Original Article

Dorsal approaches to intradural extramedullary tumors of the craniovertebral junction

Daniel Refai, John H. Shin, Christopher Iannotti, Edward C. Benzel

Center for Spine Health, Department of Neurological Surgery, Cleveland Clinic, Cleveland, OH - 441 95, USA

Corresponding author: Dr. Edward C. Benzel, Cleveland Clinic, Neurological Institute, 9500 Euclid Avenue, S40, Cleveland, OH – 441 95, USA. E-mail: benzele@ccf.org

Abstract

Tumors of the craniovertebral junction (CVJ) pose significant challenges to cranial and spine surgeons. Familiarity with the complex anatomy and avoidance of injury to neurologic and vascular structures are essential to success. Multiple surgical approaches to address lesions at the CVJ have been promoted, including ventral and dorsal-based trajectories. However, optimal selection of the surgical vector to manage the pathology requires a firm understanding of the limitations and advantages of each approach. The selection of the best surgical trajectory must include several factors, such as obtaining the optimal exposure of the region of interest, avoiding injury to critical neurologic or vascular structures, identification of normal anatomical landmarks, the familiarity and comfort level of the surgeon to the approach, and the need for fixation. This review article focuses on dorsal approaches to the CVJ and the advantages and limitations in managing intradural extramedullary tumors.

Key words: Craniovertebral junction, dorsal, dorsolateral, surgical approach, tumor

INTRODUCTION

Anatomy

The craniovertebral junction (CVJ) extends rostrally from the foramen magnum and caudally to the atlantoaxial vertebrae. Anatomically, the CVJ encompasses the medulla, the cervicomedullary junction, and the upper cervical spinal cord. Spine surgeons must be familiar with this anatomy as well as the various surgical approaches to optimally address lesions affecting the CVJ. Such lesions include congenital and acquired bony defects, tumors, inflammatory conditions, and those arising from trauma. This review article focuses on dorsal surgical approaches to the CVJ for the management of intradural extramedullary (IDEM) tumors.

Surgical approach alternatives

Classically, several different approaches to the CVJ have been described.[1] These are based on either ventral or dorsal trajectories. Ventral approaches include variations of the transoral and transcervical routes. Dorsal approaches include the dorsal midline, dorsolateral, and far/extreme lateral routes. The approaches reviewed herein encompass the dorsal approaches to the CVJ.

The selection of the optimal surgical approach must include several factors, such as obtaining optimal exposure of the region of interest, avoiding injury to key neurologic or vascular structures, the identification of normal anatomical landmarks, the familiarity and comfort of the surgeon to the approach, and the need for fixation. The dorsal approaches to the CVJ offer the surgeon the ability to manage IDEM tumors while achieving the above goals more readily as compared to the ventral-based approaches.

The dorsal approach

Dorsal approaches for IDEM lesions have even been described for ventrally located arachnoid cysts,[2] nerve sheath tumors,[3] meningiomas,[4] and schwannomas.[5] The standard dorsal approach involves a midline incision from the inion to the mid-cervical spine. For a dorsolateral approach, the midline incision can be enlarged laterally to expose the transverse foramen of the

atlas, thereby exposing the vertebral artery between the atlas and axis. A cervical laminectomy of the atlas and axis is usually performed with an additional subaxial laminectomy, as needed to achieve optimal surgical exposure. A suboccipital craniotomy/craniectomy can be included, as deemed necessary. The corridor for the surgical trajectory provided by these approaches enables the surgeon access to posterior, posterolateral, and anterior IDEM tumors. Furthermore, these approaches provide for maximal intradural exposure as compared to the other surgical vectors. This is critical as it achieves maximal visualization of the pathology, while minimizing postoperative complications such as infection and cerebrospinal fluid leak. These factors make these approaches far superior to ventral-based techniques [Figures 1 and 2].

Occipito-cervical fusion

The decision to perform an occipito-cervical fusion is critical when working at the CVJ. Factors such as the amount of bone resection and the stability of the CVJ must be ascertained. Shin *et al.*[5] reported their experience of occipital-cervical fusion after resection of CVJ tumors in both the adult and pediatric populations. They performed the fusion either on the day or surgery or commonly within a 24-h period following tumor resection. The decision to fuse patients depended on several

factors including evidence of rotatory subluxation of C1 on C2, resection of greater than 70% of a unilateral occipital condyle, greater than 50% resection of the bilateral occipital condyles, extensive destruction of the C1 and C2 vertebral bodies, or the removal of the posterior elements of C1 and C2 including the facet joints.

Representative case

A 74-year-old right-handed male with no significant past medical history presented with new-onset left leg weakness and neck stiffness, and had two falls over the week prior to admission, without loss of consciousness. He was seen in an outside hospital emergency department where a head computed tomography (CT) scan identified a C1 lesion; cervical spine magnetic resonance imaging (MRI) revealed a 2.0 cm × 1.2 cm × 1.5 cm enhancing well-circumscribed intradural-extramedullary lesion with left-sided foraminal extension (dumbbell tumor) at the level of C1-2 with severe effacement of the spinal cord at this level; no flow voids were seen within the lesion [Figure 3].

Preoperative neurological examination revealed intact cranial nerves; motor strength was 5/5 in the upper extremities bilaterally. Hoffman's sign was present on the left side. Lower extremity motor examination revealed mild proximal left-sided
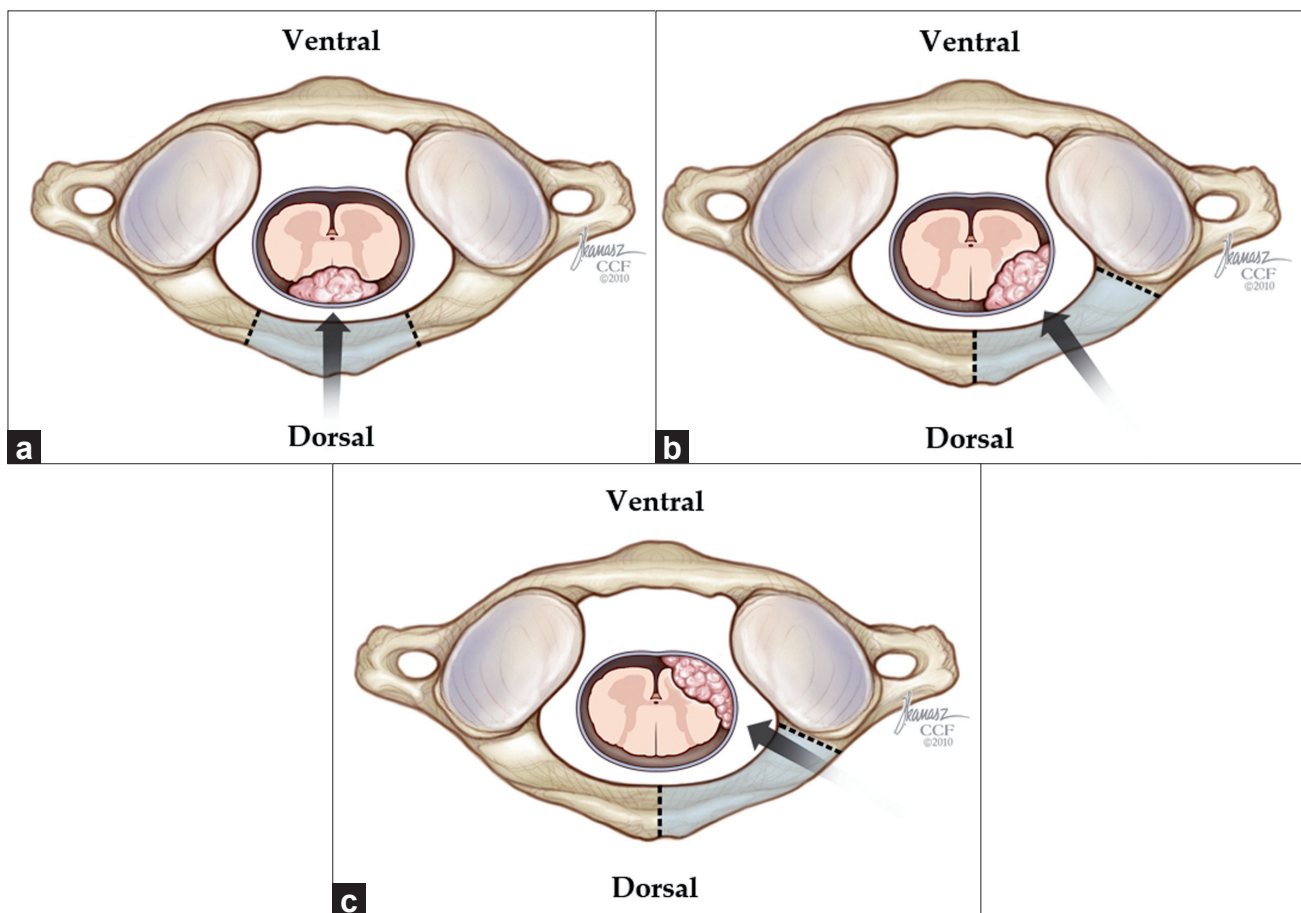

Figure 1: Dorsal approaches to intradural extramedullary tumors of the craniovertebral junction. The dorsal midline and dorsolateral approaches are illustrated. The extent of bone removal is delimited by dotted lines. Surgical trajectory is shown here with arrows directed toward the pathology. After adequate bone removal, tumors in this region can be removed successfully as long as they are visualized intradurally.
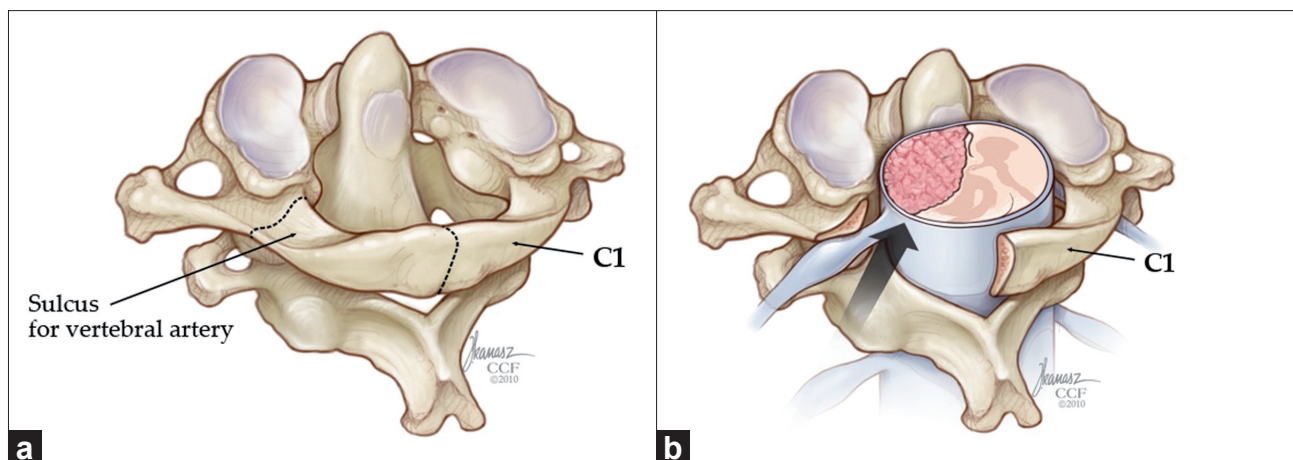
Figure 2: Dorsolateral approach. Illustration demonstrating the unique anatomy of the C1-2 joint before (a) and after (b) C1 laminectomy. After laminectomy and resection of the sulcus arteriosus, a natural corridor to the ventral canal is obtained. The trajectory is shown with an arrow.

weakness, with iliopsoas strength 4/5. Full motor strength was present upon knee flexion, knee extension, as well as dorsiflexion and plantar flexion of the feet bilaterally. Positive Babinski sign was noted bilaterally. Sensation was intact to light touch, pain/temperature, and joint position sense; hyperreflexia was present in the left upper extremity; deep tendon reflexes were symmetric throughout the lower extremities.

Surgical approach

A midline skin incision was carried from the inion to the rostral aspect of C3. The paraspinal muscles were dissected subperiosteally and a laminectomy of C1 and C2 was performed using a high-speed drill. The dura mater was opened in a linear fashion approximately 5 mm to the left of midline from the foramen magnum to C2. A pale-yellowish IDEM tumor was seen attached to the dentate ligament on the left. Although a midline dorsal exposure was utilized, a posterolateral trajectory to the tumor was obtained without difficulty. Using the microscope, the dissection of the tumor was carried out using microsurgical technique. The dentate ligaments were transected with microscissors and the traversing nerve roots were carefully dissected from the surface of the tumor. Several small nerve roots were observed penetrating the tumor; these were sacrificed. The extradural portion of the tumor within the C1-C2 foramen was removed en bloc. Intraoperative monitoring of motor-evoked and somatosensory-evoked potentials (MEPs, SSEPs) was performed, without attenuation or loss of either waveform during the dissection. The tumor was sent for frozen and permanent pathology, which yielded a diagnosis of schwannoma. An instrumented fusion was deemed unnecessary, as the C1-2 facets and capsules remained intact. A watertight dural closure was achieved using a running 5-0 prolene suture, followed by the placement of DuraGen onlay and fibrin glue. The patient awoke at neurological baseline. Postoperative MRI revealed gross total resection of the IDEM tumor [Figure 4]. At one year postoperatively, mild proximal left lower extremity weakness was present as baseline. However, the patient was ambulating independently.

DISCUSSION

Meningiomas, neurofibromas, and schwannomas are the most common IDEM tumors of the CVJ.[6] There are several surgical limitations and considerations that must be addressed when tailoring an appropriate approach to these tumors. The optimal approach provides maximal exposure with the least manipulation of neural elements. As a general rule, lesions posterior to the spinal cord can be reached using a dorsal midline approach while anterior and lateral lesions can be accessed with a dorsolateral approach. However, most IDEM tumors of this region can be reached and successfully resected from a dorsal midline approach even if situated lateral and anterior to the spinal cord. Once intradural exposure is made and the tumor is visualized, microsurgical dissection allows for gradual resection of the pathology without retraction of the nearby neural elements. This trajectory takes advantage of the natural retraction provided by the growing pathology and allows for direct access for successful resection. The main limitation to this vector is access to tumors that are strictly anterior to the spinal cord and hidden from view once dural opening is made. In these cases, a dorsolateral approach provides an unencumbered view of the anterior CVJ. The most appropriate trajectory to the underlying pathology must be considered when planning a surgical approach, with careful consideration to neighboring vascular and neural structures. Thus, the appropriate surgical approach must be based on an understanding of the underlying anatomy, the type and location of the lesion, and the stability of the CVJ.[7]

Emphasis on trajectory: Dorsal midline and dorsolateral approaches

A key component to the selection of the appropriate surgical vector to the CVJ is the ability to adequately visualize the underlying pathology. The dorsal and dorsolateral approaches
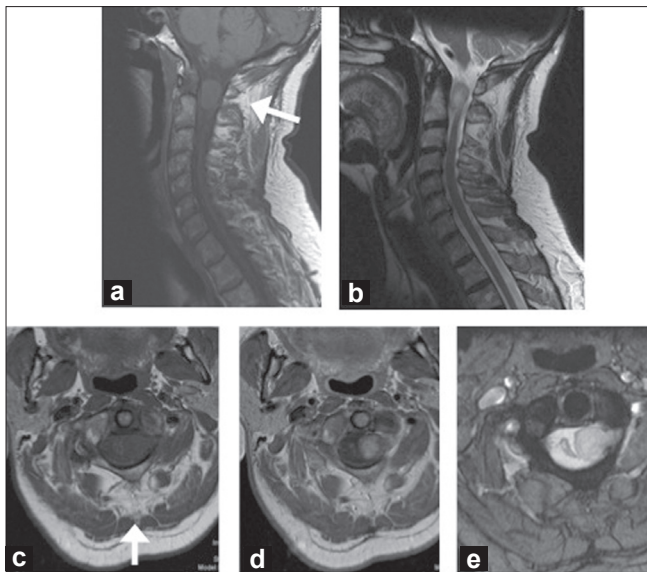
Figure 3: Preoperative sagittal T1-weighted (a) and T2-weighted (b) and axial T1-weighted (c), T1-weighted with contrast (d), and T2-weighted with contrast (e) MR images demonstrating a homogenously enhancing IDEM tumor at the C1 spinal cord level (d), predominantly on the left side. The surgical trajectory using a midline dorsal exposure to the tumor is shown (arrow).
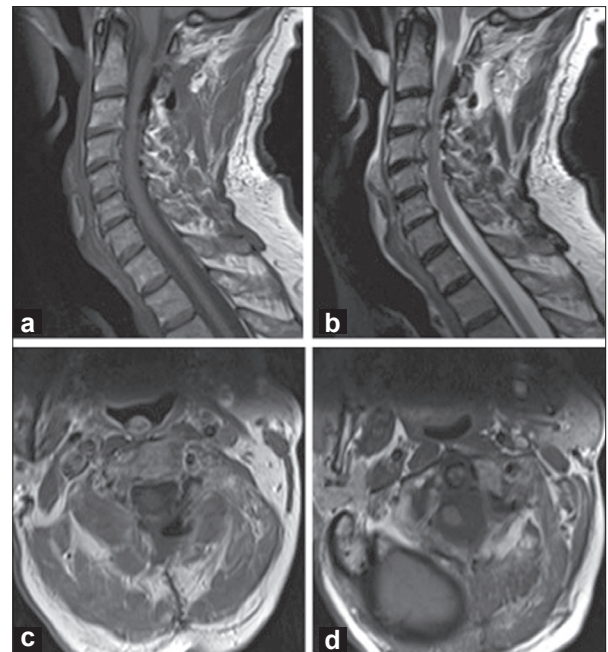

Figure 4: Postoperative T1- and T2-weighted sagittal (a, b) and T1-weighted (c) and T1-weighted with contrast (d) axial MR images. Note the absence of contrast enhancement (d), indicating a gross total resection of the tumor.

to the CVJ enable the surgeon a wide trajectory to resect the pathology without significant bone removal or neural retraction. By removing the C1 and C2 lamina, surgeons may easily access a directly posterior or posterolateral lesion as long as it is visible. If the lesion is more anterior in nature, a dorsolateral approach provides the surgeon better visualization because of the unique anatomy of the C1-2 joint, specifically the relationship of the exiting C2 nerve root to the joint. This unique feature differentiates the C1-2 joint from the subaxial spine. Whereas in the subaxial spine the lateral masses are posterior to the exiting nerve root, at C1-2, the facet and pars interarticularis are anterior to the exiting C2 nerve root. With ligation of the C2 nerve root, direct access anterior to the spinal cord is possible. Thus a much wider dorsolateral trajectory is possible to resect an anterior lesion without the need for additional bone removal. If needed, the medial C2 lateral mass can be partially drilled while preserving the pars interarticularis to allow for an even wider surgical corridor. Even if more than half of the C2 lateral mass is removed, the dorsolateral approach permits placement of stabilizing instrumentation through the same exposure. These critical regional anatomic differences of the C1-2 facet complex make the dorsal and dorsolateral approach to the CVJ the best route for resection of IDEM lesions. In addition, limitations inherent to ventral-based approaches such as a higher rate of cerebrospinal fluid (CSF) leaks and a smaller operative field of exposure are avoided.

Dorsal midline approach

The dorsal midline approach to the CVJ for the treatment of IDEM tumors is well described and utilizes a suboccipital craniotomy and cervical laminectomy.[7-11] This approach is familiar with most neurosurgeons. The exposure provided allows

for direct access to the posterior foramen magnum and to 90 degrees of both sides of the midline. If needed, the lateral most aspects of the posterior arch of the atlas or axis beyond the facets to the vertebral arteries may be exposed. The vertebral arteries may be mobilized as needed if bone removal lateral to the sulcus arteriosus is required for adequate exposure. Most IDEM tumors of the CVJ may be resected by this approach. Paramount to the successful use of this approach is an understanding of the intradural exposure provided. From the dorsal midline approach, tumors that are visible are resectable. Given the nature of most IDEM tumors in this location, these tumors are often resected piecemeal with careful microsurgical technique. With time, persistence, and microsurgical dissection, these tumors often deliver themselves into the operative field with minimal retraction of neural elements. Tumors strictly posterior to the spinal cord are easily accessed through this approach as they are evident as soon as the dura is opened. Tumors that are lateral and anterior to the spinal cord may also be removed successfully as long as part of the tumor can be seen once the dura is opened. Lateral lesions that extend anteriorly can be followed to the anterior most aspect of the tumor. This is possible due to the fact that most IDEM tumors at the CVJ are slow-growing tumors that over time have gradually shifted native neural elements away from it. Surgical resection is thus aided by this retraction and helps facilitate removal.

Should more than half of the atlantoaxial joints be removed, the facile application of instrumentation through the same exposure is another advantage of this approach. For most IDEM tumors involved in this region, aggressive bony resection is seldom needed as gradual, expansile growth of the underlying tumor

often displaces the neural elements to the contralateral side, in effect retracting the neural elements and providing a larger intradural exposure than would normally be encountered. Reported complications for this approach stem mainly from the dural closure and risk of CSF leak.[10] Brainstem dysfunction and lower cranial nerve palsies have been reported with resection of tumors in this area, however, such complications are due to the underlying pathology and microsurgical dissection from the nearby structures and not necessarily due to the operative exposure.

A major limitation to this approach is that it is not as effective a route for purely ventral lesions. Lesions that are strictly ventral are limited by the brainstem or cervical cord. Particularly if the spinal cord drapes over an anterior lesion, the tumor is hidden from view from a dorsal midline approach and thus inaccessible. Only lesions directly dorsal are easily accessed with little brainstem manipulation.[10] Interestingly, small series have reported success with resection of ventral tumors from a dorsal midline suboccipital approach, though these studies fail to describe their morbidity.[9]

Far lateral/extreme approach

In addition to the dorsal midline and dorsolateral approaches, other techniques often used to reach the CVJ include the far lateral transcondylar and the extreme lateral approaches. These approaches have been used for many years to access the lateral and anterior CVJ.[12-17] Variations of these approaches have been described with the distinction among them being the extent of bone removal at the skull base.[18] These variants include the transfacetal, retrocondylar, partial transcondylar, transcondylar, transtubercular, and transjugular approaches.[19] Significant controversy exists regarding the most appropriate approach to ventral lesions among these techniques.[20-23] Management of the occipital condyle, vertebral artery, and need for occipitocervical fusion are still debated.[20]

The far lateral transcondylar approach was developed to provide access to the pontomedullary junction, the lower third of the clivus, the ventrolateral foramen magnum, and the upper cervical spinal cord without retraction of the cerebellum or the spinal cord.[12,14,15] With this approach, the skin incision extends from the midline to the inion, then across to the mastoid tip. The posterior arch of C1 including the transverse process and lamina of C2 are exposed as needed. Bony removal includes the arch of C1 from the midline to the sulcus arteriosus of the vertebral artery. Dissection of the vertebral artery is determined by the required exposure and removal of the dorsal aspect of the foramen transversarium is often necessary. A suboccipital craniotomy is performed, including up to two-thirds of the occipital condyle. Venous bleeding may be encountered if the posterior condylar emissary vein is violated as it passes toward the jugular bulb. The resection of bone into the condylar fossa and into the medial aspect of the posterior condyle is vital in obtaining the ideal trajectory to the front of the brainstem and spinal cord without retraction.[24] Laterally based compressive lesions can be resected with minimal brain retraction as most

tumors have forced neural elements to the contralateral side. This provides direct access to the lower third of the clivus, extracranial and intracranial vertebral artery, and exposure of the lower cranial nerves.[25]

Disadvantages include venous sinus injury, hypoglossal injury, lower cranial nerve injury, and potential destabilization. Potential complications are directly related to the angle and vector of the approach. Adequacy of intradural exposure and dural closure are also of concern. Complications reported with this technique include brainstem edema, air embolism, lower cranial nerve palsies, hydrocephalus, CSF leak, and death.[23,24] In particular, the hypoglossal nerve is subject to increased potential injury as it lies in the anterior one-third and superior margin of the occipital condyle. Most of the reported complications involve patients with anterior and lateral tumors requiring brain retraction and cranial nerve dissection. In some series, ninth and tenth cranial nerve deficits were the most common complications.[15] Should instrumentation need to be applied, it can be done at the same setting.

Similar to the transcondylar approach, the extreme lateral approach was developed to provide extended access to the ventral foramen magnum, ventral brainstem, and craniocervical junction.[18] This approach has been described in the treatment of vertebrobasilar dolichoectasia, posterior circulation aneurysms, meningiomas, schwannomas, dermoids, epidermoids, paragangliomas, and arachnoid cysts.[18,19,23] Whereas anterior approaches, such as the transoral or transcervical approach provide the most direct access to the ventral aspect of the foramen magnum and craniocervical junction, the extreme lateral approach obviates the potential complications and disadvantages of the former, including limited lateral exposure and increased incidence of CSF leak, infection, and fistulae.

As with the far lateral transcondylar approach, the posterior third of the occipital condyle is removed until the jugular bulb is encountered. The hypoglossal canal, located at the junction of the posterior and middle thirds of the condyle, is then skeletonized by removing bone superiorly toward the jugular bulb. The jugular tubercle is medial and inferior to the adjacent jugular bulb. The jugular tubercle is then drilled and the remaining bone dissected from the dura. This drilling of the jugular tubercle maximizes the intradural exposure of the ventral aspect of the brainstem.[18] Advantages of this technique include direct visualization of tumor/cord interface and the entire ventral aspect of the thecal sac from the nerve roots on one side to the other. The vertebral artery and the vascular supply to the tumor can be controlled as well.[18] Structures at greatest risk during this exposure are cranial nerves IX, X, and XI as they travel from the brainstem to the jugular foramen.

Significant complications include CSF leak, lower cranial nerve palsies, meningitis, pseudomeningocele, and vertebral artery injury.[20,23] In Margalit's series, approximately 50% of patients with postoperative cranial nerve palsies improved to normal function by six months.[20] Despite the reported higher rates of cranial nerve injury and CSF leak, when compared to the far lateral transcondylar approach, advocates of this approach

emphasize the excellent exposure of the ventral surface of the brainstem and upper cervical cord.[18] Nonetheless, adequacy of intradural exposure and dural closure remain issues of concern.

CONCLUSION

Intradural extramedullary tumors at the CVJ often present significant challenges to neurosurgeons because of the complex regional anatomy and relation of tumor pathology to neighboring vascular and neural structures. Surgeons must be familiar with the anatomy as well as the various surgical approaches to optimally address lesions affecting the CVJ. The dorsal approaches reviewed here highlight the importance of understanding the limitations of each approach in designing an appropriate trajectory for ultimate success. Most IDEM tumors of the CVJ can be successfully resected with the dorsal midline and dorsolateral approaches with significantly less morbidity than the far/extreme lateral approaches. The latter are associated with increased morbidity and pose greater risk to underlying neural structures, while providing a somewhat limited intradural exposure.

REFERENCES

1. Ryu SI, Kim DH. Surgical approaches for decompression and fixation of the craniovertebral junction. In: Dickman CA, Fehlings MG, Gokaslan ZL, editors. Spinal cord and spine column tumors: Principles and practice. New York: Thieme Medical Publishers, Inc; 2006. p. 420-36.
2. Mohindra S, Gupta SK, Chhabra R, Gupta R, Kumar A, Radotra BD. Ventrally placed craniovertebral junction arachnoid cysts in children: a report of 2 cases and literature review. Surg Neurol 2007;68:85-8.
3. Maurya P, Singh K, Sharma V. C1 and C2 nerve sheath tumors: analysis of 32 cases. Neurol India 2009;57:31-5.
4. Menezes AH. Craniovertebral junction neoplasms in the pediatric population. Childs Nerv Syst 2008;24:1173-86.
5. Shin H, Barrenechea IJ, Lesser J, Sen C, Perin NI. Occipitocervical fusion after resection of craniovertebral junction tumors. J Neurosurg Spine 2006;4:137-44.
6. Wu Z, Hao S, Zhang J, Zhang L, Jia G, Tang J, *et al.* Foramen magnum meningiomas: experiences in 114 patients at a single institute over 15 years. Surg Neurol 2009;72:376-82.
7. Rhoton AL Jr. The foramen magnum. Neurosurgery 2000;47:S155-93.
8. Arnautović KI, Al-Mefty O, Husain M. Ventral foramen magnum meninigiomas. J Neurosurg 2000;92:71-80.
9. Kandenwein JA, Richter HP, Antoniadis G. Foramen magnum meningiomas-experience with the posterior suboccipital approach. Br J Neurosurg 2009;23:33-9.
10. Menezes AH. Complications of surgery at the craniovertebral junction--avoidance and management. Pediatr Neurosurg 1991;17:254-66.
11. Goel A, Desai K, Muzumdar D. Surgery on anterior foramen magnum meningiomas using conventional posterior sub-occipital approach: A report on an experience with 17 cases. Neurosurgery 2001;49:102-7.
12. Heros RC. Lateral suboccipital approach for vertebral and vertebrobasilar artery lesions. J Neurosurg 1986;64:559-62.
13. Dowd GC, Zeiller S, Awasthi D. Far lateral transcondylar approach: dimensional anatomy. Neurosurgery 1999;45:95-9.
14. Lanzino G, Paolini S, Spetzler RF. Far-lateral approach to the craniocervical junction. Neurosurgery 2005;57:367-71.
15. al-Mefty O, Borba LA, Aoki N, Angtuaco E, Pait TG. The transcondylar approach to extradural nonneoplastic lesions of the craniovertebral junction. J Neurosurg 1996;84:1-6.
16. Matsushima T, Matsukado K, Natori Y, Inamura T, Hitotsumatsu T, Fukui M. Surgery on a saccular vertebral artery-posterior inferior cerebellar artery aneurysm via the transcondylar fossa (supracondylar transjugular tubercle) approach or the transcondylar approach: surgical results and indications for using two different lateral skull base approaches. J Neurosurg 2001;95:268-74.
17. Day JD, Fukushima T, Giannotta SL. Cranial base approaches to posterior circulation aneurysms. J Neurosurg 1997;87:544-54.
18. Sen CN, Sekhar LN. An extreme lateral approach to intradural lesions of the cervical spine and foramen magnum. Neurosurgery 1990;27:197-204.
19. Salas E, Sekhar LN, Ziyal IM, Caputy AJ, Wright DC. Variations of the extreme-lateral craniocervical approach: anatomical study and clinical analysis of 69 patients. J Neurosurg 1999;90:206-19.
20. Margalit NS, Lesser JB, Singer M, Sen C. Lateral approach to anterolateral tumors at the foramen magnum: factors determining surgical procedure. Neurosurgery 2005;56:324-36.
21. Bruneau M, George B. Foramen magnum meningiomas: detailed surgical approaches and technical aspects at Lariboisiere Hospital and review of the literature. Neurosurg Rev 2008;31:19-32.
22. Borba LA, de Oliveira JG, Giudicissi-Filho M, Colli BO. Surgical management of foramen magnum meningiomas. Neurosurg Rev 2009;32:49-58.
23. Kumar CV, Satyanarayana S, Rao BR, Palur RS. Extreme lateral approach to ventral and ventrolaterally situated lesions of the lower brainstem and upper cervical cord. Skull Base 2001;11:265-75.
24. Menezes AH. Surgical approaches: postoperative care and complications "posterolateral-far lateral transcondylar approach to the ventral foramen magnum and upper cervical spinal canal". Childs Nerv Syst 2008;24:1203-7.
25. Wen HT, Rhoton AL Jr, Katsuta T, de Oliveira E. Microsurgical anatomy of the transcondylar, supracondylar, and paracondylar extensions of the far-lateral approach. J Neurosurg 1997;87:555-85.