiogram with 3D reconstructions, that allowed to highlight the close relationship between the tumor and the right vertebral artery (VA). The right V3 segment run dorsally, medially, and inferiorly to the tumor. The tumor caused osteolytic alterations of the right posterior arc and articular mass of C1 and presented intertumoral calcifications (Figure 1).

A post-contrast enhancement MRI revealed the presence of an extradural mass, sited at the level of the CVJ, mainly at the level of the right articular mass of C1. The tumor caused bony erosion at the level of the right foramen of the VA, of the posterior arc of C1 and is directed toward the antero-lateral paraspinal space, exerting mass effect at the level of the extracranial tract of the internal jugular vein. The medial part of the neoplasm embosses the dural sac. The tumor presents 35×36 mm axial extension, and 30 mm cranio-caudal maximum diameter. The neoplasm shows hypointense appearance in T1, while is hyperintense in T2-weighted sequences (SPAIR/STIR). After administration of gadolinium, the tumor shows diffuse post-contrast enhancement. The paraspinal muscles of the right side showed slight atrophy, compared to those of the opposite side. Lymph nodes showed slight enlargement bilaterally, the jugular-digastric ones. The radiological findings suggested the presence of a schwannoma (Figure 2).

After multidisciplinary assessment, involving the gamma-knife and neurosurgical teams, we performed a microsurgical resection of the tumor. A midline skin incision from inion to C5 was performed, and a right monolateral skeletonization was accomplished, exposing the occipital bone, C1 arc, C2 and C3 laminae. Under microscopic magnification, the tumor was exposed, and dissected from the VA aided by the intraoperative micro-Doppler, that clearly detected the VA. The neoplasm was then incised, fragments were sent for histological examination, and debulked with ultrasound aspirator. Complete tumor removal was achieved with microsurgical technique, sparing the VA, and with minimal muscle damage. Venous bleeding of the VA complex was easily controlled with topic hemostatic agents.[6] The intraoperative neuromonitoring was stable, during the procedure. Immunohistochemical assessment documented S100 positivity, suggestive for the diagnosis of schwannoma and concordant with the morphological histological examination (Figure 3).

Postoperative MRI showed complete tumor removal (Figure 4). At 1 year follow-up the patient is doing well and complains unmodified slight intermittent tinnitus to the right.

3 | DISCUSSION

Craniovertebral junction schwannomas are a relatively rare entity and their management include surgery and more recently radiosurgery.

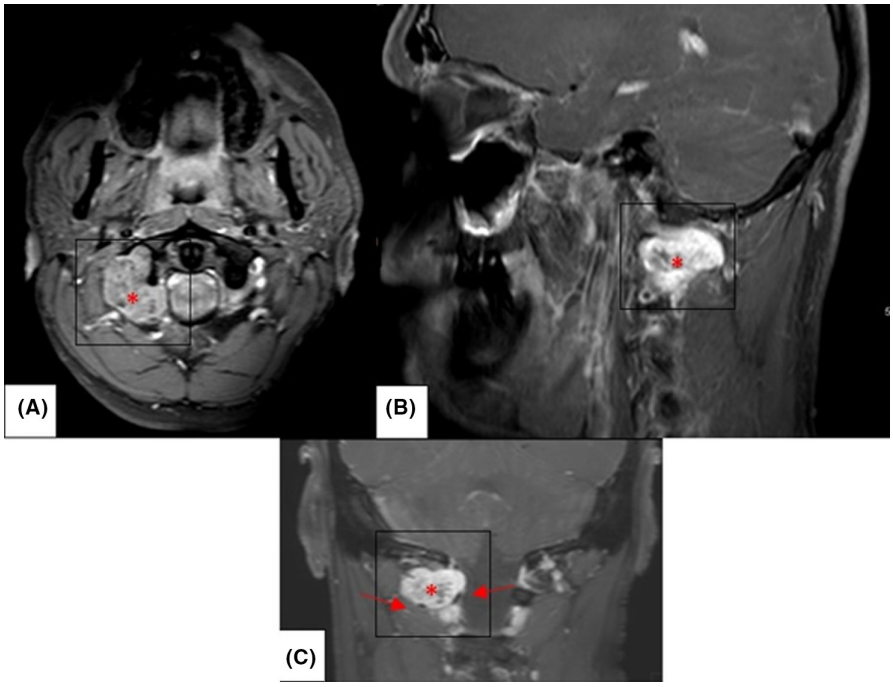

FIGURE 1 T1-weighted MRI with Gadolinum, documenting the right C1 schwannoma (red asterisk) and its close relation with VA (red arrow), in axial (A), sagittal (B) and coronal views (C).

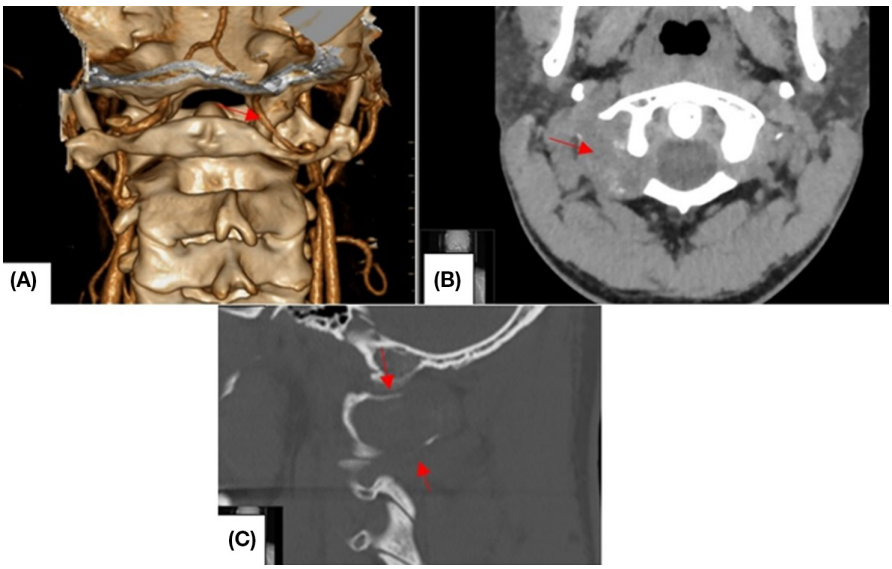

FIGURE 2 (A) CT angiogram with 3D reconstructions showing the right V3 segment running dorsally, medially, and inferiorly to the tumor (red arrow); (B) axial view of CT angiogram showing intertumoral calcifications (red arrow); (C) sagittal view of CT scan with bone algorithm showing bony erosion of the C1 lateral mass and posterior arc (red arrows).

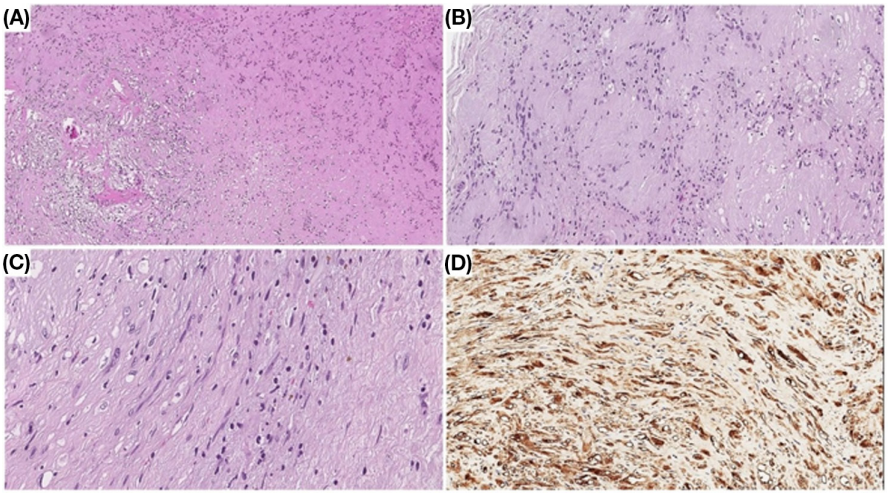

FIGURE 3 Hematoxylin and eosin (H&E) stained sections demonstrating the main morphological and immunohistochemical findings of the present case. (A) Low power magnification (5×) illustrating a spindle cell neoplasm composed of an admixture of hypocellular and hypercellular areas. (B) Nuclear palisading of neoplastic cells around fibrillary process leading to formation of Verocay bodies is depicted (10×). (C) Neoplastic cells showed bland morphology with elongated and wavy nuclei with ill-defined cytoplasm. (D) Diffuse immunohistochemical expression of S100 protein by neoplastic cells, consistent with sChwannian differentiation, is depicted.

Both techniques show risks of complications: Surgical removal takes risk of immediate injury to neural and/or vascular structures, that can be limited by careful preoperative planning and meticulous intraoperative dissection[7]; SRS treatment may cause late vessel occlusion, especially for those tumors in tight contact with eloquent arteries of this area, namely the vertebral artery.[4,8–11] Our decision to perform a surgical resection was driven by the young age of the patient and the close relation with the VA segment. The related risks of late occlusion of VA, in a patient in good clinical condition were the reasons for our surgical indication. Of notice, we discussed about the two options, surgery, and SRS, with the patient and his family members, to allow him to have all the necessary information to decide if endorse our main suggestion or to choose a second therapeutic option. However, literature lacks exhaustive information on this topic, due to the rarity of this tumor. More data on long follow-up of patients treated with gamma-knife radiosurgery (GKSRS) should be collected and based on the risk of late artery occlusion obtained, noninvasive techniques for the treatment of tumors located in eloquent anatomical area could be considered to become a new first line of care.

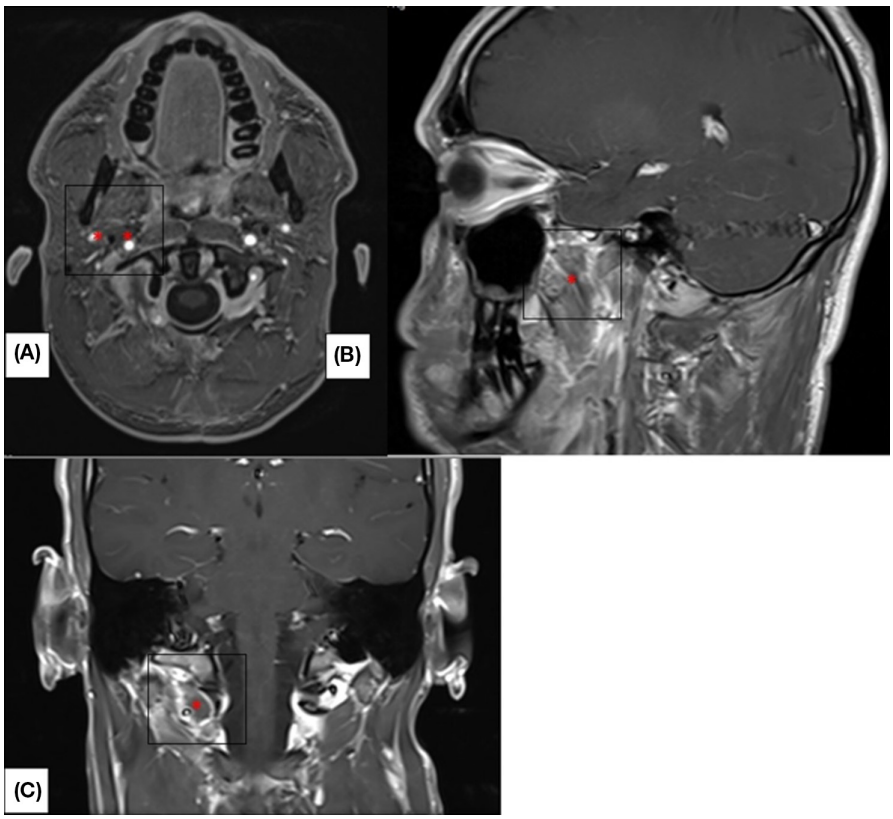

FIGURE 4 (A) postoperative T1 weighted MRI with Gadolinum documenting the complete removal of the tumor and the residual C2 nerve, split in two halves (red asterisk for each half of the nerve). (B, C) sagittal and coronal postoperative T1-weighted MRI scans with Gadolinum documenting the radical resection of the tumor and its surgical bed (red asterisk).

4 | CONCLUSIONS

CVJ schwannomas are rare tumors, sited in an eloquent anatomical area. The current standard of care is surgical resection, but if future studies will document GKSRS similar efficacy and safety, patients could benefit of a noninvasive treatment, changing the actual standard of care, even in the young-adult population. Longitudinal studies are required and should promoted promptly since the recent introduction of the new version of GKSRS that allow the treatment of CVJ's lesions.

AUTHOR CONTRIBUTIONS

Giuseppe Emmanuele Umana: Conceptualization; data curation; formal analysis; investigation; supervision; writing – original draft; writing – review and editing. **Maurizio Passanisi:** Supervision; visualization. **Bipin Chaurasia:** Validation. **Gianluca Scalia:** Data curation; investigation; visualization; writing – original draft; writing – review and editing.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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