Case Report

Recurrent paraganglioma of Meckel’s cave: Case report and a review of anatomic origin of paragangliomas

Anna Prajsnar, Naci Balak1, Gerhard F. Walter2, Alexandru C. Stan2, Wolfgang Deinsberger, Leyla Tapul3, Cicek Bayindir4

Department of Neurosurgery, Klinikum Kassel, Kassel, Germany, 1Goztepe Education and Research Hospital, Istanbul, Turkey, 2Department of Pathology, Klinikum Kassel, Kassel, Germany, 3Department of Histology, Istanbul Faculty of Medicine, 4Department of Neuropathology, Istanbul Faculty of Medicine, University of Istanbul, Turkey

E-mail: Anna Prajsnar - aniaprajsnar@poczta.onet.pl; *Naci Balak - drnacibalak@yahoo.com; Gerhard F. Walter - gf.walter@klinikum-kassel.de; Alexandru C. Stan - alex.stan@gmx.net; Wolfgang Deinsberger - wolfgang.deinsberger@klinikum-kassel.de; Leyla Tapul - tapul@istanbul.edu.tr; Cicek Bayindir - cicekbayindir@gmail.com

*Corresponding author

Received: 2 February 11
Accepted: 22 March 11
Published: 19 April 11

This article may be cited as:
Prajsnar A, Balak N, Walter GF, Stan AC, Deinsberger W, Tapul L, Bayindir C. Recurrent paraganglioma of Meckel’s cave: Case report and a review of anatomic origin of paragangliomas. Surg Neurol Int 2011;2:45.
Available FREE in open access from: http://www.surgicalneurologyint.com/text.asp?2011/2/1/45/79763

Copyright: © 2011 Prajsnar A. This is an open-access article distributed under the terms of the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original author and source are credited.

Abstract

Background: Paragangliomas are rare, usually benign tumors of neural crest origin. They account for only 0.6% of all head and neck tumors. In the craniocervical area, they are more common in the carotid body and tympanic-jugular regions. To the authors’ knowledge, a case of paraganglioma in Meckel’s cave has not yet been reported in the medical literature. The pathogenesis and natural history of paragangliomas are still not well understood. We present a case of recurrent paraganglioma in Meckel's cave.

Case Description: A 53-year-old woman was diagnosed with trigeminal neuralgia, dysesthesia and hypoesthesia on the left side of the face, hearing disturbance and a history of chronic, persistent temporal headaches. Magnetic resonance imaging (MRI) showed a lesion located in Meckel’s cave on the left side, extending to the posterior cranial fossa and compressing the left cerebral peduncle. The lesion was first thought to be a recurrence of an atypical meningioma, as the pathologist described it in the tissue specimen resected 3 years earlier, and a decision for re-operation was made. A lateral suboccipital approach to the lesion was used under neuronavigational guidance. The tumor was removed, and histological examination proved the lesion to be a paraganglioma. Five months later, the follow-up MRI showed local regrowth, which required subsequent surgical intervention.

Conclusions: A paraganglioma in Meckel’s cave is an uncommon tumor in this location. Although ectopic paragangliomas have been described in the literature, a paraganglioma atypically located in Meckel’s cave makes a topographic correlation difficult, mainly because paraganglionic cells are usually not found in Meckel’s cave. Another peculiarity of the case is the local recurrence of the tumor in a relatively short time despite an attempted, almost gross total resection.

Key Words: Intracranial tumors, Meckel’s cave, neurocristopathies, paraganglioma, transmission electron microscopy
INTRODUCTION

The vast majority of paragangliomas present as spinal intradural tumors in the cauda equina region. Paragangliomas or glomus tumors are rare tumors of the head and neck. They usually occur in the fifth and sixth decade of life. They may be sporadic or familial, and solitary or multicentric, although they most commonly appear solitarily.\cite{11,13,41,74} They account for only 0.6% of all head and neck tumors.\cite{73} They are more common in Caucasians\cite{21} and occur more frequently in women.\cite{13,21,41} Paraganglia are clusters of neuroendocrine cells dispersed throughout the body, and connected with the autonomic nervous system.\cite{11} The largest collection of these cells is found in the adrenal medulla, where they give rise to pheochromocytomas.\cite{11} Extra-adrenal paragangliomas develop in two general locations: paravertebral paraganglia, close to the spinal cord, and paraganglia close to, occasionally touching, the large vessels of the head and neck. The former distinction into chromaffin and non-chromaffin paragangliomas has been abandoned since the reaction of tumor cells with chromic acid does not reliably reflect their functional activity, i.e., catecholamine secretion. The fact that the tumor cells arise from paraganglionic cells explains, and also indicates, their site and origin.\cite{2,30,35,41,49,57,64,67,68,74} More than 90% of paragangliomas in the cranial cavity occur in the jugular foramen.\cite{73}

Magnetic resonance imaging (MRI) is the procedure of choice. Radiologically, the lesion usually appears as a well-limited, circumscribed mass. It occasionally contains cystic compartments. After administration of contrast media, the lesion enhances markedly. The low-intensity rim, the so-called “cap sign”, seen well in T2-weighted images, which reflects the architecture of serpentine-like, elastic vessels around the Zellballen areas, is considered diagnostically helpful. Nevertheless, paragangliomas may be confused with meningiomas and schwannomas, but they can be distinguished from these by examining pre-operative radiographic imaging meticulously.\cite{9,13} We present a case of paraganglioma in Meckel’s cave that is atypical with regard to its location as well as to its multiple local recurrences.

CASE REPORT

A 53-year-old woman was admitted to the clinic with a 2-month history of trigeminal neuralgia, dysesthesia and hypoesthesia in the left side of the face, combined with a lengthy history of persistent temporal headaches and a progressive hearing problem. An MRI revealed a tumor in the left Meckel’s cave, with partial compression of the trigeminal nerve and extension to the posterior cranial fossa [Figure 1a]. A temporobasal craniotomy was performed and the lesion was completely removed. The histological pattern of the specimen was classified initially as an atypical meningioma (WHO grade II) in 2005. Two years later, in 2007, a follow-up MRI showed a contrast-enhanced lesion, a seemingly recurrent meningioma mass in Meckel’s cave. A local stereotactic radiotherapy (10 × 4 Gy) was applied. However, the patient was re-admitted to the hospital 1 year later, in 2008, because of progressive clinical disturbance and increased volume of the lesion seen in follow-up MRI scans [Figure 1b], and a decision for re-operation was made. A left lateral suboccipital approach was used under neuronavigational guidance. The tumor was almost completely removed. The histopathologic examination of the specimen [Figure 2a–f] showed fragments of well-organized, compacted and nested epitheloid cells of the tumor dispersed in lobules (Zellballen architecture, composed

Figure 1: (a) The first manifestation of the tumor. The axial and coronal T1-weighted, contrast-enhanced MR scan shows a strongly enhanced lesion in the Meckel’s cave. The large tumor is compressing the preoptic cistern, the pontocerebellar cistern and the pons. A clear border among the tumor, trigeminal nerve and the internal cerebral artery is indistinguishable. (b) Pre-operative axial T1-weighted MRI with contrast media revealing the first reoccurrence of the tumor in the left Meckel’s cave. The volume of the tumor in the medial fossa is approximately 2.5 × 1.8 cm. Regressive changes and cystic compartment with liquid mirror image are visible within the lesion. Part of the tumor is located in the posterior fossa; its volume is 1.3 × 1.6 cm. The left trigeminal nerve inside the tumor is no longer distinguishable. Additionally, a compression of the pons as well as the left cerebral peduncle in the ponto-mesencephalic junction can be seen. The part of the tumor in the middle fossa has contact with the C2–C5 segments of the left internal carotid artery
of nests of polygonal chief cells enclosed by trabeculae of fibrous and sustentacular elongated cells) which were histologically and immunohistochemically classified as a paraganglioma (WHO grade I). A comparative histopathologic reinvestigation of the tumor specimen surgically removed during the first operation gave the result that the first tumor was a paraganglioma too, not an atypical meningioma. Five months after the second surgical intervention, in 2009, another local recurrence of the tumor was diagnosed in a follow-up MRI. Consequently, the tumor was removed and again classified as a paraganglioma (WHO grade I).

**Immunohistochemistry**

In addition to the examination of hematoxylin and eosin-stained tumor tissue [Figure 2a and b], an immunohistochemistry investigation was performed. Several markers, including neuron-specific enolase (NSE), synaptophysin, neurofilament (NF) proteins, chromogranin A, Comori reticulin stain, and S-100 protein, were used to identify immunohistochemical features of the tumor. NSE immunoreactivity was strongly positive [Figure 2c]. Immunostaining for NF was also positive [Figure 2d]. These reactions, as well as a positive reaction for synaptophysin, permitted identification of the chief cells. Comori reticulin stain showed septae delineating Zellballen [Figure 2e]. Chief cells were not immunoreactive for chromogranin A, and sustentacular cells were not immunoreactive for S-100 protein. All of the primary and recurrent tumors had similar histopathologic features.

The proliferative activity, given as the percentage of Ki-67/MIB-1 immunoreactive cells, was calculated using a 10 × 10 square ocular grid. The tumor cells showed a mean of 20% proliferative activity, as measured by Ki-67/MIB-1 staining [Figure 2f]. Ki-67/MIB-1 expression levels were similar in all primary and recurrent tumor specimens.

**Electron microscopy studies**

It was decided to confirm further the diagnosis of paraganglioma using an electron microscopic (EM) examination. For this purpose, a transmission electron microscopy study was carried out in specific areas of the formalin-fixed, paraffin-embedded blocks, using a modified protocol of previously described various methods.[12,36,61] The areas that were rich in chief cells as identified by immunohistochemistry and light microscopy were marked on the surface of the paraffin-embedded blocks. In these marked areas, tissue fragments of 1 mm³ were cut with a sharp razor blade. Samples of 1 mm³ that were cut out were deparaffinized using toluene solution at room temperature for 90 minutes. The specimens were transferred to an absolute alcohol solution, and after a reprocessing protocol, the re-embedded epon blocks of the tissue pieces were sectioned and ultrathin sections of 400–600 Å were obtained using an ultramicrotome (LKB Ultramicrotome, Stockholm, Sweden). Thick sections were stained with toluidine blue and examined. The
ultrathin sections were contrasted with uranyl acetate and lead citrate. Finally, these ultrathin sections were analyzed and documented photographically using a transmission electron microscope (JEOL 100 C, Tokyo, Japan). The ultrathin sections permitted the identification of neurosecretory granules, which are the distinctive ultrastructural feature of chief cells and confirmed the diagnosis of paraganglioma in this case [Figure 4].

**DISCUSSION**

The tumors encountered most frequently within Meckel’s cave are adenoid cystic carcinoma, meningioma, and schwannoma.[25] Only a few cases of supratentorial paraganglioma have been reported in the international literature. In one of the largest investigated series, 71 patients with paragangliomas of the craniocervical region, there was a notable trend of local recurrence.[32] These rare tumors have been found mainly in the middle cranial fossa – cavernous sinus, sphenoid ridge, and also, some in the parasellar region.[2,30,56,57,69] The presence of a paraganglioma in Meckel’s cave is likely an indication of its ectopic origin.

It is known that paraganglionic cells are derived from embryonic cells of the neural crest.[34,62] The neural crest is the major source of mesenchymal cells in the head and neck, and in addition, gives rise to sensory ganglia, autonomic and enteric ganglia, as well as to pigment cells of the skin.[62] Endocrine tissues, including adrenomedullary cells and adrenergic paraganglia, also calcitonin parafollicular cells of the thyroid gland and carotid body, are derived from the neural crest.[34,62] Leptomeninges and Schwann’s sheath cells, which give rise to meningiomas and schwannomas, respectively, are also derivatives of the neural crest.[62] Despite the morphological differences between these tumors and paragangliomas, the possibility of their coexistence can be connected to the same embryonic origin. A common embryonic origin of two morphologically completely different tumors, but having the same embryonic background, can also be shown in cases describing the coexistence of a meningioma with a pheochromocytoma.[34] These entities have been defined as “neurocristopathies”, originally defined in 1974 as a category of diseases arising from neural crest development.[5,6] On the other hand, Tong et al. used the term paraganglioma-like meningioma and reported four cases in ultrastructural and immunohistochemical studies, exhibiting the possible morphological similarity of meningioma and paraganglioma in single cases.[64]

Ectopic locations of paraganglioma have been reported. For example, Nielsen et al. described a case of paraganglioma in the tongue.[43] Another unusual

---

**Figure 3:** Pre-operative axial T1-weighted MRI with contrast media, axial T2-weighted MRI, and coronal T1-weighted contrast-enhanced MRI show a markedly enhanced lesion located at the site of the previously removed tumor in Meckel’s cave.

**Figure 4:** Transmission electron microscopy shows identification of neurosecretory granules (arrow) in chief cells. Higher magnification in the inset.
location of paraganglioma was found by Yamuchi et al., who described a paraganglioma in the frontal skull base connected to the sphenoid ridge. As an explanation, it was suggested that paraganglionic cells strayed to the sphenoid ridge following the endothelium of the middle meningeal artery and external carotid artery whose rich innervations derive from the superior cervical ganglion. From an anatomical point of view, the tumor in our patient may have originated from parasym pathetic paraganglionic cells along the lesser petrosal nerve, or from sympathetic paraganglionic cells in the walls of arteries, or in the external and deep petrosal nerves. Anatomically, the lesser petrosal nerve is composed of fibers from three cranial nerves: the tympanic branch of the glossopharyngeal nerve, the nervus intermedius of the facial nerve, and the auricular branch of the vagus nerve.[23] The sympathetic nerve fibers, originating in the superior cervical ganglion, travel along the external carotid artery, follow the maxillary and middle meningeal arteries and pass through the foramen spinosum, which is close to Meckel’s cave. The external and deep petrosal nerves are simply intracranial extensions of the cervical sympathetic trunk. Ultimately, so-called ectopic paraganglioma is an unusual pathological entity even though there may be considerations as to why paraganglioma may appear in Meckel’s cave, and the term “ectopic location” is still not accepted by all.[19,35,75]

Conventional methods for EM examination include immediate fixation of small pieces of tumor tissue in 2.5% cacodylate buffered glutaraldehyde solution, post-fixation in 1% osmic acid for an hour, dehydration through a graded series of alcohol and embedding it in Epon 812.[50] As in the present case this procedure was not initially feasible, the formalin-fixed, paraffin-embedded tissue was used for EM. There are various techniques for re-embedding of formaldehyde-fixed paraffin-embedded material for EM investigation.[12,39,50] Formaldehyde is not as useful as glutaraldehyde for ultrastructural investigation since it leads to lipid extraction during re-embedding, and subsequently, structural damages and weak contrast. Mitochondria, polyribosomes, and microtubules are among the organelles that exhibit various degrees of degradation in re-processed paraffin-embedded tissue. However, accurate sampling of highly specific tissue areas from large complex lesions can reliably be performed using archival paraffin-embedded material.[12] In our case, the EM quality approached that of material immediately processed for EM. Hence, it was possible to recognize clearly neurosecretory granules of the chief cells.

Current trends in the management of paragangliomas

In the last decades, various treatment concepts for paragangliomas of the head and neck region have been introduced and implemented. They include observation, microsurgery with or without selective endovascular embolization, conventional radiotherapy, and stereotactic radiosurgery (SRS) using tools such as the Leksell Gamma Knife or linear accelerator (Linac)-based CyberKnife.[4,10,24,54,55] In order to achieve better quality of microsurgical tumor resection, and consequently prolonged survival, these procedures have also been used in combination with staged stereotactic radiosurgery following tailored surgery.[16,72] Microsurgical removal is the generally accepted procedure of choice for the treatment of patients with non-infiltrative paragangliomas, in the same way as for other truly benign brain tumors. The beneficial effects of microsurgical management seem to be related to normal life spans and avoiding the possible risks of long-term adverse effects of irradiation (>15 years), including tumor regrowth and radiation-induced neoplasms, although these issues remain in debate in the medical literature.[29,33,48] However, the morbidity and mortality ratio of surgery may be high because of close proximity of the tumor to cranial nerves and vascular structures. Radical resection is almost impossible without sacrifice of one or more cranial nerves.[41,48] Additionally, the vascular nature of paraganglioma itself may make the surgery complicated. Multicentric paragangliomas represent another difficulty in the treatment of paragangliomas.[131] Due to the high risk associated with surgery, some authors recommend radiation therapy as the initial treatment for paragangliomas.[48] Many reports appeared recently in the medical literature regarding promising results shown with the use of irradiation/stereotactic radiosurgery in the treatment of neck and some head paragangliomas, with a mean/median of 4–13 years of follow-up including large case series.[6,45,47,48,49] These results have been found to be equally positive as, or even better than, surgery. In these studies, irradiation/stereotactic radiosurgery has been used as the primary treatment, after surgery for residual or recurrent tumors or following embolization. Radiosurgery was also found to be effective in reducing the catecholamine secreting capacity of paraganglioma. However, there is some controversy about the use of radiation for treatment of paragangliomas because of the presence of persistent chief cells years after treatment.[6,32] The volume of tumors usually ranges from 0.38 to 33.5 cm3 in patients treated using SRS.[14,15,54] Although SRS has been used as primary management modality because it is an effective alternative to surgery, there are some patient selection criteria for SRS that include size and extension of tumor, presence of residual or recurrent tumor after surgery, age and pre-existing medical conditions of the patient, endocrinological activity as well as neurological manifestation.[31] The outcomes of SRS have been evaluated using one or more of three criteria: Subjective, neurological and radiological improvements. A comparison of the biological behavior of our case, which was proved histopathologically to be a paraganglioma,
with those of previous studies is difficult, mainly for two
reasons: a lack of histopathologic diagnosis in some cases
which were primarily treated using SRS and the rarity of
paragangliomas in the regions of sella turcica, cavernous
sinus and Meckel’s cave. However, it should be borne
in mind that a paraganglioma in the region of Meckel’s
cave might not be completely benign, as our case showed
multiple recurrences even after gross total or near total
resection and despite the use of SRS.

Prognostic factors
Paragangliomas are listed under the heading “Neuronal
and mixed neuronal-glial tumors” according to the
2007 WHO classification of tumors of the central
nervous system.[38] Paragangliomas have remarkably
uniform microscopic features; they are composed of
nests of polygonal chief cells enclosed by trabecula of
fibrous and sustentacular elongated cells.[11] There is
little pleomorphism and mitoses are relatively rarely
seen. The cells are agyrophilic and stain positively for
neuroendocrine markers.

Although paragangliomas are usually benign tumors
classified as WHO grade I tumors, they can display
features of malignancy and manifest aggressive behavior,
such as the possibility of metastasizing, which is rarely
observed.[36,66] Malignant paraganglioma of the sellar
region may also mimic a pituitary macroadenoma.[36] In
contrast to benign tumors, malignant paragangliomas
tend to show focal necrosis areas as well as vascular
invasion. Mitotic figures, which are atypical and occur very
seldom in the benign cases, are usually seen in malignant
paragangliomas.[21] On the contrary, some authors stated
that foci of hemorrhagic necrosis and scattered mitotic
figures as well as nuclear pleomorphism are not of
prognostic significance.[38] On the other hand, it has been
reported that anaplastic and metastasizing extracranial
paragangliomas are either devoid or markedly depleted of
sustentacular cells.[27,28,63] Although sustentacular cells are
usually uniformly reactive for S-100 protein, sustentacular
cells in our case did not show immunoreactivity for
S-100. This could be an important finding that might
explain the recurrences and aggressive behavior of the
tumor in this case and be relevant to the previous
medical literature. It should also be kept in mind that
immunologic profile seems to vary slightly, depending
on the type of paraganglioma.[71] Hence, histological
criteria alone are not enough to predict the biological
behavior of paragangliomas.[34] Tumor location has been
reported to be important in determining recurrences and
metastasis of paragangliomas.[28] Meckel’s cave may be
one such location of recurrent paragangliomas. A
Correlation of MIB-1 labeling and malignant behavior has
been previously demonstrated in pheochromocytoma,[7]
whereas there is no established relationship between
indices and recurrence in intracranial paragangliomas.
In our case, the significance of the, however elevated, Ki-67/
MIB-1 labeling index remains open, since the prognostic
importance of Ki-67/MIB-1 labeling index in intracranial
paragangliomas has not been well described so far.

Genetic factors have a role in the pathogenesis of
paragangliomas.[1,3,4,46,59,60] Recent studies have
verified that succinate dehydrogenase (SDH) gene
mutations in germ line occur in at least 11% of
non-familial head and neck paragangliomas, 8% of
non-familial pheochromocytomas, 28% of malignant
pheochromocytomas and 33% of extra-adrenal
pheochromocytomas.[3] Recent studies have shown that
at least one-third of paragangliomas are familial ones,
which are often indistinguishable from truly sporadic
paragangliomas.[15] However, the pathogenesis of a
substantial percentage of truly sporadic paragangliomas is
still unknown. We were unable to study the chromosomal
changes in our patient. Nevertheless, our case was
probably a sporadic paraganglioma, since there were
no clinical indicators of familial paraganglioma, such as
multiple, bilateral paragangliomas, coexistence of
paragangliomas and pheochromocytomas, and young age
of onset.

CONCLUSIONS

Although ectopic paragangliomas have been described
in the literature, a paraganglioma atypically located in
Meckel’s cave makes a topographic correlation difficult,
mainly because paranganglionic cells are usually not
found in Meckel’s cave. In the present case, there
were no elements allowing for a distinct diagnosis of a
paranganglioma-like meningioma as described by Tong
et al.[64] Because SDH gene evaluation was not carried out
in our case, additional genes and the role of SDH genes
should be studied, especially those that are also involved
in the pathogenesis of recurrent paragangliomas.

REFERENCES

1. Antonello M, Piazza M, Menegolo M, Opocher G, Deriu GP, Grego F. Role of
the genetic study in the management of carotid body tumor in paraganglioma
syndrome. Eur J Vasc Endovasc Surg 2008;36:517-9.
2. Arkiha Y, Boutarbour M, Lamalini N, Halefdi S, Laamarti A, Derraz S, et al. Paraganglioma of the latero-sellar area. Case report. Neurochirurgie
2003;49:540-4.
3. Baysal BE. Clinical and molecular progress in hereditary paraganglioma. J
Med Genet 2008;45:689-94.
4. Bianchi LC, Marchetti M, Brat L, Bergantin A, Milanesi I, Broggi G, et al.
Paragangliomas of head and neck: A treatment option with CyberKnife
radiosurgery. Neurol Sci 2009;30:479-85.
5. Bolande RP. Neuroradiosurgery: Its growth and development in 20 years. Pediatr
Pathol Lab Med 1997;17:1-25.
6. Bolande RP. The neuroradiopathies: A unifying concept of disease arising in
neural crest maldevelopment. Hum Pathol 1974;5:409-29.
7. Brown HM, Komorowski RA, Wilson SD, Demereu MJ, Zhu YR. Predicting
metastasis of pheochromocytomas using DNA flow cytometry and
immunohistochemical markers of cell proliferation: A positive correlation
between MIB-1 staining and malignant tumor behavior. Cancer 1999;86:
1583-9.
8. Castrucci WA, Chiang VL, Hulinsky I, Kniely JP. Biochemical and clinical responses after treatment of a catecholamine-secreting glomus jugulare tumor with gamma knife radiosurgery. Head Neck 2010;32:1720-7.

9. Chen ZC, Wang CP, Hsiao JK, Ko JT, Tseng HM, Yso YT. Angiomatous type of jugular foramen meningioma with neck extension: Differential diagnosis from paraganglioma and schwannoma. Head Neck 2007;29:793-8.

10. Chung SM, Kim HS, Jung J, Lee HK. Lee WS. Clinical presentation and management of jugular paraganglioma. Clin Exp Otorhinolaryngol 2009;2:28-32.

11. Cotran RS, Kumar V, Collins T. Pathologic Basis of Disease. 6th ed. Philadelphia, Pennsylvania: W.B. Saunders Company; 1999. p. 768-69.

12. Dingemans KP, Rankema M. Immunoelectron microscopy on material retrieved from paraffin. Accurate sampling on the basis of stained paraffin sections. Ultrastruct Pathol 2001;25:201-4.

13. Erkmen K, Al-Mefty O, Adada B. Tumors of the skull base. In: Tonon J, Westphal M, J. R. Grossman SA, editors. Neuro-Oncology of CNS Tumors. Berlin Heidelberg: Springer-Verlag; 2006. p. 242-68.

14. Eustacchio S, Trummer M, Unger F, Schrottner O, Sutter B, Bendí G. The role of Gamma Knife radiosurgery in the management of glomus jugulare tumors. Acta Neurochir Suppl 2002:84:91-7.

15. Feigl GC, Horstmann GA. Intracranial glomus jugulare tumors: Volume reduction with Gamma Knife surgery. J Neurosurg 2006;105:161-7.

16. Foote RL, Pollock BE, Gorman DA, Schomberg PJ, Stafford SL, Link MJ, et al. Glomus jugulare tumor: Tumor control and complications after stereotactic radiosurgery. Head Neck 2002;24:332-8; discussion 338-9.

17. Genc A, Bicer A, Abacioglu U, Peker S, Panır MK, Kılıç T. Gamma knife radiosurgery for the treatment of glomus jugulare tumors. J Neurooncol 2010;97:101-8.

18. Gottfried ON, Liu JK, Coulwdell WT. Comparison of radiosurgery and conventional surgery for the treatment of glomus jugulare tumors. Neurorsurg Focus 2004;17:64.

19. Helfner DK. Sphenoid sinus paraganglioma. Am J Otolary 2009;30:70. author reply 71.

20. Hinerman RW, Andrus BJ, Morris CG, Kirwan J, Mendenhall WM. Definitive radiotherapy in the management of paragangliomas arising in the head and neck: A 15-year experience. Head Neck 2008;30:1431-8.

21. Jackson CG. Glomus tympanicum and glomus jugulare tumors. Otolaryngol Clin North Am 2001;34:941-70, vii.

22. Jordan JA, Roland PS, McManus C, Weiner RL, Giller CA. Stereotactic radiosurgery for glomus jugulare tumors. Laryngoscope 2000;110:35-8.

23. Kakizawa Y, Abe H, Fukushima Y, Hong K, Kihouly H, Rhoton AL Jr. The course of the lesser petrosal nerve on the middle cranial fossa. Neurorsurgery 2007;61:15-23; discussion 23.

24. Karaman E, Yilmaz M, Isildak H, Korkut N, Devranoglu I, Karaman C. Jugular foramen meningioma with neck extension: Differential diagnosis from recurrent meningioma and metastatic meningioma in the base of the skull. J Neuro-Oncol 2007;28:347-9.

25. Liess BD, Horst JA, Zitsch RP. Sphenoid sinus paraganglioma: first reported case. Am J Otolaryngol 2007;28:347-9.

26. Lighezan R, Baderca F, Alexe A, Iacovlev M, Bonte D, Murarescu ED, et al. The value of the reprocessing method of paraffin-embedded biopsies for transmission electron microscopy. Rom J Morphol Embryol 2009;50:613-7.

27. Lim M, Gibbs IC, Adler JR, Chang SD. Efficacy and safety of stereotactic radiosurgery for glomus jugulare tumors. Neurosurg Focus 2004;17:E11.

28. Louis DN, Oghalii H, Wester OD, Cavenee WK, Burger PC, Jouvet A, et al. The 2007 WHO classification of tumours of the central nervous system. Acta Neurochir 2007;149:1079-109.

29. Mazrouf M, Vokes J, Landwehr P, Bramer R, Treuer H, Kocher M, et al. Stereotactic linear accelerator-based radiosurgery for the treatment of patients with glomus jugulare tumors. Cancer 2003;97:1093-8.

30. Miller JP, Semaan M, Einstein D, Megerian CA, Maciunas R. Staged Gamma Knife radiosurgery after tailored surgical resection: A novel treatment paradigm for glomus jugulare tumors. Stereotact Funct Neurosurg 2009;87:3-16.

31. Moe KS, Li D, Linder TE, Schmid S, Fisch U. An update on the surgical treatment of temporal bone paraganglioma. Skull Base Surg 1999;9:185-94.

32. Neumann HP, Ercoli Z, Boedecker CC, Rybicki LA, Robledo M, Hermsen M, et al. Clinical predictors for germline mutations in head and neck paraganglioma patients: Cost reduction strategy in genetic diagnostic process as fall-out. Cancer Res 2009;69:3650-6.

33. Nielsen TO, Sejean G, Onerheim RM. Paraganglioma of the tongue. Arch Pathol Lab Med 2000;124:877-9.

34. Perren A, Kominioth P. Familial pheochromocytomas and paragangliomas: Stories from the sign-out room. Endocr Pathol 2006;17:337-44.

35. Persky MS, Setton A, Nini Y, Hartman J, Frank D, Berenstein A. Combined endovascular and surgical treatment of head and neck paragangliomas—a team approach. Head Neck 2002;24:423-31.

36. Persu A, Hanoir M, Greigore V, Garian P, Duvvier E, Reyehler H, et al. High prevalence of SDHB mutations in head and neck paragangioma in Belgium. J Hypertens 2008;26:1395-401.

37. Pollock BE. Stereotactic radiosurgery in patients with glomus jugulare tumors. Neurosurg Focus 2004;17:E10.

38. Ramina R, Maniglia JF, Fernandes Y, Paschohal JR, Neto MC. Jugal Foramen Tumors – Diagnosis and Management. In: Ramina R, Aguilar PH, Tatagiba M, editors. Sami’s Essentials in Neurosurgery. Berlin Heidelberg New York: Springer; 2008.

39. Reithmeier T, Gumprecht H, Stolzle A, Lumenta CB. Intracerebral paraganglioma. Acta Neurochir (Wien) 2000;142:1063-6.

40. Robinson G, Gray T. Electron microscopy 2: Practical procedures. In: Bancroft JD, Stevens A, editors. Theory and practice of histological techniques. London, New York, Madrid, san Francisco: Churchill Livingstone; 1996. p. 856-262.

41. Rzepekowska A, Osuch-Wojcikiewicz E, Kulesza A, Badejiewicz A, Niemczyk K. Paragangliomas of the neck: A 10-year experience of the Department of Otolaryngology of Warsaw Medical University. Otolaryngol Pol 2010;64:65-72.

42. Schwaber MK, Gussack GS, Kirkpatrick W. The role of radiation therapy in the management of catecholamine-secreting glomus tumors. Otolaryngol Head Neck Surg 1988;98:150-4.

43. Sevilla MA, Hermsen MA, Weiss MM, Grimbergen A, Balbin M, Lorrente JL, et al. Chromosomal changes in sporadic and familial head and neck paragangliomas. Otolaryngol Head Neck Surg 2009;140:724-9.

44. Sharma MS, Gupta A, Hale SS, Agrawal D, Mahapatra AK, Sharma BS. Gamma knife radiosurgery for glomus jugulare tumors: Therapeutic advantages of minimalism in the skull base. Neuro India 2008;56:56-71.

45. Sheehan J, Kondziolka D, Flickinger J, Lunsford LD. Gamma knife surgery after tailored surgical resection: A novel treatment paradigm for glomus jugulare tumors. Stereotact Funct Neurosurg 2009;87:3-16.

46. Sinha S, Sharma MC, Sharma BS. Malignant paraganglioma of the sellar region mimicking a pituitary macroadenoma. J Clin Neurosci 2008;15:937-9.

47. Steel TR, Dailey AT, Born D, Berger MS, Mayberg MR. Paragangliomas of the sellar region: Report of two cases. Neurosurgery 1993;33:844-7.

48. Steiner HJ, Peringer P, Moser P, Schmid T, Kern MA, Steger CM, et al. Improved ultrastructural resolution by a new re-embedding procedure for formaldehyde-fixed paraffin-embedded tissues. Histopathology 2009;54:775-7.
Commentary

Paraganglioma

I read with interest this case report describing a recurrent paraganglioma occurring in the region of the Meckel’s cavium. The authors not only have made a wide search in the literature but have presented a good discussion regarding the tumorigenesis and histopathological confirmation of similar cases which can be quite informative for the researchers in the field of neuropathology.

As the authors mentioned, paragangliomas are the tumors arising from neuroectodermal-derived extra-adrenal paraganglia or chemoreceptor bodies. The central nervous system involvement may occur by: 1) direct extension from primary tumors of the glomus jugulare or glomus tympanicum, 2) spinal cord or brain compression from metastatic deposits in bone or directly within the brain and 3) primary involvement of the pineal gland or filum terminale.[1] This case can be either a primary tumor originating within the Meckel’s cavium or from the paraxially located paraganglionic cells.

REFERENCE

76. Kamalian N, Abbassioun K, Amirjamshidi A, Shams-Shahrabadi M. Paraganglioma of the filum terminale internum. Report of a case and review of the literature. J Neurol 1987;235:65-9.

Abbas Amirjamshidi
Department of Neurosurgery, Sina Hospital, Tehran University of Medical Sciences, Tehran, Iran
E-mail: alamirjamshidi@yahoo.com