

Foramen Magnum Meningioma Presenting with Cough Syncope (Case Report and Review of the Literature)

Meningioma de forame magno apresentando com síncope da tosse (Relato de caso e revisão da literatura)

Metin Kaplan¹ Omer Batu Hepgünsel¹ Selman Kok¹ Murat Gonen²

¹Department of Neurosurgery, Firat Universitesi Hastanesi, Elazig, Turkey

²Department of Neurology, Firat Universitesi Hastanesi, Elazig, Turkey

Address for correspondence Metin Kaplan, MD, Department of Neurosurgery, Firat Universitesi Hastanesi, Beyin ve Sinir Cerarhisi AD., Kat:6 23200, Elazig, Turkey (e-mail: mtkaplan02@yahoo.com.tr).

Arq Bras Neurocir 2018;37:334–338.

Abstract

Foramen magnum meningiomas cause different symptoms based on the size and the location of the tumor. They often present with involvement of the long tracts and of the lower cranial nerves. Ataxia and occipitocervical headache are other common symptoms. In the present study, we report a case of foramen magnum meningioma presenting with cough syncope. A mass lesion located anterolateral to the foramen magnum was detected in a 38-year-old man during a magnetic resonance imaging (MRI) exam; the lesion extended from the inferior clivus to the level of the C2 vertebra. The neural axis has pushed towards posterior and contralateral side by the mass. We think that syncope occurred due to the encasement of the vertebral arteries by the tumor in addition to the compression of the neural axis. The posterolateral approach without condylar resection provides a safe surgical plane for total excision of these tumors. In our case, the tumor was totally removed and the syncope episodes were resolved.

Keywords

- ▶ far lateral approach
- ▶ foramen magnum meningioma
- ▶ syncope

Resumo

Os meningiomas do forame magno causam sintomas diferentes com base no tamanho e na localização do tumor. Eles frequentemente apresentam envolvimento dos longos tratos e dos nervos cranianos inferiores. Ataxia e cefaleia occipitocervical são outros sintomas comuns. No presente estudo, relatamos um caso de meningioma do forame magno que apresentava síncope por tosse. Uma lesão em massa, localizada anterolateralmente ao forame magno, foi detectada em um homem de 38 anos durante um exame de ressonância magnética (RM). A lesão se estendia do clivus inferior ao nível da vértebra C2. O eixo neural foi empurrado para o lado posterior e contralateral pela massa. Pensamos que a síncope ocorreu devido ao encapsulamento das artérias vertebrais pelo tumor, além da compressão do eixo neural. A abordagem posterolateral sem ressecção condilar oferece um plano cirúrgico seguro para a excisão total desses tumores. No nosso caso, o tumor foi totalmente removido e os episódios de síncope foram resolvidos.

Palavras-chave

- ▶ abordagem lateral
- ▶ meningioma do forame magno
- ▶ síncope

received
September 19, 2018
accepted
October 30, 2018

DOI <https://doi.org/10.1055/s-0038-1676532>.
ISSN 0103-5355.

Copyright © 2018 by Thieme Revinter
Publicações Ltda, Rio de Janeiro, Brazil

License terms



Introduction

Approximately 0.2 to 3.2% of meningiomas are located in the foramen magnum which is between one-third inferior part of clivus and the level of the C2 vertebra. The majority of mass lesions are meningiomas in the foramen magnum.^{1,2} This small area harbors dense neural and vascular structures; thus, pathologies involving the foramen magnum display a wide spectrum of symptoms. Foramen magnum meningiomas mainly manifest with motor and sensorial deficits (hemiparesis, paresthesia), lower cranial nerve palsy, cervico-occipital pain, gait disorder, and cerebellar ataxia.^{3,4} It has been reported that they present with drop attacks in rare instances.⁵

Syncope is a mild form of temporary loss of consciousness. It is generally caused by transient global cerebral hypoperfusion and the patient recovers spontaneously within several minutes.⁶ It is known that congenital anomalies of the craniocervical junction lead to syncope and that cough may trigger syncope in these cases.⁷⁻¹²

In the present article, we report a case of foramen magnum meningioma presenting with cough syncope. To the best of our knowledge, no such case has been reported in the literature so far. In the present case report, we discuss the radiological appearance, the mechanism underlying the syncope episodes, and the treatment of the patient.

Case Report

A 38-year-old man presented with recurrent syncope episodes following cough. It was found that he was an active smoker but had no history of medication. He reported that he had infrequent occipitocervical headache, which worsened with cough. In addition, it was found that he had transient paresthesia involving different body regions.

Physical Examination

Motor and sensorial assessments were normal in the neurological examination. The lower cranial nerves were intact.

There were no pathological reflexes, with normoactive deep tendon reflexes. The cerebellar tests were found to be normal. In the cardiac examination, no pathology was detected. The electrocardiogram and echocardiogram exams were normal. Blood pressure was within the normal range. Complete blood count and biochemical tests were normal.

Radiological Evaluation

In a cranial magnetic resonance imaging (MRI) exam, a mass lesion with dense contrast enhancement compatible with meningioma was detected, which appeared hypointense on T1 weighted sequences and hyperintense in T2-weighted sequences. The mass lesion was anterolateral to the foramen magnum on the left side, extending from the inferior clivus to the level of the C2 vertebra. It was compatible with an L-type tumor, according to the classification system proposed by Kano et al,³ and, according to the classification system based on the relation with the vertebral artery (VA) proposed by Bernard et al, it was compatible with a both-sided tumor.^{13,14} The brainstem and the cervical cord were pushed posterior and contralateral side (►Fig. 1a and 1b). The V4 segment of both VAs were encased by the tumor (►Figs. 1a and 2). On the computed tomography (CT) scan, the calcification on the wall of the left V4 segment was striking (►Fig. 3).

Surgical Treatment

Depending on the size and location, several surgical approaches have been used in the treatment of foramen magnum meningiomas.^{3,14-17} In our case, we performed the surgery via the posterolateral approach (far lateral approach/retrocondylar far lateral approach). The VA transposition is generally unnecessary in intradural tumors of the foramen magnum.^{3,14} We did not perform a VA transposition in our case. There was a wide surgical plane, since the tumor shifted the neural axis toward the posterolateral space. Thus, no condylar resection or drilling of the jugular tubercle was required during surgery. The tumor was in the anterior side of the dentate ligament. Bernard et al. proposed another

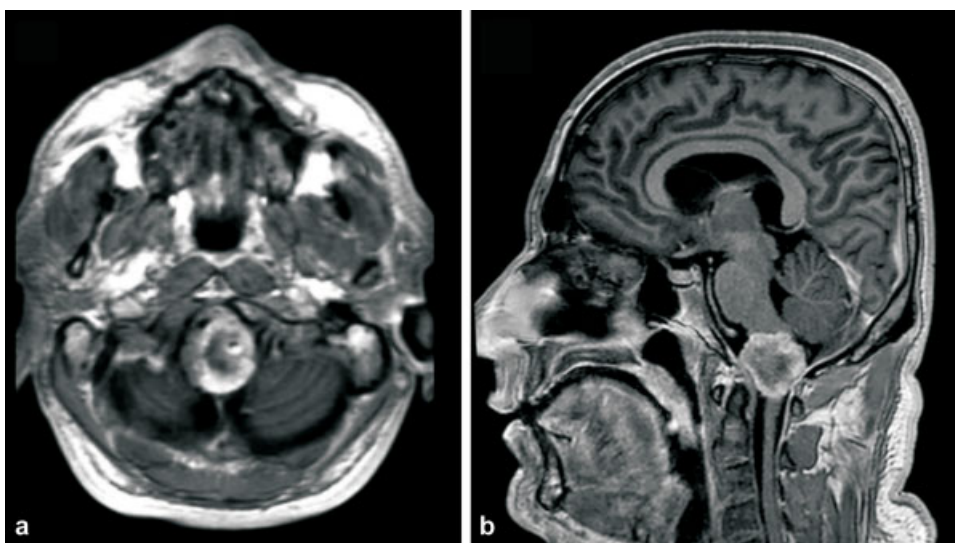


Fig. 1 (a–b) The T1-weighted magnetic resonance imaging (a; axial, b; sagittal) shows a mass lesion with dense contrast enhancement on the anterolateral foramen magnum, extending from the inferior clivus to the level of the C2 vertebra.

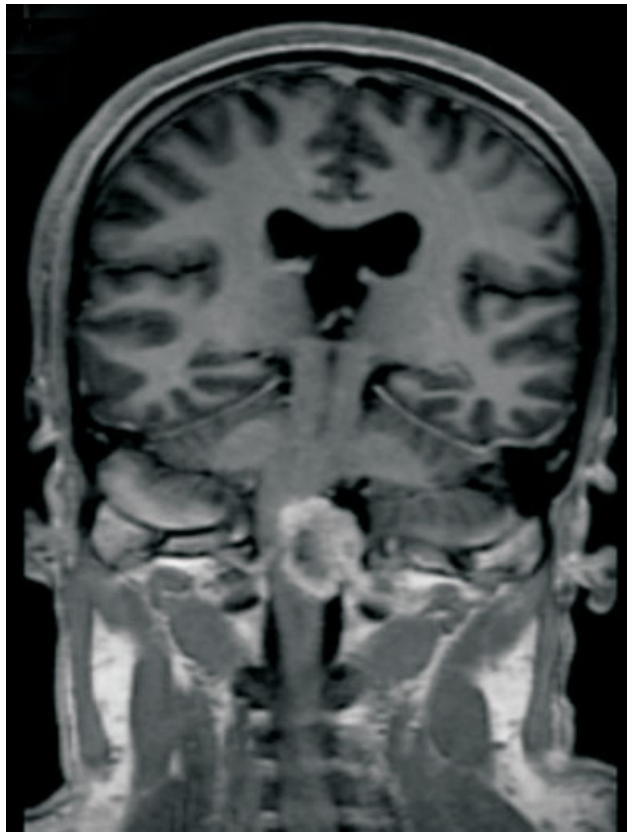


Fig. 2 T1-weighted contrast-enhanced coronal magnetic resonance imaging; The V4 segment of both VAs was encased by the tumor.

classification based on the relation of tumor with the dentate ligament.¹³ In our case, the tumor was compatible with lateral foramen magnum tumor according to this classification. The dentate ligament was transected to release the

neural axis. First, debulking was performed. The left VA was encased by the tumor; thus, the left VA and its branches were carefully dissected from the tumor tissue. After debulking, the contralateral VA was appeared. The tumor was totally excised while preserving the roots of the C1 and C2 vertebrae and the lower cranial nerves (► **Fig. 4a** and **4b**).

In the postoperative neurological examination, there was no motor or sensorial deficit. The lower cranial nerves were found to be normal. The syncope episodes following cough were fully resolved. At the follow-up visits at 3, 6, and 12 months, no headache, syncope episodes or abnormal neurological finding were observed. No recurrence was observed in the cranial MRI images.

Discussion

Foramen magnum meningiomas cause different symptoms, and findings vary depending on the localization and on the size of the tumor. They often present with involvement of the long tracts and of the lower cranial nerves. Cerebellar findings such as ataxia and headache, generally in the occipitocervical region, are also commonly seen symptoms.^{3,4,13,17,18} In our case, the presenting complaint were syncope episodes triggered by repetitive cough. This type of syncope episode has not been reported in association with foramen magnum meningiomas in the literature so far. Syncope or drop attacks can be rarely seen in association with craniocervical junction anomalies. The pathophysiology underlying syncope has not been fully understood in craniocervical junction anomalies. Mainly, midbrain ascending reticular activating system compression and insufficiency due to compression in vertebrobasilar flow has been discussed.^{7-12,19,20} Ireland et al reported transient medullar compression as the cause of syncope.²⁰ The presence of

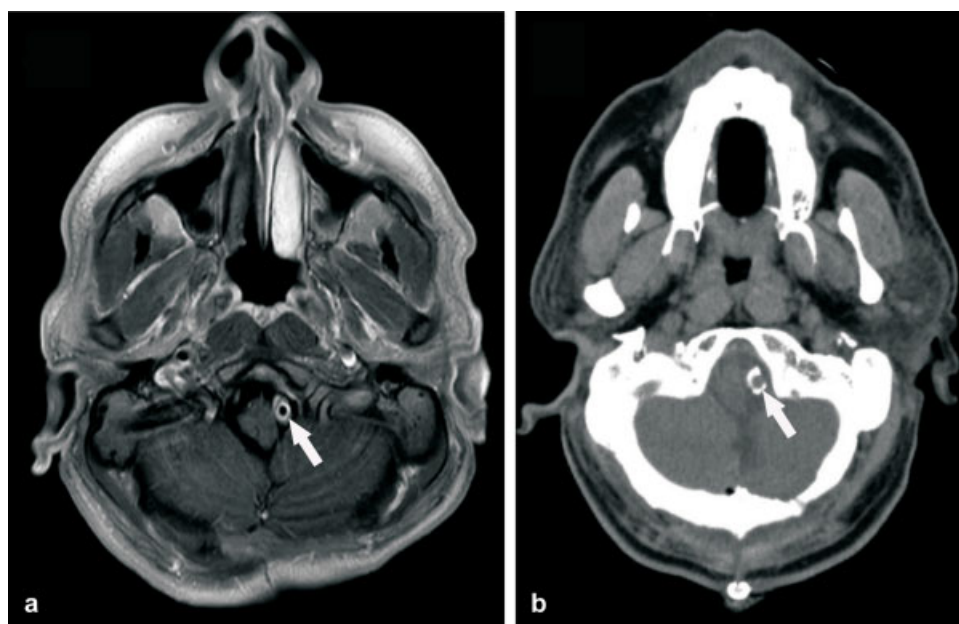


Fig. 3 (a–b) (a) Axial T1-weighted magnetic resonance imaging (b) computed tomography scan, showing the calcification on the wall of the left V4 segment (white arrows).

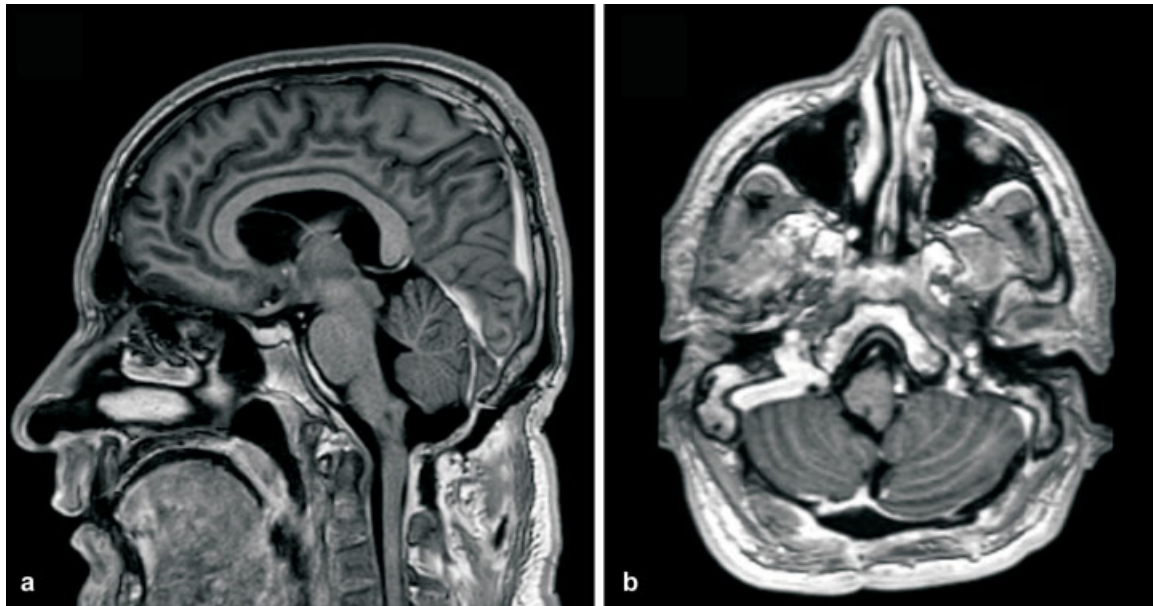


Fig. 4 (a–b) T1-weighted axial (a), sagittal (b) magnetic resonance imaging. The tumor was totally excised.

cardiovascular efferent sympathetic and parasympathetic neural pathways (cranial nerves IX and X, nucleus of the solitary tract) in the medulla oblongata supports this hypothesis.

There is a striking report on the relationship between the mechanism of syncope following cough and cerebellar tonsil herniation by Larson et al.¹² Cough induces syncope by increasing the medullar compression in cases with craniocervical junction anomalies (for example, Chiari, basilar invagination, atlas assimilation). Cough enhances the compression effect of the foramen magnum tumor on the medulla by elevating the intracranial pressure; thus, the increased medullar compression influences the cardiovascular neural pathways, resulting in reflex cardiac inhibition/vasodepression. This causes syncope. We called this mechanism as a “direct mechanism” which explains syncope that occurs due to the neural axis compression by the foramen magnum tumor.

Vertebrobasilar insufficiency (VBI) is another pathophysiological mechanism implied in syncope in craniocervical junction anomalies.^{9,10} The elevated intracranial pressure due to cough exacerbates the compression on the vertebrobasilar system, with a decrease in cerebral perfusion. It is well known that bilateral VA compression is a cause of syncope.²¹ In this hypothesis, anomalies of the vertebrobasilar system or vertebrobasilar malposition play an important role.⁷ In our case, the tumor encased the V4 segments of both VAs. Prolonged cough with increased compression by the tumor on the VAs may be the cause of syncope by decreasing cerebral perfusion with an elevated intracranial pressure. We think that the thickening and the marked calcification on the wall of the left VA had an impact on the decreased blood flow in our case. In addition, elevated intrathoracic pressure during prolonged cough decreases the venous return, resulting in a decreased cardiac output.⁶ The decreased cardiac output enhances the vertebrobasilar insufficiency. We have called this mechanism as an

“indirect mechanism” that explains syncope that occurs because of vertebrobasilar insufficiency.

Are the syncope episodes seen in our case episodes of primary reflex syncope or are they caused by the mass lesion in the foramen magnum? It is extremely challenging to differentiate these two conditions. In our case, the complete resolution of the syncope episodes after the resection of the mass lesion indicates that the foramen magnum mass was the cause of the syncope episodes.

The surgery in tumors located anterolateral to the foramen magnum is a specialized procedure due to the presence of neural and vascular structures in this region, and to the anatomy of the craniovertebral junction. Tumor size and location are important parameters in the selection of the surgical approach. The posterolateral approach (far lateral approach) including partial and complete condylar resection and drilling of the jugular tubercle, or the midline posterior approach, are commonly used in the surgery of these tumors.^{2–4,14,17,22} The encasement of the vertebral artery by the tumor is another factor limiting the complete resection of these tumors. In our case, there was a marked encasement of bilateral VAs, calcification, and thickening of the wall of the right VA. Similarly to the report of Kano et al,³ the large L-type meningioma in our case was totally excised by the posterolateral approach (far lateral approach) without condylar resection. This approach provided a safe surgical plane for the excision of the tumor without insulting the calcified VA and the branches encased by the tumor.

In conclusion, foramen magnum meningiomas may present with cough syncope. The encasement of the VA by the tumor, tumor invasion of the arterial wall, and calcification may play a role in syncope. The posterolateral (far lateral) approach provides a safe surgical plane for total excision of the tumor without condylar resection. Total excision provides cure of the syncope episodes.

Conflicts of Interest

The authors have no conflicts of interest to declare.

References

- 1 Yasargil MG, Mrtara RW, Curcic M. Meningiomas of basal posterior cranial fossa. *Adv Tech Stand Neurosurg* 1980;7:1–115
- 2 Arnautović KI, Al-Mefty O, Husain M. Ventral foramen magnum meningiomas. *J Neurosurg* 2000;92(1, Suppl):71–80
- 3 Kano T, Kawase T, Horiguchi T, Yoshida K. Meningiomas of the ventral foramen magnum and lower clivus: factors influencing surgical morbidity, the extent of tumour resection, and tumour recurrence. *Acta Neurochir (Wien)* 2010;152(01):79–86, discussion 86
- 4 Flores BC, Boudreaux BP, Klinger DR, Mickey BE, Barnett SL. The far-lateral approach for foramen magnum meningiomas. *Neurosurg Focus* 2013;35(06):E12
- 5 Mahore A, Ramdasi R, Mavani S, et al. Meningioma of foramen magnum causing drop attacks. *Case Rep Neurol Med* 2015; 2015:214563
- 6 Adkisson WO, Benditt DG. Pathophysiology of reflex syncope: A review. *J Cardiovasc Electrophysiol* 2017;28(09): 1088–1097
- 7 Miyakoshi N, Hongo M, Kasukawa Y, Shimada Y. Syncope caused by congenital anomaly at the craniovertebral junction: a case report. *J Med Case Reports* 2014;8:330
- 8 Corbett JJ, Butler AB, Kaufman B. 'Sneeze syncope', basilar invagination and Arnold-Chiari type I malformation. *J Neurol Neurosurg Psychiatry* 1976;39(04):381–384
- 9 Mangubat EZ, Wilson T, Mitchell BA, Byrne RW. Chiari I malformation associated with atlanto-occipital assimilation presenting as orthopnea and cough syncope. *J Clin Neurosci* 2014;21(02): 320–323
- 10 Shin HS, Kim JA, Kim DS, Lee JS. Type I. Chiari malformation presenting ortostatic syncope who treated with decompressive surgery. *Korean J Pediatr* 2016;59(Suppl 1):149–151
- 11 Prilipko O, Dehdashti AR, Zaim S, Seeck M. Orthostatic intolerance and syncope associated with Chiari type I malformation. *J Neurol Neurosurg Psychiatry* 2005;76(07):1034–1036
- 12 Larson SJ, Sances A Jr, Baker JB, Reigel DH. Herniated cerebellar tonsils and cough syncope. *J Neurosurg* 1974;40(04):524–528
- 13 Bruneau M, George B. Foramen magnum meningiomas: detailed surgical approaches and technical aspects at Lariboisière Hospital and review of the literature. *Neurosurg Rev* 2008;31(01):19–32, discussion 32–33
- 14 George B, Lot G, Boissonnet H. Meningioma of the foramen magnum: a series of 40 cases. *Surg Neurol* 1997;47(04):371–379
- 15 Bertalanffy H, Seeger W. The dorsolateral, suboccipital, transcondylar approach to the lower clivus and anterior portion of the craniocervical junction. *Neurosurgery* 1991;29(06):815–821
- 16 Pamir MN, Kiliç T, Ozduman K, Türe U. Experience of a single institution treating foramen magnum meningiomas. *J Clin Neurosci* 2004;11(08):863–867
- 17 Samii M, Klekamp J, Carvalho G. Surgical results for meningiomas of the craniocervical junction. *Neurosurgery* 1996;39(06):1086–1094, discussion 1094–1095
- 18 Bassiouni H, Ntoukas V, Asgari S, Sandalcioğlu EI, Stolke D, Seifert V. Foramen magnum meningiomas: clinical outcome after microsurgical resection via a posterolateral suboccipital retrocondylar approach. *Neurosurgery* 2006;59(06):1177–1185, discussion 1185–1187
- 19 Straus D, Foster K, Zimmerman F, Frim D. Chiari drop attacks: surgical decompression and the role of tilt table testing. *Pediatr Neurosurg* 2009;45(05):384–389
- 20 Ireland PD, Mickelsen D, Rodenhouse TG, Bakos RS, Goldstein B. Evaluation of the autonomic cardiovascular response in Arnold-Chiari deformities and cough syncope syndrome. *Arch Neurol* 1996;53(06):526–531
- 21 Felbaum DR, Ryan JE, Stemer AB, Anaizi A. Bilateral subaxial rotational vertebral artery occlusion in setting of a prior cervical construct. *World Neurosurg* 2017;97:762.e5–762.e10
- 22 Sohn S, Chung CK. Conventional posterior approach without far lateral approach for ventral foramen magnum meningiomas. *J Korean Neurosurg Soc* 2013;54(05):373–378