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Surgical outcomes of craniocervial junction meningiomas: A series of 22 consecutive patients

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ABSTRACT

Objective: We present our experience in managing craniocervical junction meningiomas and discuss various surgical approaches and outcomes.
Methods: We retrospectively reviewed 22 consecutive cases of craniocervical junction meningiomas operated on between August 1995 and May 2012.
Results: There were 15 female and 7 male patients (mean age: 54 years). Meningiomas were classified based on origin as spinocranial (7 cases) or craniospinal (15 cases). Additionally, the tumors were divided into anatomical location relative to the brainstem or spinal cord: there were 2 anterior tumors, 7 anterolateral, 12 lateral, and 1 posterolateral. Surgical approaches included the posterior midline suboccipital approach (9 cases), the far lateral approach (12 cases) and the lateral retrosigmoid approach (1 case). Gross-total resection was achieved in 45% of patients and subtotal in 55%. The most common post-operative complications were cranial nerve (CN) IX and X deficits. The mortality rate was 4.5%. There have been no recurrences to date with a mean follow-up was 46.5 months and the mean Karnofsky score at the last follow-up of 82.3. In this series, spinocranial tumors were detected at a smaller size ($p = 0.0724$) and treated earlier ($p = 0.1398$) than craniospinal tumors. They were associated with a higher rate of total resection ($p = 0.0007$), fewer post-operative CN IX or X deficits ($p = 0.0053$), and shorter hospitalizations ($p = 0.08$).
Conclusion: Our experience suggests that posterior midline suboccipital or far-lateral approaches with minimal condylar drilling and vertebral artery mobilization were suitable for most cases in this series.

1. Introduction

Meningiomas are generally benign tumors that generally carry a good prognosis. Craniocervical junction (CCJ) meningiomas account for 1.8–3.2% of all the meningiomas [1]. These lesions are often large at the time of diagnosis [2]. Tumor involvement of neighboring neurovascular structures poses significant technical challenges for surgeons. The tumor often encases the vertebral or basilar artery, and the lower cranial nerves, and may invade the bony structures of the craniocervial junction. Surgical management includes tumor resection and stabilization of the CCJ when necessary. In our report, two main surgical approaches, the posterior midline suboccipital approach and the far lateral approach, were used to facilitate resection and limit post-operative morbidity. We present our experience in the past 17 years and discuss the factors that dictate the surgical approach to adopt, that determine surgical outcome, and that affect the resectability of the tumor.

2. Patients and methods

From August 1995 to May 2012, a total of 22 patients with CCJ meningiomas were treated surgically at our institution. Clinical notes, operative notes, and radiological findings were evaluated. The study was approved by the Johns Hopkins Institutional Review Boards located in Baltimore, MD. For surgical outcomes, the Karnofsky Performance Scale (KPS) score and a scoring system proposed by Samii et al. [3] were employed. Preoperative imagining consisted of magnetic resonance imaging (MRI) or computer tomographic (CT) scans. In some cases, CT angiogram and

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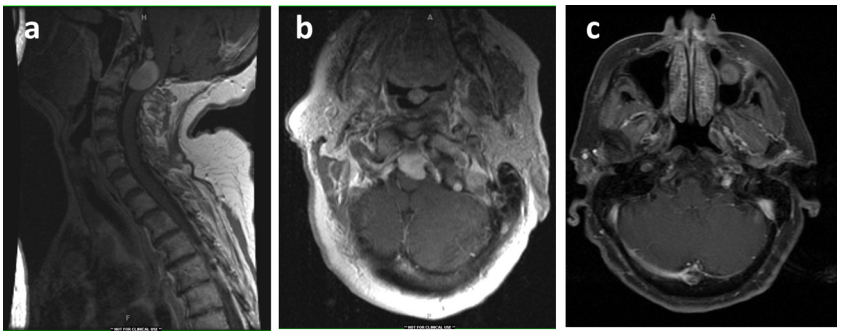

Fig. 1. This 84-year-old female patient experienced a significant deterioration for her neurological condition several months pre-op and presented with neck pain, dysesthesias and was unable to walk properly. MRI scans (a–b) revealed a large spinocranial anterior meningioma (2.3 cm × 1.3 cm × 1.5 cm) displacing neuroaxis posteriorly. She underwent a posterior suboccipital midline approach with C1 and C2 laminectomy. Post-operatively, she had a very good recovery, with headache and dysesthesias resolved and was able to walk normally. MRI (c) at follow-up showed complete excision of the tumor.

magnetic resonance angiogram (MRA) were also used pre-operatively and intra-operatively.

We categorized the meningiomas based on the origin of the tumor matrix or dural insertion. Craniospinal meningiomas originate from the skull base and extend into the spinal canal through the foramen magnum; spinocranial meningiomas, in contrast, originate from the spinal canal and extend rostrally into the cranial cavity. The tumors were further categorized by their relative location to the medulla in the axial plane: anterior (tumor mass predominantly anterior to the neuraxis, displacing it dorsally) (Fig. 1a–c), lateral (Fig. 2a–f), anterolateral (Fig. 3a–f), posterior and posterolateral. The anterior and anterolateral tumors are located ventral to the dentate ligament while the posterior and posterolateral tumors are dorsal to the dentate ligament. The extent of tumor resection was categorized into complete or subtotal. Complete tumor resection was defined as total removal of the tumor mass, including the capsule.

Post-operatively, every patient underwent an MRI scan immediately after surgery to detect edema, hemorrhage, or residual tumor. Typically, another MRI scan was obtained post-operatively at 2 months, 6 months and 1 year in order to determine the resection status of the tumor, pseudomeningocele, and monitor for growth of the residual tumor. Further scans were obtained if new clinical symptoms developed. If the tumor remained stable, patients were instructed to undergo annual MRI scans. Additional follow-up information was obtained by regular outpatient follow-ups and telephone calls. Clinical and radiological characteristics of all 22 patients are summarized in Table 1.

Means plus or minus the standard deviation are presented. For statistical analysis, Student's *t*-tests were used for continuous variables, and the Chi-squared test for categorical variables. A difference was considered significant with a *p*-value less than 0.05.

Table 1
Clinical and radiological characteristics of 22 patients.

Pt. no.	Tumor type	Tumor location	VA/CN involvement	Surgical approach	Condyle drilling	VA transposed	Extent of removal	Complications	Karnofsky score (pre/post-op)
1	CS	Lat, IE	VA	FL	Partial	No	Sub	–	90, 95
2	SC	AL, I	–	FL	Partial	No	Total	–	80, 100
3	SC	Lat, I	–	PM	No	No	Total	–	80, 90
4	SC	Lat, IE	VA, CN	FL	No	No	Total	–	70, 90
5	CS	Lat, I	VA, CN	FL	Partial	No	Sub	Infection, dysphagia	70, 80
6	CS	Lat, I	VA, CN	FL	Total	No	Sub	Dysphagia, vocal cord paralysis, new CN VI, VII, XII deficits	60, 95
7	CS	Lat, I	VA, CN	FL	Partial	No	Sub	Dysphagia, vocal cord paralysis, motor sensory deficit, bladder disturbance, quadriplegia	40, 0
8	SC	Lat, I	CN	FL	No	No	Total	–	70, 85
9	SC	Ant, I	–	PM	No	No	Total	–	70, 100
10	CS	Lat, I	CN	FL	Partial	No	Sub	Dysphagia, new CN XI, XII deficit	65, 80
11	SC	PL, I	–	PM	Total	No	Total	New CN deficits,	80, 85
12	CS	AL, I	–	PM	No	Yes	Total	New muscle weakness	70, 90
13	CS	Al, IE	CN	FL	Partial	No	Sub	Dysphagia,	60, 80
14	CS	AL, I	CN	Lat	Partial	No	Total	Dysphagia, vocal cord paralysis	85, 90
15	SC	Lat, I	VA, CN	PM	No	No	Total	Infection, new CN VI deficit, CSF leak	75, 75
16	CS	AL, I	VA, CN	PM	No	Yes	Sub	Dysphagia, new muscle weakness, balancing difficulty	80, 70
17	CS	Ant, I	CA, CN	FL	Partial	Yes	Sub	Balancing difficulty	70, 90
18	CS	AL, I	VA, CA, CN	PM	No	No	Sub	Dysphagia, vocal cord paralysis, new CN XI, XII deficits, brain infarction	70, 75
19	CS	Lat, IE	VA, BA, CA, CN	FL	Partial	Yes	Sub	Infection, meningitis, hydrocephalus, Dysphagia, CSF leak, respiratory failure	70, 75
20	CS	Lat, I	VA, CN	PM	No	No	Sub	Dysphagia, motor sensory, new CN XI deficit, respiratory failure, quadriplegia	50, 20
21	CS	AL, IE	VA, CA, CN	PM	No	No	Total	Dysphagia, vocal cord paralysis,	75, 85
22	CS	Lat, I	VA, CA	FL	No	No	Sub	Infection, meningitis, hydrocephalus	N/A

CS, craniospinal; SC, spinocranial; Lat, lateral; Ant, anterior; AL, anterolateral; PL, posterolateral; I, intradural; IE, intraextradural; VA, vertebral artery; CN, cranial nerve; CA, cerebellar artery; BA, basilar artery; FL, far lateral approach; PM, posterial midline suboccipital approach; Lat, lateral approach; sub, subtotal removal.

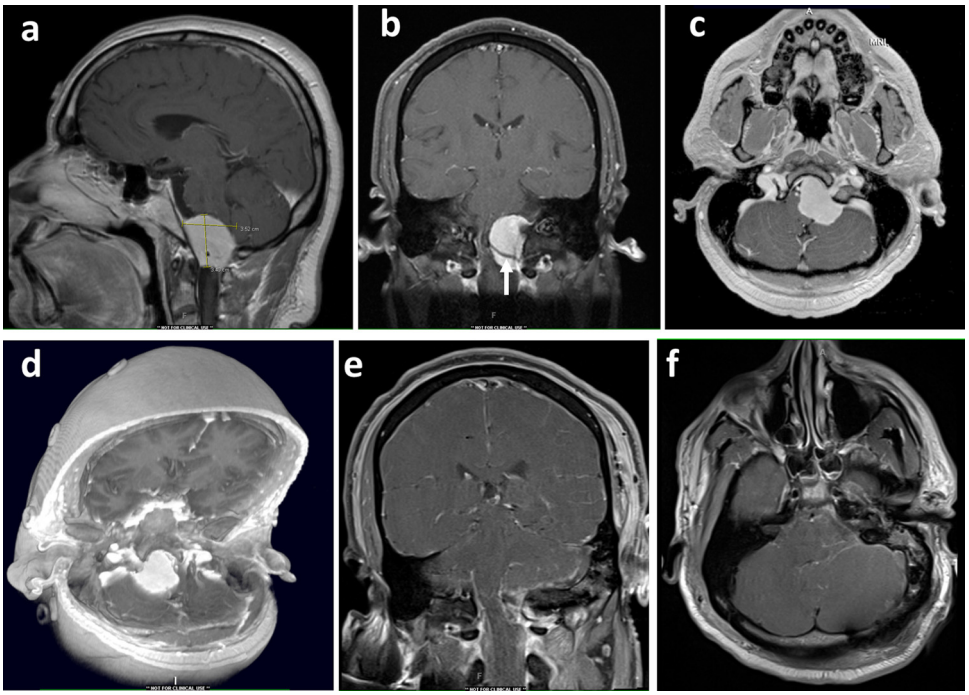

Fig. 2. This 45-year-old male patient experienced neck pain, headache, vertigo and numbness in his left thigh. MRI scans (a–c) and CT reconstruciton (d) revealed a very large craniospinal meningioma (3.4 cm × 3.5 cm × 2.5 cm) displacing significantly the brain stem laterally and wrapped around the vertebral artery (arrow). He underwent left suboccipital craniectomy with a far-lateral approach through a transcondylar route to the posterior fossa. Post-operatively, he recovered reasonably well after a transient dysphagia after surgery. He developed some difficulty in temperature sensation on the left side of the trunk. MRI (e and f) at follow-up showed evidence of small amount of enhancement at the level of the insertion of the tumor, but no significant compression of the brain stem.

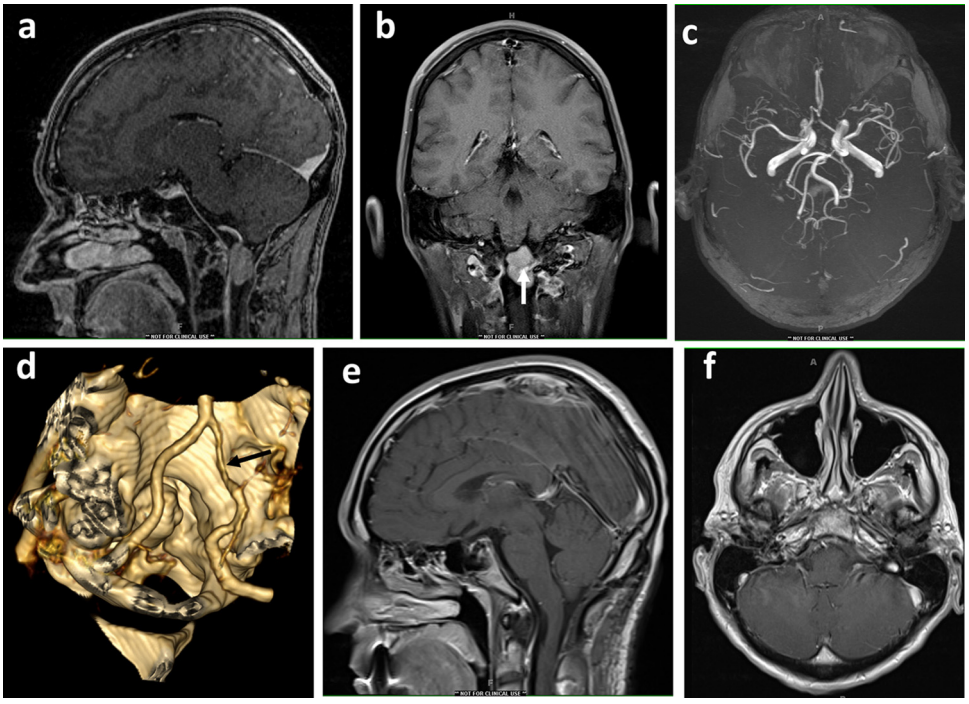

Fig. 3. This 36-year-old male patient presented with numbness in both hands, headache and gait instability. MRI scans (a–b) revealed an anterolateral spinocranial meningioma (3 cm × 1.9 cm × 1.6 cm) encasing the vertebral artery (arrow). Pre-operative CT angiogram (c) and intra-operative MRA (d) shows the narrowing of the branch of the VA encased by the tumor. He underwent a suboccipital craniectomy and C1 laminectomy with left lateral approach to the C1 and foramen magnum area with an exposure of the vertebral arteries. Post-operatively, he recovered well except developing some atrophy of the infraspinatus and supraspinatus muscle and possibly a part of the deltoid. MRI (e and f) at follow-up showed complete resection of the tumor.

Table 2
Preoperative characteristics.

Patient characteristics	Total (%)
Age	54.0 ± 15.9
Male	7 (31.8)
Co-morbidities	
CAD	1 (4.76)
Hyperlipidemia	6 (28.57)
Diabetes	4 (19.05)
Osteoporosis	0 (0)
Obesity	2 (9.52)
Smoking	3 (14.29)
COPD	0 (0)
Hypertension	9 (42.86)
Depression	2 (9.52)
Previous surgery	2 (9.52)
Symptoms	
Neck pain	12 (57.14)
Headache	12 (57.14)
Facial pain	2 (9.52)
Dysphagia	4 (19.04)
Ataxia	8 (38.1)
Motor weakness	5 (23.81)
Dizziness/vertigo	5 (23.81)
Cranial nerve deficits	3 (14.29)
Sensory changes	15 (71.43)
Hydrocephalus	0 (0)
Hemiparesis	0 (0)
Fatigue	5 (23.81)
Bowel/bladder dysfunction	4 (19.05)

3. Results

3.1. Patient data

The current series includes 22 patients (15 females and 7 males; average age: 54 years; range: 30–96) who underwent surgery for resection of craniocervical junction meningiomas between August 1995 and May 2012. The mean follow-up time was 46.5 months. The mean duration of disease, which is time between symptoms to admission, was 21.5 ± 24.67 months. Symptoms and neurological signs are listed in Table 2. The predominant symptoms at the time of presentation were neck pain, headache and sensory changes (mainly hypoesthesia, paresthesias and dysesthesias). Preoperative Disability was assessed by the Karnofsky score: 8 patients were able to carry put daily activities entirely independently (>70), 12 only sufficient for self-care (70–60) and 2 required total assistance (<60) (Table 1).

Seven (32%) meningiomas were spinocranial and 15 (68%) were craniospinal. In terms of position relative to the spinal cord and brainstem: 2 (9%) tumors were anterior, 7 (32%) were anterolateral, 12 (55%) were lateral, and 1 (4%) was posterolateral. In 17 cases (77%), the tumor was intradural and in 5 cases, intra- and extradural (23%) (Table 3). All tumors were encapsulated, and all were operated on for the first time.

3.2. Surgical approach and operative findings

A variety of surgical approaches were employed (Table 4). The posterior midline approach was used in 9 cases (41%), the far lateral approach in 12 cases (54%), and the lateral approach in 1 case (4%) (Table 4). The vertebral arteries (VA) were involved in 12 cases (55%) and the basilar artery in 1 case (5%). In most of those cases, the vasculature structure was encased by the tumor (Table 3). Cranial nerves appeared to be encased radiographically or intraoperatively in 15 cases (68%), although patients exhibit symptoms of pre-operative cranial nerve deficit in only 3 cases (14%).

Table 3
Tumor types.

Tumor classifications	Total (%)
Location	
Spinocranial	7 (31.82)
Craniocervical	15 (68.18)
Anterior	2 (9.09)
Anterolateral	7 (31.82)
Lateral	12 (54.55)
Posterior	0 (0)
Posterolateral	1 (4.55)
Type	
Capsule	22 (100)
En plaque	0 (0)
Intradural	17 (77.27)
Intraextradural	5 (22.73)
Extradural	0 (0)
Recurrent tumor	2 (9.09)
Arteries/CN involved	
Vertebral arteries	12 (54.55)
Basilar arteries	1 (4.55)
Carotid arteries	0 (0)
Cerebellar arteries	5 (22.73)
Cranial nerves	15 (68.18)

3.3. Surgical results

The median hospital stay was 5 days. Total resection was achieved in 10 cases (45%) and subtotal removal in 12 cases (55%) (Table 4). In the subtotal removal group, remnants were left attached to the vertebral or basilar arteries. Two of the 12 patients with subtotal removal underwent radiotherapy after the operation. These patients were followed with routine MRI scans and in all cases, the residual tumor remained stable over the years. No patients with subtotal removal required re-operation. Peri-operative complications are summarized in Table 4. The most prevalent complication after surgery was dysphagia, which

Table 4
Intra-operative variables and post-operative complications.

Intra-operative variables	Total (%)
Surgical approach	
Posterior midline	9 (40.91)
Lateral	1 (4.55)
Far lateral	12 (54.55)
Extreme lateral	0 (0)
Tumor removal	
Total	10 (45.45)
Subtotal	12 (54.55)
Complications	Total (%)
Infection	4 (18.2)
Meningitis	2 (9.1)
Hydrocephalus	2 (9.1)
Pneumocephalus	0 (0)
Pneumonia	0 (0)
GI hemorrhage	0 (0)
CN deficit (IX, X)	10 (45.4)
Feeding tube	5 (22.7)
Long-term deficit[a]	0 (0)
CN deficit (others)	8 (36.4)
Long-term deficit[a]	1 (5.3)
Bowel/bladder disturbance	1 (4.6)
Long-term bowel/bladder disturbance[a]	0 (0)
New muscle weakness	2 (9.1)
Long-term weakness[a]	0 (0)
CSF leak	2 (9.1)
Craniocervical instability	0 (0)
Respiratory failure	3 (13.6)
New onset ataxia	2 (9.1)
Brain infarct	1 (4.6)
Quadriplegia	2 (9.1)

[a] For long term symptoms, 19 patients were analyzed, excluding 2 patients whose follow-up time was less than 3 months and 1 patient who died peri-operatively.

Table 5
Neurological scoring system.

Score	Sensory deficits, pain, dysesthesias	Motor weakness	Gait ataxia	Bladder function	Bowel function
5	No symptom	Full power	Normal	Normal	Normal
4	Present, not significant	Movement against resistance	Unsteady no aid	Slight dist., no catheter	Slight dist., full control
3	Significant, function not restricted	Movement against gravity	Mobile with aid	Residual, no cath.	Laxatives, full control
2	Some restriction of function	Movement without gravity	Few steps with aid	Sometimes catheter	Sometimes loss of control
1	Severe restriction of function	Contraction without	Standing with aid	Often catheter	Often loss of control
0	Incapacitated	Plegia	Wheel chair	Permanent catheter	No control

Dist. = disturbance; Cath. = catheter.

was present in 10 cases (45%). In 5 cases, a feeding tube was necessitated. Of the 10 patients with post-operative dysphagia, the majority (70%) were able to swallow a sufficient solid diet independently 1 year after surgery. The second most common complications after surgery was new cranial nerve deficit, most commonly associated with CN XI, XII, VI and VII.

We scored patient neurologic outcomes via the model proposed by Klekamp and Samii [4] (Table 5). Scores between 3 and 5 indicate satisfactory levels of function and scores between 0 and 2 indicate unsatisfactory or severely compromised function. The scores of the 22 patients were averaged at various time points (Pre-op, at discharge, 1–2 months, 6–12 months and last follow-up) and plotted over time, allowing for visualization of a dynamic change in clinical picture (Fig. 4a–g). Hypoesthesias, paresthesias, pain and sphincter disturbance generally improved steadily after surgery. However, although weakness and gait ataxia generally improved at the last follow-up compared to pre-op, there is a noticeable transient worsening of the symptoms right immediately the surgery. The average score for cranial nerve deficits slightly worsened at last follow up compared to pre-op. In terms of cranial nerve deficits, among the 22 patients, 4 patients improved (18.1%), 16 remain unchanged (72.7%), and 2 worsened (9.2%), consistent with previous reports [3,5]. Functional recovery (Karnofsky score >70) was achieved in all but 2 patients (90.9%) who presented with severe disability before the surgery. There was one case of mortality in our series. The patient had multiple cardiovascular risk factors and unfortunately developed bilateral cerebellar and inferior vermian infarcts post-operatively, which were responsible for his near "locked in" syndrome. He also developed cervical medullary edema resulting in bulbar dysfunction resulting in respiratory failure, for which he underwent tracheostomy and PEG placement. His family decided in keeping with his wishes to withdraw aggressive treatment and move toward comfort measures. The patient died 3 weeks after the operation.

4. Discussion

4.1. Surgical approach

Various surgical approaches have been advocated for craniocervical junction meningiomas, depending on the anatomical location of the tumor relative to the brainstem. Tumors situated posterior or posterolateral to the spinal cord or the brainstem can be safely resected via a posterior midline suboccipital approach combined with C-1 laminectomy. Tumors situated anteriorly [6] may be accessed with the far-lateral approach described by Heros [7], or the extreme lateral modification described by George et al. [8].

In our case series, the posterior midline approach was used for 9 cases (40.9%) mostly for spinocranial meningiomas. In some cases, this approach was also used to remove craniospinal tumors that have a lateral component. For craniospinal tumors that are anteriorly based, the far lateral approach was frequently utilized. In our experience, there is no significant correlation between tumor resection status (total versus subtotal) and surgical approaches: 6/9 (66.7%) patients who received surgery via a midline posterior approach had total resections, compared to 4/13 (30.8%) patients who had lateral retrosigmoid or posterolateral approaches ($p = 0.1920$). Additionally, there was no significant correlation between surgical approaches and new cranial nerve deficit post-operatively ($p = 0.6619$) or change in the Karnofsky score ($p = 1.000$).

4.2. Drilling of the occipital condyle and mobilization of the vertebral artery

In order to achieve a wider surgical corridor, surgeons have combined the traditional surgical approaches with partial drilling of the occipital condyle and transposition of the vertebral artery [1,6,8–17]. There has been considerable controversy in the neurosurgical literature about the extent of occipital condyle drilling in these cases, with some surgeons supporting partial condyle resection [1,11,18–21], and others advocating against resection altogether [3,20,22]. In our series, partial condylar resection was performed in 9 cases (40.9%), and total resection in only 1 case (4.5%). Our experience is similar to that of Pamir et al. [15] and Goel et al. [20], where with the exception of small anterior tumors, debulking large tumors frequently creates sufficient surgical space enabling access to the anterior area. In cases with limited access to an anteriorly located tumor, the far-lateral approach combined with a partial condylectomy was utilized.

In some cases, complete resection of anteriorly based tumors may require mobilization of the vertebral artery [23]. Although some surgeons deem transposition of the vertebral artery necessary to facilitate condylar drilling [1,6,9,14,16,17,24], other investigators have found mobilization of the artery not to be necessary [10,13]. As the vertebral artery carries the highest risk of injury, we attempted to leave it intact except in cases where it was necessary to release the vertebral artery from its dural attachment in order to gain sufficient access to the anterior most part of the tumor. In our series, the vertebral artery was mobilized only in 4 cases (18.1%).

4.3. Surgical and clinical outcomes

A review of the surgical and clinical outcomes in the literature is presented in Table 6. Differences in total resection rate likely reflect the differences in tumor recurrence, extradural growth, vasculature and cranial nerve encasement, which are all widely accepted as factors against radical resection. We showed that vertebral artery involvement significantly affected the rate of radical resection. 7/9 (77.8%) patients who had no artery involvement had total resections, which was statistically higher compared to 3/13 (23%) patients who did have VA involvement ($p = 0.0274$). VA involvement also tended to impact clinical outcome. 1/11(9.1%) patients who had arterial involvement had increases in Karnofsky scores, which approached but did not reach statistical significance compared to 4/9 (44.4%) patients who improved without arterial involvement ($p = 0.1194$). However, VA involvement had no significant effect on new cranial nerve deficit post-operatively ($p = 0.3742$). In contrast, we failed to find any significant

Table 6
A review of the craniocervical junction meningioma literature.

Authors	Year	Nb Pt	Tumor location (%)			Approach	VA encasement (%)	CN encasement (%)	Complications	Total removal (%)	Mortality (%)	Recurrence (%)	OC resection	VA mobilization
			Ant	Lat	Post									
Yasuoka et al. [31]	1978	37	~60	~20	~20	PM	–	–	Respiratory failure, extradural hemorrhage	100	3	0	–	–
Guidetti et al. [32]	1980	11	27, 55 (AL)	0	18 (PL)	PM	–	–	–	–	11	0	–	–
Meyer et al. [33]	1984	78	60	20	10	PM	–	–	–	–	5	–	–	–
Guidetti et al. [34]	1988	17	23, 59 (AL)	0	18 (PL)	PM	6	–	Lateromedullary infarction, respiratory failure	100	12	0	100	–
Kratimenos et al. [14]	1993	7	14, 86 (AL)	0	0	FL	–	–	Facial nerve palsy, respiratory failure	86	29	12.5	–	100
Babu et al. [9]	1994	9	100	0	0	EL	–	–	CN deficit, CSF leak, meningitis	88	13	0	100	100
Akalan et al. [35]	1994	8	12.5, 87.5 (AL)	0	0	PM	–	–	–	100	0	0	0	0
Bertalanffy et al. [11]	1996	19	100 (incl. AL)	0	0	FL	–	–	No	100	0	0	100	0
Samii et al. [3]	1996	38	95	5	5	PM, DL	40	–	Infection, hydrocephalus, pneumonia, CN deficit	63	6	5	18	–
George et al. [13]	1997	40	45	52.5	2.5	PL, AL, PM	38	–	No	94	8	0	78	–
Pirotte et al. [16]	1998	6	100 (AL)	0	0	TC	–	–	Hemiplegia, palsy, hydrocephalus	100	17	–	100	100
Salas et al. [24]	1999	24	100	0	0	EL variations (TFA, RCA, PTCA, CTCA, ETJA, TTA)	43	–	Hydrocephalus, CSF leak, CN deficit	66	4	–	75 (PTCA, CTCA, TTA)	79 (TFA, PTCA, CTCA, ETJA)
Sharma et al. [36]	1999	10	50	0	50	PM, FL	–	–	–	100	15	–	0	0
Arnautovic et al. [1]	2000	18	100	0	0	TC	–	–	CN deficits (IX, X)	75	0	11.1	>75	–
Goel et al. [20]	2001	17	100	0	0	SO	59	–	CN deficits	82	0	0	–	–
Roberti et al. [27]	2001	21	–	–	–	EL, TC	–	–	CN deficits	76	10	–	–	–
Nanda et al. [22]	2002	6	100	0	0	FL	–	–	–	100	0	0	0	100
Marin et al. [37]	2002	7	71	0	29	TC, PM, TO	–	–	Tetraparesis, lower CN paresis	100	14	0	28	28
Parlato et al. [6]	2003	7	–	–	–	PTCA, RCA	–	–	Dysphagia, CSF leak	86	0	0	85	85
Boulton et al. [2]	2003	10	60 (incl. AL)	10	30	PM, TC	–	–	CSF leak	90	0	0	0	–
Pamir et al. [15]	2004	22	91	0	9	FL, PM	40	–	CN palsies and VA injury	95	0	0	96	–
Margalit et al. [28]	2005	18	100	0	0	Lat	67	–	CN deficit, CSF leak	61	6	6	64	100
Bassiouni et al. [38]	2006	25	36, 64 (AL)	0	0	FL	43	–	CSF leak, CN XI deficit	96	4	0	0 (intradural)	0
Borba et al. [39]	2009	15	53	47	0	Lat	–	–	CN deficits	80	7	7	53	–
Kandenwein et al. [30]	2009	16	19	75	6	Posterier SO	88 (Adhere)	–	CN deficits	88	6	0	–	0
Wu et al. [26]	2009	114	70, 21 (AL)	0	9 (PL)	FL, PM, extended FL	40	–	Dysphagia, Dyspnea	86	2	1	9	–
Kano et al. [25]	2010	23	39	61	0	SO, TC	–	–	CN palsies	62	0	4	Very low	0
Bruneau et al. [40]	2010	107	39	55	6	PL	5	–	–	86	2	1	–	–
Pirotte et al. [29]	2010	22	38	62	0	PL	38	42	CN palsies (23%), CSF leak (7%)	73	4	–	38	73
Talacchi et al. [5]	2012	64	38, 62 (AL)	0	0	DL	39	IX-X: 34 XII: 31 XI: 27	Dysphagia, cranial nerve deficits	81	0	0	55	0
Present study	2013	22	9, 32 (AL)	55	4 (PL)	PM, FL, Lat	55	68	CN deficit, infection	45	4	0	45	18

PM, posterior midline; FL, far lateral; DL, dorsal lateral; EL, extreme lateral; Lat, lateral; PL, posterolateral; TC, transcondylar; SO, suboccipital; TO, transoral; AL, anterolateral; TFA, transfacetal approach; RCA, retrocondylar approach; PTCA, partial transcondylar approach; CTCA, complete transcondylar approach; ETJA, the extreme-lateral transjugular approach; TTA, transtubercular approach.

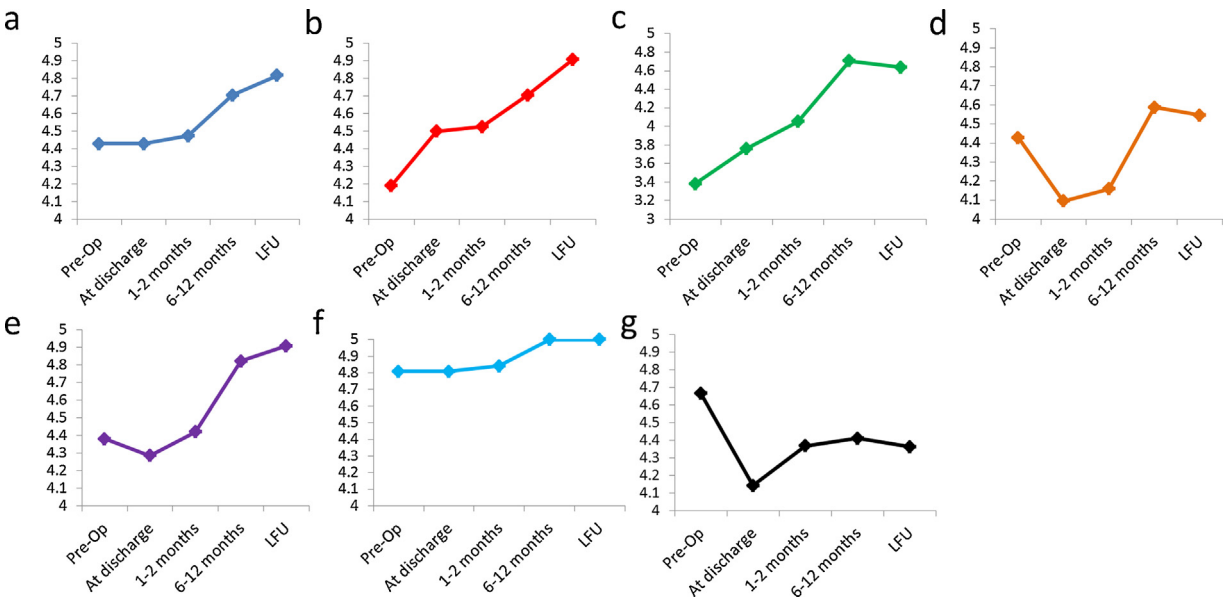

Fig. 4. Change in clinical pictures. The neurological scale is described in Table 5. (a) Hypoesthesias, (b) paresthesias, (c) pain, (d) motor weakness, (e) gait ataxia, (f) sphincter disturbance, and (g) cranial nerve deficits.

correlation between cranial nerve encasement by the tumor and tumor resection status ($p = 0.1718$) or new post-operative cranial nerve deficit ($p = 0.1932$) or change in Karnofsky score ($p = 0.6126$).

Total resection of craniocervical junction meningiomas in previous studies was typically achieved in 61–100% of the cases (Table 6). In our series, tumors that engulfed critical neurovascular structures were treated conservatively; therefore, total removal was not attempted. Our series demonstrates that generally patients who underwent total resection had better outcomes than those with subtotal resection. Notably, 9/10 (90.0%) patients who had a total resection had increases in Karnofsky scores, which was statistically higher compared to 7/11 (63.6%) patients who improved after subtotal resections ($p = 0.0367$). Radiotherapy was used in conjunction with surgery in 2 cases in this study. We employed early post-operative radiation when there was substantial infiltration of the tumor to the surrounding structures or when there was significant residual tumor left. We also prescribed radiotherapy in cases where there was a change in the size of the residual tumor during follow-ups.

Multiple studies have shown that the most common complication post-operatively is cranial nerve IX and X, and XII deficit, occurring in 39.1–55.5% of the cases [1,5,24–28]. In our series, dysphagia caused by cranial nerve IX and X deficit remains to be the most serious complication, occurring in 10 cases (45.4%). In 5 cases, this required feeding tube placement. The second most frequent complication was impairment of other cranial nerves (mainly XI and XII), occurring in 7 cases (31.8%). However, they were transient in almost in all cases (Table 4) and did not contribute to the final Karnofsky score. In some studies, CSF leak has been described as the most common post-operative complications, occurring in nearly 16–20% of patients [15]. In contrast, we only had 2 cases (9.1%) of CSF leak in our series. The overall post-operative morbidity in our series is 72.7%, consistent with the reported literature [3,5,27,29,30]. Two patients developed quadriplegia post-operatively. One patient had no movement in either of the upper or lower extremities consistent with likely spinal cord injury. He had relatively preserved sensation which made his injury suggestive of a medullary pyramidal disturbance. The other patient developed bilateral cerebellar and inferior vermian infarcts. The right cerebellar infarct was in the right superior cerebellar artery distribution which was responsible for his poor neurological exam and near "locked in" syndrome. One patient developed acute infarction in the right frontal lobe. However, the infarction was very small and did not cause any noticeable neurological deficits.

In the current series, the mean Karnofsky score of the patients improved from 70.5 pre-operatively to 82.3 at the last follow-up, with 66.7% of the patients having a normal life (Karnofsky score of 80–100). The Karnofsky score improved in 76.2%, remained unchanged in 9.5% and worsened in 14.3% of the patients. The final outcomes of our patients compare favorably with others in the literature, despite the advanced age of our patients ($n = 9$ where age >65 years) and serious pre-operative functional impairment ($n = 13$ where Karnofsky score ≤ 70).

4.4. A comparison of craniospinal tumors and spinocranial tumors

Craniospinal and spinocranial meningiomas differ in their origin, anatomical location, and interaction with neighboring structures. In this series, we compared these two types of tumors in terms of patient data, tumor size, surgical approach, surgical outcome and clinical improvement. The results are summarized in Table 7. Spinocranial tumors are associated with a higher rate of total resection (100% vs. 20%, $p = 0.0007$), fewer post-operative CN IX/X deficits (0% vs. 66.7%, $p = 0.0053$) and shorter hospitalization (3.4 ± 1.3 days vs. 12.57 ± 18.0 days, $p = 0.08$). There was no significant difference in the percentage of patients with improved Karnofsky score (85.7% vs. 71.4%, $p = 1.0$). Additionally, the mean tumor volume of the spinocranial tumors was $4.6 \pm 2.0\,cm^3$. While the difference was not statistically significant, the spinocranial tumors were smaller than the craniospinal tumors (mean volume $12.2 \pm 9.2\,cm^3$, $p = 0.0724$). This may be due to the fact that spinocranial tumors impinge on the spinal cord earlier than their craniospinal counterparts, thereby causing symptoms at a smaller size and resulting in earlier diagnosis.

It should be noted that the current study is retrospective in nature and shares some common limitations associated with this type of study, including selection bias, multiple surgeons operating and multiple residents involved in care. Despite these limitations, ours is one of the largest studies on craniocervical junction meningiomas and will contribute to a better understanding of the

Table 7
A comparison of craniospinal and spinocranial tumors.

Characteristics	Total cases (%)	Craniospinal (%)	Spinocranial (%)	*p*
No. cases	22	15 (68.2)	7 (31.8)	–
Age	53.7 ± 12.5	49.3 ± 15.0	59.7 ± 6.5	0.2778
Sex (male, female)	7 (31.8), 15 (68.2)	7 (46.7), 8 (53.3)	0 (0.0), 7 (100.0)	0.0513[b]
Length of hospitalization	9.5 ± 15.1	12.57 ± 18.0	3.4 ± 1.3	0.0800[b]
Symptom onset until surgery	21.5 ± 24.7	26.2 ± 27.0	10.3 ± 14.4	0.1398
Tumor volume (cm^3)	9.5 ± 8.2	12.2 ± 9.2	4.6 ± 2.0	0.0724[b]
Surgical approach				
Posterior midline approach	9 (40.9)	5 (33.3)	4 (57.1)	0.3762
Far lateral approach	12 (54.5)	9 (60.0)	3 (42.8)	0.6517
Resection of OC	10 (45.4)	9 (60.0)	1 (14.3)	0.0743[b]
Total removal	10 (45.4)	3 (20.0)	7 (100.0)	0.0007[a]
Surgical complications				
Infection	4 (18.2)	3 (20.0)	1 (14.3)	1.0
Meningitis	2 (9.1)	2 (13.3)	0 (0.0)	1.0
Hydrocephalus	2 (9.1)	2 (13.3)	0 (0.0)	1.0
CN deficit (IX, X)	10 (45.4)	10 (66.7)	0 (0.0)	0.0053[a]
CN deficit (others)	7 (31.8)	5 (33.3)	2 (28.6)	1.0
New muscle weakness	2 (9.1)	2 (13.3)	0 (0.0)	1.0
CSF leak	2 (9.1)	1 (6.7)	1 (14.3)	1.0
Respiratory failure	3 (13.6)	3 (20.0)	0 (0.0)	1.0
New onset ataxia	2 (9.1)	2 (13.3)	0 (0.0)	1.0
Quadriplegia	2 (9.1)	2 (13.3)	0 (0.0)	1.0
Clinical improvement				
Hypoesthesia	6 (30.0)	4 (23.5)	2 (28.6)	1.0
Paresthesia	8 (40.0)	6 (46.1)	2 (28.6)	0.6424
Pain	15 (75.0)	9 (69.2)	6 (85.7)	0.6126
Weakness	2 (10.0)	1 (7.7)	1 (14.3)	1.0
Ataxia	4 (20.0)	3 (23.1)	1 (14.3)	1.0
Sphincter disturbance	3 (15.0)	2 (15.4)	1 (14.3)	1.0
CN deficit	1 (5.0)	1 (7.7)	0 (0.0)	1.0
Karnofsky score	16 (76.2)	10 (71.4)	6 (85.7)	1.0

[a] Statistically significant ($p < 0.05$)
[b] Approaching statistical significance.

pathophysiology of the disease as well as an improvement in the surgical management.

5. Conclusions

Surgical management of craniocervical junction meningiomas continues to present as a challenge to neurosurgeons especially in cases involving neighboring neurovascular structures. Here we summarize our experience at a single institution of 22 patients over 17 years. We show that patients who had vertebral artery involvement were more likely to receive subtotal resection of their tumor. Our series demonstrates that patients who underwent total resection had statistically better outcomes than those with subtotal resection. Interestingly, we found that spinocranial and craniospinal tumors have distinct characteristics and therefore should be managed differently. We demonstrrate that the posterior midline approach is often appropriate for spinocranial meningiomas while the far lateral approach can be utilized for craniospinal tumors that are anteriorly based. Spinocranial tumors are often detected at a smaller size and treated earlier. They are also associated with a higher rate of total resection, fewer postoperative CN IX or X deficits, and shorter hospitalizations.

Disclosures

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