

# Surgical Management of Extracranial Meningiomas Arising in the Head and Neck



Moustafa Mourad, MD,\* David Chan, MD,† and Yadranko Ducic, MD‡

**Purpose:** To determine outcomes in the management of extracranial meningiomas of the head and neck.

**Patients and Methods:** This is a retrospective single-surgeon series performed at a tertiary-care referral center. In all, 23 patients met the inclusion criteria, consisting of 12 men and 11 women. The mean age of patients treated was 60.5 years (range, 32 to 71 years). Subsite involvement included the infratemporal fossa (n = 8), greater wing of the sphenoid and orbit (n = 7), clivus (n = 2), and parapharyngeal space (n = 6).

**Results:** In all, recurrence occurred in 21% of patients (n = 5) who underwent gross tumor resection. Two patients underwent subtotal resection because of the tumor's location within the clivus with adjuvant CyberKnife therapy (Accuray, Sunnyvale, CA). Both patients had persistent disease with no new neurologic symptoms. Examination showed that 100% of patients (N = 23) had dural involvement. Post-ablative complications occurred in 43% of patients (n = 10).

**Conclusions:** We presently report the largest series of surgical treatment for extracranial meningiomas. Gross tumor resection should be the mainstay of therapy, except in anatomically restricted regions such as the petrous apex and clivus. Adjuvant therapy including CyberKnife therapy may be used in such lesions. All lesions showed dural involvement. A more unified nomenclature is required for the characterization of these lesions.

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Meningiomas are nonglial neoplasms arising from arachnoid granulation from the axial or intracranial central nervous system.<sup>1</sup> Meningiomas are the most common intracranial tumors of nonglial origin, constituting 18% of all primary intracranial tumors.<sup>1</sup> In contrast, extracranial and extra-axial meningiomas are rare entities, occurring at a rate of 1 to 2%, most commonly documented within the head and neck.<sup>2,3</sup>

Different authors have proposed classification systems distinguishing different types of extradural meningiomas with critical implications in their management.<sup>3,4</sup> In 1960 Hoyer et al<sup>4</sup> distinguished extracranial meningiomas based on the site of origin. In the computed tomography (CT) era, Lang et al<sup>3</sup> further classified primary extradural meningiomas

(Hoyer types B and C) based on the relative origin to the calvaria. However, because of the paucity of reported cases within the literature, most of which were reported in the pre-CT era, in addition to poorly defined nomenclature, the biological behavior of true extracranial meningiomas is poorly understood.<sup>3</sup> We document the largest case series to date of extracranial lesions arising within the head and neck.

## Patients and Methods

After approval by the John Peter Smith Institutional Review Board, all patients with extracranial meningiomas treated at our institution between 1997-2013 were identified. Only patients with a minimum of

\*Resident Physician, Division of Facial Plastic Surgery, Department of Otolaryngology-Head & Neck Surgery, The New York Eye & Ear Infirmary, New York, NY.

†Fellow Physician, Department of Otolaryngology Head and Neck Surgery, Otolaryngology and Facial Plastic Surgery Associates, Fort Worth, TX.

‡Professor, Department of Otolaryngology Head and Neck Surgery, Otolaryngology and Facial Plastic Surgery Associates, Fort Worth, TX.

Address correspondence and reprint requests to Dr Ducic: Otolaryngology and Facial Plastic Surgery Associates, 923 Pennsylvania Ave, Ste 100, Fort Worth, TX 76104; e-mail: [yducic@sbcglobal.net](mailto:yducic@sbcglobal.net)  
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24 months' follow-up were included in the study. For the purposes of the study, only lesions with extracranial extension based on CT and/or magnetic resonance imaging, in addition to intraoperative findings during surgical resection, were included (Lang types I and III) (Fig 1). Tumors that had a clear intradural point of origin or were completely intracranial were excluded from the study (Lang type II). Tumors were classified according to the point of origin, dural involvement, and extension in relation to the calvaria. Demographic as well as patient clinical information was reviewed. Findings of the preoperative workup, including radiographic mapping of tumors, were analyzed. Intraoperative details were further reviewed, as were patient follow-up and disease-related outcomes. All patients were treated with primary surgery, with some patients receiving adjuvant radiotherapy. Robotic radiosurgery was used between 1998-2013 once the technology was made available at our institution. All remaining patients were treated with external beam radiotherapy (EBRT) or intensity-modulated radiotherapy at a minimum dose of 50 Gy. Complications were defined as new and permanent deficits or patient complaints arising after intervention.

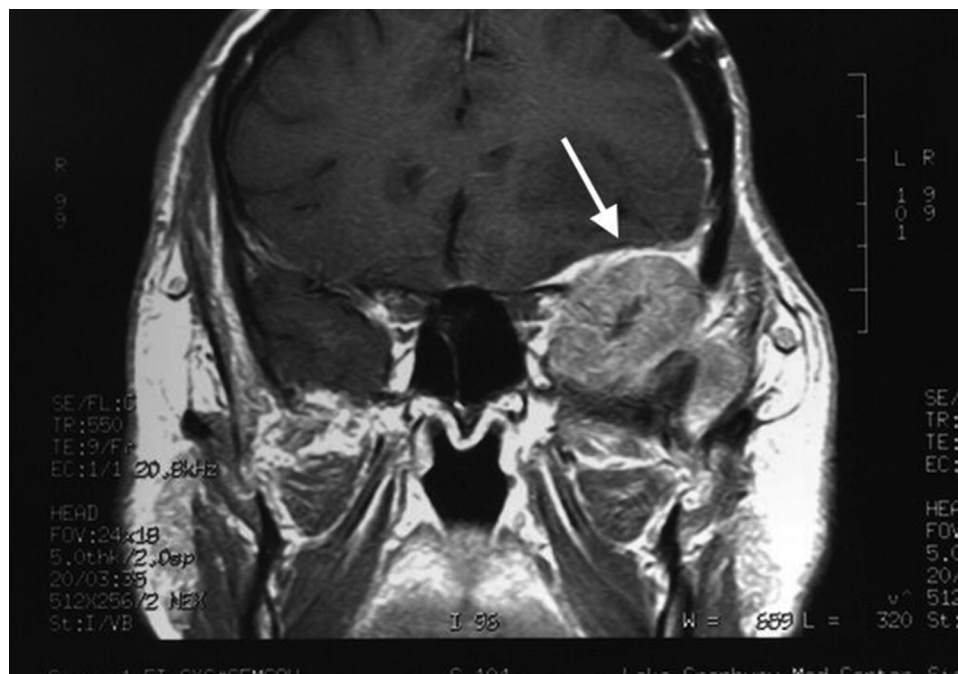
## Results

In all, 23 patients met the inclusion criteria, consisting of 12 men and 11 women. The mean age of

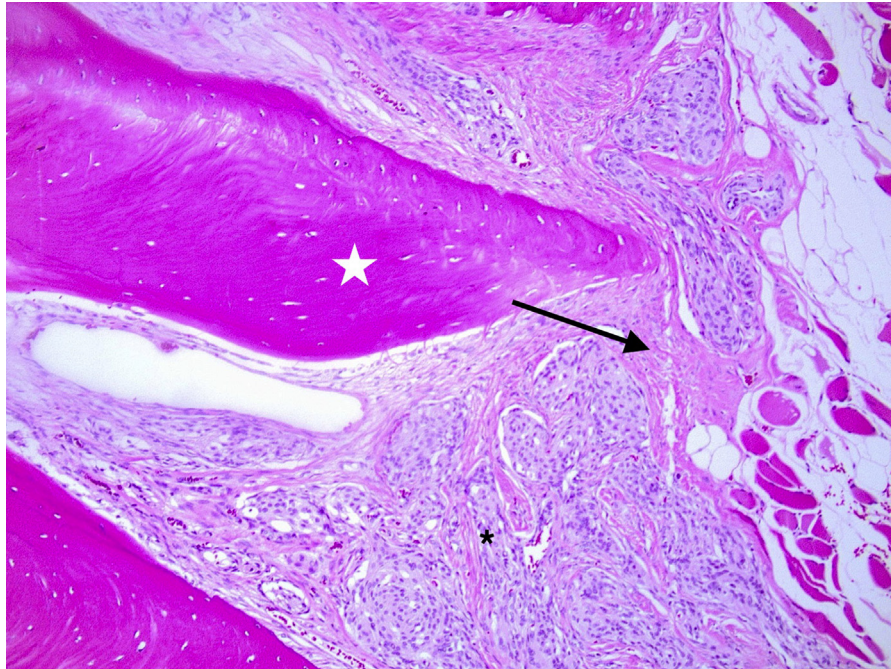
patients treated was 60.5 years (range, 32 to 71 years). Subsite involvement included the infratemporal fossa (n = 8), greater wing of the sphenoid and orbit (n = 7), clivus (n = 2), and parapharyngeal space (n = 6). Histopathologic review of the specimens showed 100% of lesions (N = 23) to be benign (grade I by World Health Organization [WHO] classification), without evidence of malignancy (WHO grade III) or atypical cells (WHO grade II). In total, 47.8% of patients (n = 11) received adjuvant radiotherapy, whereas 8% (n = 2) underwent subtotal resection because of the tumor's location. The recurrence rate was 26.1% (n = 6). Of note, 100% of lesions (N = 23) to some extent had involvement of the skull base and dura (Fig 2). No disease-related deaths or distant metastatic disease was noted in any patient (Table 1).

### GREATER WING OF SPHENOID AND ORBIT

In total, 7 patients were treated for extracranial tumors involving the greater wing of the sphenoid and orbit, consisting of 4 men and 3 women (Fig 3). The mean age at diagnosis was 49 years (range, 32 to 68 years). Adjuvant radiotherapy was performed in a single patient. All patients (n = 7) had preoperative proptosis, with 6 patients exhibiting decreased mobility of the extraocular muscles. In addition, 4 patients presented with decreased visual acuity. Postoperatively, movement normalized in 4 patients and persisted in 2 patients. The 2 patients with limited



**FIGURE 1.** T1-weighted magnetic resonance image with contrast in the coronal plane showing a Lang type III lesion with extracalvarial extension within the infratemporal fossa. The arrow indicates dural enhancement.



**FIGURE 2.** Histologic section showing relationship of calvarial bone (*star*) with meningioma (*asterisk*), showing intimate involvement with dura (*arrow*) (Hematoxylin and Eosin [H&E] stain, magnification  $\times 200$ ).

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extraocular muscle movement underwent ophthalmologic surgery to normalize the centric gaze. Therefore, no patients had permanent centric neutral gaze diplopia. Visual acuity was improved in 3 patients postoperatively and was stable in the remaining patient with previous complaints of decreased visual acuity. Local recurrence occurred within the dura in a single patient 3 years postoperatively and was treated surgically. All patients were disease free at last follow-up.

#### INFRATEMPORAL FOSSA

In total, 8 patients were treated for extracranial tumors involving the infratemporal fossa, including 5

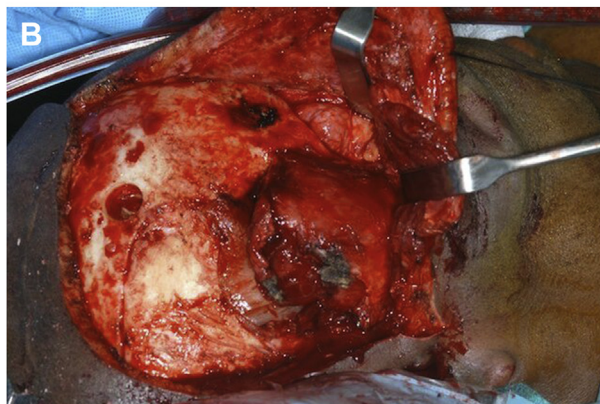
men and 3 women (Fig 4). The mean age at the time of diagnosis was 58 years (range, 49 to 71 years). All were treated with surgical excision through a combined intracranial and extracranial approach. In total, 4 patients received postoperative EBRT. Recurrence developed in 2 patients, neither of whom had received postoperative radiation. One patient with recurrence underwent salvage surgery 5 years postoperatively, whereas the second patient with recurrence died of an unrelated cardiac arrest, 2.5 years postoperatively. Subsequent complications included temporomandibular joint issues in 3 patients, permanent frontal nerve paralysis in a single patient, and House-Brackmann grade III palsy in 2 patients that resolved.

**Table 1. SUMMARY OF PATIENT DEMOGRAPHIC DATA, TREATMENT, AND OUTCOME-RELATED MEASURES**

	Total, n	Mean Age (Range), yr	Male/Female, n	Radiation	Recurrence	Complications
Infratemporal fossa	8	58 (49-71)	5/3	50% (4)	25% (2)	63% (5)
Greater wing of sphenoid and orbit	7	49 (32-68)	4/3	14% (1)	14% (1)	0%
Clivus and nasopharynx	2*	64 (61-66)	2/0	100% (2)	100% (2)	0%
Parapharyngeal space	6	59 (43-69)	2/4	100% (6)	33% (2)	83% (5)

\* Tumors of the clivus and nasopharynx were treated with subtotal resection followed by radiotherapy; recurrence was measured by disease progression.

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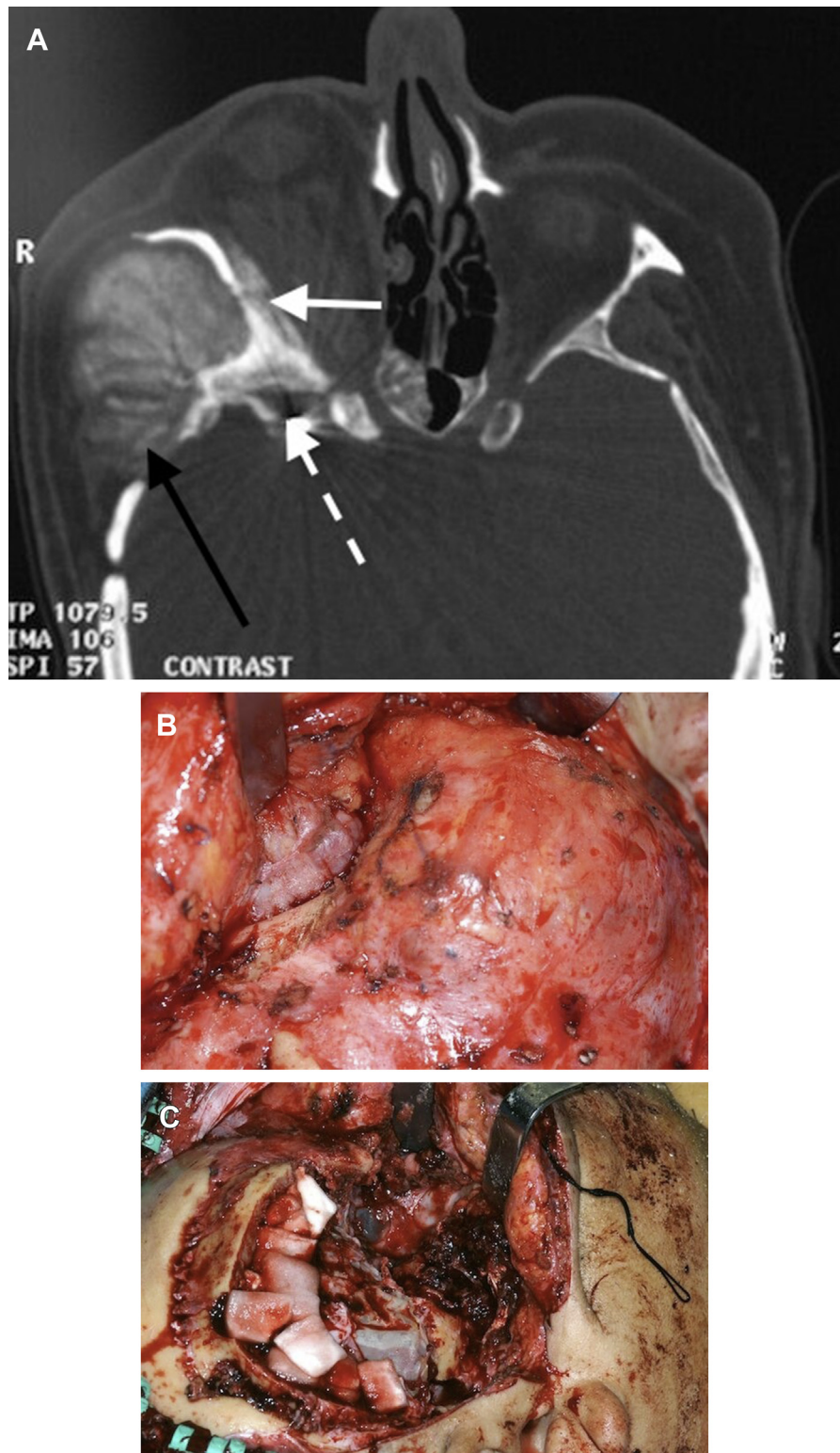
**FIGURE 3.** A, T1-weighted magnetic resonance image with contrast in the coronal plane showing a lesion involving the greater wing of the sphenoid and orbit (*black arrow*). The *white arrow* indicates secondary soft tissue edema. B, Intraoperative photograph showing coronal flap and tumor extent.

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#### CLIVUS AND NASOPHARYNX

Two patients, both of whom were men aged 66 and 61 years, had lesions involving the clivus. Both underwent subtotal resection with postoperative

CyberKnife radiation (Accuray, Sunnyvale, CA). One patient had no evidence of disease 4.5 years after combined treatment, with no evidence of new neurologic deficits. The other patient had progressive disease at



**FIGURE 4.** A, Computed tomography axial image with contrast showing an infratemporal fossa lesion (*black arrow*) with extension into the orbit (*white arrow*) and middle cranial fossa (*dashed arrow*). B, Intraoperative photograph showing extracalvarial extension. C, Postoperative defect after tumor excision.

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the clivus 18 months postoperatively, but died of unrelated metastatic lung carcinoma.

#### PARAPHARYNGEAL SPACE

In total, 6 patients had lesions involving the parapharyngeal space, including 4 women and 2 men. They presented at a mean age of 59 years (range, 43 to 69 years). All patients underwent complete resection with adjuvant radiotherapy. Postoperative EBRT was performed in 5 patients, with 1 patient receiving robotic radiosurgery. Recurrence occurred in 2 patients, of whom 1 received salvage surgery (4.5 years postoperatively). The other patient refused surgical salvage and had non-progressive disease 6 years postoperatively. Complications included temporomandibular joint issues (n = 4), facial palsy (n = 2), and osteoradionecrosis (n = 2). In the patients who had facial palsy, it was graded as House-Brackmann grades II and IV.

## Discussion

Extracranial meningiomas are rare tumors, accounting for less than 2% of all meningiomas, but are most often encountered in the head and neck.<sup>2,3</sup> The relative rarity, in addition to inconsistencies in nomenclature, limits the study of these lesions. Historically, they have been referred to as *extradural*, *extracranial*, *extraneuraxial*, and *ectopic tumors* arising outside of the central nervous system.<sup>5-7</sup> Different classification systems distinguishing different types of extradural meningiomas have been proposed, with the most widely referenced schemes proposed by Hoyer et al<sup>4</sup> (Table 2) and Lang et al<sup>3</sup> (Table 3). In 1960 Hoyer et al distinguished extracranial meningiomas based on the site of origin (Table 2). In the CT era, Lang et al further classified primary extradural meningiomas (Hoyer types B and C) based on the relative origin to the calvaria (Table 3).

There exists a lack of consensus on the presence of dural involvement, with implications in whether these lesions extend from central nervous system structures or if these are ectopically derived lesions. In 1960 Hoyer et al<sup>4</sup> noted the presence of arachnoid cell clusters at points of penetration of the dura where meningiomas were found. They therefore postulated that extracranial meningiomas may be extensions of intracranial tumors or may arise from a skull base foramen or cranial nerve, all with some degree of dural origin (Table 2).<sup>4</sup> Furthermore, extradural meningiomas arising from the spinal cord have all been implicated to extend from points of dural attachments.<sup>6,8,9</sup>

Other authors have classified extradural lesions as true ectopic tumors, without dural attachments.<sup>7,10,11</sup> Lopez et al<sup>10</sup> documented and characterized

**Table 2. HOYE CLASSIFICATION SYSTEM<sup>4</sup> FOR EXTRACRANIAL MENINGIOMAS DELINEATED BASED ON SITE OF ORIGIN**

Type A: extracranial extension of meningioma with intracranial origin (secondary)
Type B: extracranial extension of meningioma arising from skull base foramen (primary)
Type C: ectopic tumor without connection to foramen, cranial nerve, or intracranial extension (primary)
Type D: extracranial metastasis with documented intracranial lesion (secondary)

*Note:* Tumors with intracranial extension or origin are classified as secondary, with no associated cranial involvement denoted as primary.

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cutaneous meningiomas as follows: type I, primary cutaneous meningioma; type II, meningioma of the skin and soft tissue; and type III, meningioma of the central nervous system and skin. Type I lesions are the only true ectopic cutaneous lesions without any dural involvement, and these are thought to represent congenital abnormalities caused by trapping of arachnoid cell rests, trapped mesenchyme, delayed neural tube closure with herniation, or premature closure of the neural tube with meningeal trapping.<sup>10</sup> Other authors also have postulated that extradural meningiomas are ectopic tumors arising from ectopic meningocytes, leading to development of intraosseous lesions.<sup>11</sup> However, owing to most intraosseous lesions being related to cranial vault sutures, other authors have hypothesized that these extradural lesions may arise because of trapped arachnoid cell caps during development.<sup>12</sup>

The point of origin of these lesions becomes imperative in characterizing these lesions and their relationship to the dura. Some authors do not consider any lesions with any dural abutment or involvement as extradural because of the inability to rule out an

**Table 3. LANG CLASSIFICATION SYSTEM<sup>3</sup> FOR PRIMARY EXTRACRANIAL MENINGIOMAS DETERMINED BASED ON CALVARIAL OR EXTRACALVARIAL EXTENSION**

Type I: purely extracalvarial
Type II: purely calvarial
Subtype C: convexity lesion
Subtype B: skull base lesion
Type III: calvarial with extracalvarial extension
Subtype C: convexity lesion
Subtype B: skull base lesion

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intradural point of origin.<sup>13</sup> In a meta-analysis by Lang et al,<sup>3</sup> they determined that 62% of documented extradural meningiomas in the head and neck (n = 104), in addition to 60% of patients (n = 10) within their own series, had normal-appearing dura after craniectomy. They maintained that these tumors likely have epicenters in the extracalvarial space and likely do not involve the dura. Hoye et al,<sup>4</sup> however, maintained that a true ectopic lesion should not arise near cranial nerve foramina or have any intracranial extension. Therefore, given the proximity of the studied tumors within the head and neck to the central nervous system, as well as cranial nerves, the likelihood of being truly ectopic tumors without any dural relation should be better validated. Within our study, 100% of all head and neck extracranial meningiomas (N = 23) had some degree of dural involvement. Therefore, in the absence of metastatic disease, referring to these tumors as *extradural* does not explicate them as arising from an ectopic site.

In 2000 Lang et al<sup>3</sup> introduced a classification system to characterize extradural meningiomas in relation to the calvaria (Table 3). Type I tumors were characterized as tumors completely located outside of the cranium without attachment to bone, type II lesions were purely intracranial lesions, and type III lesions were intracranial lesions with soft tissue extension outside of the cranium. Nomenclature relating tumor location using the Lang classification does not replace referring to these lesions as *extradural*. Rather, the Lang characterization is a subclassification of extradural meningiomas. Within our study, we documented our experience with extracranial (type I and type III) lesions.

In total, 100 extracranial meningiomas have been documented in the CT-era literature.<sup>3</sup> Of these, 55 were purely extracalvarial (type I), whereas 65 tumors had extension from within the calvaria. In contrast, 100% of extracranial lesions (N = 23) treated at our institution had both an intracranial and extracranial component (type III). There was no gender predilection, in line with previous studies.<sup>3</sup> All lesions were identified as benign (WHO grade I) meningiomas.

Gross total resection is often recommended in the treatment of extracranial meningiomas, owing to the 5% disease-related mortality rate for benign lesions, as well as the 30% disease-related mortality rate for atypical or malignant lesions.<sup>14</sup> Complete resection was possible in most cases except for lesions of the clivus and nasopharynx. The 2 clival lesions treated at our institution underwent subtotal resection, followed by radiotherapy. Disease progression was noted in both patients. Only a single case of petro-clival meningioma has been documented in the literature (WHO grade II).<sup>15</sup> Similarly, there was disease progression

despite complete surgical excision in conjunction with radiotherapy, with the patient ultimately dying of the disease.

Radiotherapy is recommended for WHO grade I lesions after incomplete resection, recurrence, or inoperability because of medical comorbidities. Adjuvant radiotherapy is further recommended in all cases of WHO grade II and III lesions after total or subtotal resection.<sup>14</sup> Adjuvant radiotherapy was used in 57% of our patients (n = 13). The recurrence rate was 26.1% (n = 6), in line with systematic reviews.<sup>3</sup> Recurrence occurred in 4 patients who had received adjuvant radiotherapy.

In addressing extracranial meningiomas, a more unified nomenclature, as well as a consensus on the anatomic and pathophysiological origin of these lesions, is needed. The management of these lesions is largely surgical, with additional benefit from adjuvant radiotherapy. Complete surgical resection is often possible except in cases of clival lesions. Radiation has a pivotal role in cases of subtotal resection.

## References

1. Friedman CD, Costantino PD, Teitelbaum B, et al: Primary extracranial meningiomas of the head and neck. *Laryngoscope* 100: 41, 1990
2. Whicker JH, Devine KD, MacCarty CS: Diagnostic and therapeutic problems in extracranial meningiomas. *Am J Surg* 126:452, 1973
3. Lang FF, Macdonald OK, Fuller GN, DeMonte F: Primary extradural meningiomas: A report on nine cases and review of the literature from the era of computerized tomography scanning. *J Neurosurg* 93:940, 2000
4. Hoye SJ, Hoar CS, Murray JE: Extracranial meningioma presenting as a tumor of the neck. *Am J Surg* 100:486, 1960
5. Farr HW, Gray GF, Vrana M, Panio M: Extracranial meningioma. *J Surg Oncol* 5:411, 1973
6. Rosencrantz M, Stattin S: Extradural meningiomas. Report of two cases. *Acta Radiol Diagn (Stockh)* 12:419, 1972
7. Saade R, Hessel A, Ginsberg L, et al: Primary extradural meningioma presenting as a neck mass: Case report and review of the literature. *Head Neck* 37:E92, 2015
8. Rasmussen TB, Kernohan JW, Adson AW: Pathologic classification, with surgical consideration, of intraspinal tumors. *Ann Surg* 111:513, 1940
9. Hallpike JE, Stanley P: A case of extradural spinal meningioma. *J Neurol Neurosurg Psychiatr* 31:195, 1968
10. Lopez DA, Silvers DN, Helwig EB: Cutaneous meningiomas—A clinicopathologic study. *Cancer* 34:728, 1974
11. Azar-Kia B, Sarwar M, Marc JA, Schechter MM: Intraosseous meningioma. *Neuroradiology* 6:246, 1974
12. Arana E, Diaz C, Latorre FF, et al: Primary intraosseous meningiomas. *Acta Radiol* 37:937, 1996
13. Crawford TS, Kleinschmidt-DeMasters BK, Lillehei KO: Primary intraosseous meningioma. Case report. *J Neurosurg* 83:912, 1995
14. Mattox A, Hughes B, Oleson J, et al: Treatment recommendations for primary extradural meningiomas. *Cancer* 117:24, 2010
15. Juratli TA, Geiger KD, Weigel P, et al: A five year-old child with clear cell petro-clival meningioma: Case report with clinical and histopathological long-term follow-up. *Childs Nerv Syst* 31:2193, 2015