Intracranial/Extracranial Meningioma Arising in the Hypoglossal Canal: Case Report

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ABSTRACT

A case of a patient with a posterior fossa meningioma extending through the hypoglossal canal to the cervical region as described in this article has not been previously described in the literature. Investigations and surgical management are outlined and pathological classifications are discussed. A literature review including recent reports of extracranial meningiomas is presented. Extracranial meningiomas are exceedingly rare and a high index of suspicion is necessary to make the diagnosis.

KEYWORDS: Extracranial meningioma, posterior fossa meningioma, carotid artery occlusion, thyroplasty, meningioma classification

Meningiomas are common tumors of the nervous system, representing 18% of primary intracranial tumors and 25% of primary intraspinal tumors.¹ Meningiomas appear to be more common in Africans and represent almost 30% of primary intracranial tumors.² Extracranial and extraspinal meningiomas are rare with an incidence of 1 to 2%.³ Extension of intracranial meningiomas to the parapharyngeal and cervical regions is exceedingly rare.⁴ The clinical diagnosis of cervical meningiomas can be difficult as more common cervical tumors such as metastatic epithelial tumors,

schwannomas, or paragangliomas are likely to be suspected first.⁵

We report on a meningioma with intracranial and extracranial components that presents with a cervical mass.

CASE REPORT

A 28-year-old African man presented with a 7-year history of a slowly enlarging right cervical mass and

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Figure 1 Contrast-enhanced (A) coronal and (B) axial T1-weighted magnetic resonance images. (A) The tumor is seen above and below the skull base (white arrows) extending through the hypoglossal canal (black arrows) into the neck. (B) The parapharyngeal component (white arrows) is seen encasing the cervical carotid artery (black arrow).

increasing dysarthria. Initial examination revealed a solitary 7- × 6.5-cm mass in the right cervical region extending to level 3. It was mobile, not attached to skin, and nonpulsatile. The oropharynx was displaced medially. Cranial nerve examination revealed a right hypoglossal nerve palsy with no deficits of the remaining cranial nerves. There was no abnormality in the nose or ears. An initial fine-needle aspiration (FNA) of the mass was inconclusive but an incisional biopsy suggested a diagnosis of meningioma.

Further investigations included computed tomography (CT) and magnetic resonance imaging (MRI) scans. These confirmed the presence of the large parapharyngeal mass and showed it to be continuous through a widened hypoglossal canal with a small mass in the right posterior fossa related to the posterior surface of the petrous bone, causing slight indentation of the brainstem. It had a

broad attachment to the petrous bone typical of meningioma (Fig. 1A). The internal carotid artery was encased in tumor and on angiography there was a fusiform filling defect of the artery (Figs. 1B and 2). The internal jugular vein did not fill.

The patient underwent tracheostomy in preparation for a test occlusion of his right carotid artery, which showed good filling of the right cerebral circulation from the contralateral carotid system and ipsilateral vertebral artery (Fig. 3). The right internal carotid artery was thus occluded in preparation for tumor resection 3 days later of the parapharyngeal component of the tumor.

At surgery the internal carotid artery, the internal jugular vein, and the lower cranial nerves (IX, X, XI, XII) could not be separated from tumor and were sacrificed. The major vessels were secured inferiorly and the cervical part of the tumor mobilized and completely removed up to the skull base.

Figure 2 An angiogram (lateral view) of a balloon-test occlusion of the right internal carotid artery. The position of the inflated balloon is indicated (arrows).

Figure 3 Anteroposterior view of a cerebral angiogram; the right internal carotid artery is occluded by intravascular coils (black arrows); collateral blood supply from the left internal carotid artery (white arrow) is seen filling the right anterior and middle cerebral arteries.

Postoperatively, the patient's neurological deficit was related to the loss of the lower cranial nerves. He initially required gastrostomy feeding. The tracheostomy tube and gastrostomy tube were removed following a medialization thyroplasty and intensive speech and swallow therapy. Two months after surgery he could swallow without difficulty and had a good voice. The intracranial component of the tumor is currently being managed conservatively and is monitored on regular MRI.

Histological examination of the mass confirmed a grade 1 meningioma (WHO grade 1, transitional subtype) (Table 1) which was locally invasive but had no malignant features.

DISCUSSION

Meningiomas account for 18% of intracranial tumors. They are usually benign but locally aggressive neoplasms that are thought to arise from the cap cells lining the arachnoid villae. The majority of meningiomas are supratentorial but about 8% arise in the posterior cranial fossa, and the neurotologist frequently encounters them in the cerebellopontine angle. Malignant features are seen in a small percentage of meningiomas, but there is controversy in neuropathological circles as to just what the criteria for malignancy should be.⁶ Cushing and Eisenhardt,⁷ in their classic 1938 monograph, were the first to propose a pathological classification of meningiomas. The current classification by the World Health Organization (WHO) 2000 is shown in Table 1. Grade 1 meningioma of the transitional subtype found in this case is one of the most common subtypes. The risk of aggressive growth and recurrence of this subtype is low if completely excised.

Extracranial meningiomas are not common. They occur in several ways⁸:

1. Primary intracranial meningioma with extracranial extension
2. From arachnoid cell rests, accompanying cranial nerve sheaths

Table 1 WHO 2000 Classification of Meningiomas

WHO Grade 1

Benign (90%), risk of recurrence and aggressive growth is low

Meningiothelial meningioma, fibrous (fibroblastic) meningioma, transitional (mixed) meningioma, psammomatous meningioma, angiomatous meningioma, microcystic meningioma, secretory meningioma, lymphoplasmacyte-rich meningioma, metaplastic meningioma

WHO Grade 2

Atypical (5–7%), recurrence and/or aggressive behavior possible

Chordoid meningioma, clear cell meningioma, atypical meningioma

WHO Grade 3

Anaplastic (3–5%), risk of recurrence and/or aggressive behavior more likely

Papillary meningioma, rhabdoid meningioma, anaplastic meningioma

3. From extracranial embryonic arachnoid cell rests
4. Metastases from intracranial meningiomas

These meningiomas may occur by direct extension of an intracranial meningioma through a natural, traumatic, or iatrogenic defect in the skull. They may arise from meningocytes in arachnoid clusters accompanying cranial nerve sheaths. They may occur without demonstrable connections with cranial nerves or intracranial primary tumors from presumed embryonic cell rests. These are primary ectopic meningiomas. Rarely, an extracranial meningioma may be metastatic from an intracranial primary.^{9,10}

The most common site of occurrence of an extracranial meningioma is the skin, followed by the orbit; the nose and sinuses may be involved either from direct extension from an anterior fossa tumor or de novo. Primary ectopic tumors are very rare with very few reported cases. Kishore and associates¹¹ described a case of primary meningioma of the soft palate. Petrulionis and colleagues¹² record the case of a child with a primary extracranial meningioma in the nose. Daneshi et al¹³ document a primary meningioma of the ethmoid sinus. Ex-

tracranial meningiomas may also occur in the ear, either as extension into the middle or external ear of an intrapetrous meningioma^{14–16} or de novo in the external meatus without obvious intracranial connection.^{15,17}

Extracranial meningiomas may also occur in the neck, as with the case we describe. Direct extension from a posterior or middle fossa mass is the most common cause and has been described in the infratemporal fossa,¹⁸ parapharyngeal space,¹⁹ the parotid gland,²⁰ and even inside the lumen of the internal jugular vein.²¹

Extension into the neck of a posterior fossa meningioma is most likely to occur through a skull base foramen, and of these the jugular foramen in the most common route. Shimono et al²² described eight cases of jugular foramen meningiomas and noted that the intracranial and extracranial components displayed different signal intensity and enhancement patterns on MRI, suggesting differences in histological composition, notably collagen.

The case described here had both intracranial and extracranial components connected to each other through a widened hypoglossal canal. It would appear that this tumor took origin from primitive meningocytes on the hypoglossal nerve as it passed through the skull base. To our knowledge, this pattern of growth in relation to the hypoglossal nerve has not been previously reported in the literature. The intracranial portion of the tumor was considerably smaller than the cervical portion. This may be due to the availability of more space for tumor expansion in the neck, or better blood supply to the tumor within the neck. It is possible but unlikely that the tumor had an extracranial origin with secondary extension through the hypoglossal foramen. The observation by Shimono et al²² that intracranial and extracranial components could have different histological composition might lead one to suggest that the two parts could have different growth characteristics.

Solares and Lacruz⁵ suggested that FNA might be a suitable technique to diagnose extracranial

meningiomas,²² but it did not yield a diagnosis in this case, making incisional biopsy necessary. Cytological features of meningiomas include a syncytial cellular pattern, laminated whorls, psammoma bodies, as well as pseudoinclusions. Psammoma bodies are also seen in papillary thyroid carcinoma, which must therefore be excluded carefully when investigating a cervical mass using FNA.

Not all meningiomas need to be removed. The extracranial mass was, however, so large with distortion of the pharynx that surgery was recommended despite the problems that involvement of the lower cranial nerves and the internal carotid artery could have presented. Balloon occlusion suggested that sacrifice of the carotid artery would be safe, and there were no complications as a result of this. The deficit from loss of the lower cranial nerves can be devastating, especially in older patients, because of swallowing difficulties and aspiration. This young man made a rapid adjustment and after 2 months was swallowing normally. A medialization thyroplasty performed soon after the original operation gave him a good voice. Many intracranial meningiomas run a fairly indolent course. For that reason and based on our previous experience of managing posterior fossa meningiomas, it was decided not to excise the intracranial portion of the tumor at this time but to look for evidence of further growth on future MRI. Furthermore, the difference in size of the intracranial and extracranial components of the tumor might suggest different growth rates of the two parts. Stereotactic radiotherapy is rarely employed in our institution for the management of meningiomas, and was not considered in this case.

CONCLUSION

Intracranial meningiomas with extracranial extension are exceedingly rare. The tumor described appears to have arisen in the hypoglossal canal, which appears not to have been described before.

The intracranial and extracranial components may show different growth patterns. Diagnosis may be suggested on CT and MRI and confirmed by FNA of the cervical component.

REFERENCES

1. Rubinstein LJ. Tumors of the central nervous system. Fascicle 6. Washington, DC: Armed Forces Institute of Pathology; 1972
2. Ohaegbulam SC. Geographical neurosurgery. *Neurol Res* 1999;21:161–170
3. Whicker JH, Devine KD, MacCarty CS. Diagnostic and therapeutic problems in extracranial meningiomas. *Am J Surg* 1973;126:452–457
4. Batsakis JG. Extracranial meningiomas. *Ann Otol Rhinol Laryngol* 1984;93:282–283
5. Solares J, Lacruz C. Fine needle aspiration cytology diagnosis of an extracranial meningioma presenting as a cervical mass. *Acta Cytol* 1987;31:502–504
6. Perry A, Scheithauer BW, Stafford SL, Lohse CM, Wollan PC. "Malignancy" in meningiomas: a clinicopathological study of 116 patients, with grading implications. *Cancer* 1999;85:2046–2056
7. Cushing HW, Eisenhardt L. Meningiomas: Their Classification, Regional Behaviour, Life History and Surgical End Results. Springfield, IL: Charles C. Thomas; 1938
8. Hoye SJ, Hoar CS, Murray JE. Extracranial meningioma presenting as a tumor of the neck. *Am J Surg* 1960;100:486–489
9. Teague SD, Conces DJ Jr. Metastatic meningioma to the lungs. *J Thorac Imaging* 2005;20:58–60
10. Younis G, Sawaya R. Intracranial osteolytic malignant meningiomas appearing as extracranial soft-tissue masses. *Neurosurgery* 1992;30:932–934
11. Kishore A, Roy D, Irvine BW. Primary extracranial meningioma of the soft palate. *J Laryngol Otol* 2000;114:149–150
12. Petrulionis M, Valeviciene N, Paulauskiene I, Bruzaite J. Primary extracranial meningioma of the sinonasal tract. *Acta Radiol* 2005;46:415–418
13. Daneshi A, Asghari A, Bahramy E. Primary meningioma of the ethmoid sinus: a case report. *Ear Nose Throat J* 2003;82:310–311
14. Tsunoda R, Fukaya T. Extracranial meningioma presenting as a tumour of the external auditory meatus: a case report. *J Laryngol Otol* 1997;111:148–151
15. Thompson LD, Bouffard JP, Sandberg GD, Mena H. Primary ear and temporal bone meningiomas: a clinicopathologic study of 36 cases with a review of the literature. *Mod Pathol* 2003;16:236–245
16. Hanna SJ, Derham C, Van Hille P, Fenwick JD. Cerebellopontine angle meningioma resulting in middle-ear polyp. *J Laryngol Otol* 2006;120:786–788

17. Uppal HS, Kabbani M, Reddy V, Kaur S. Ectopic extracranial meningioma presenting as an aural polyp. *Eur Arch Otorhinolaryngol* 2003;260:322–324
18. Pieper DR, Al-Mefty O. Management of intracranial meningiomas secondarily involving the infratemporal fossa: radiographic characteristics, pattern of tumor invasion, and surgical implications. *Neurosurgery* 1999;45:231–237
19. Shuangshoti S, Netsky MG, Fitz-Hugh GS. Parapharyngeal meningioma with special reference to cell of origin. *Ann Otol Rhinol Laryngol* 1971;80:464–473
20. Walther LE, Mentzel R, Meyer R, Eschholz A, Beleites E. Extracranial meningioma of the parotid gland [in German]. *HNO* 1999;47:573–576
21. Miyagi N, Morimitsu H, Fujimura N, et al. Case of meningioma with extracranial extension through the internal jugular vein [in Japanese]. *No Shinkei Geka* 2005;33:271–275
22. Shimono T, Akai F, Yamamoto A, et al. Different signal intensities between intra- and extracranial components in jugular foramen meningioma: an enigma. *AJNR Am J Neuroradiol* 2005;26:1122–1127