



Rare Case of Young Patient with Intraventricular Angiomatous Meningioma

Caso raro de paciente jovem com meningioma angiomatoso intraventricular

Gabriel Carvalho Heemann¹ Rafael Silva Paglioli² Ricardo Chmelnitsky Wainberg³

¹ Medicine Student, Pontifícia Universidade Católica do Rio Grande do Sul (PUC-RS), Porto Alegre, RS, Brazil

² Resident doctor, Neurosurgery Service, Hospital São Lucas, PUC-RS, Porto Alegre, RS, Brazil

³ Preceptor doctor, Neurosurgery Service, Hospital São Lucas, PUC-RS, Porto Alegre, RS, Brazil

Address for correspondence Gabriel Carvalho Heemann, Medicine Student, Pontifical Catholic University of Rio Grande do Sul (PUC-RS), Porto Alegre, RS, Brazil (e-mail: gabriel.heemann@gmail.com).

Arq Bras Neurocir 2021;40(3):e257–e262.

Abstract

Pediatric meningiomas are rare and account for only 2.2% of the central nervous system (CNS) tumors. In this age group, they are more frequently located in atypical sites, such as, mainly, the ventricular system, with a frequency of 8.8 to 13.6%. Adding this to the fact that the angiomatous subtype constitutes only 2.1% of all meningiomas, the rarity of the case reported here is corroborated. We report a 17-year-old female patient diagnosed with intraventricular angiomatous meningioma; she underwent surgical resection of the tumor in the body and frontal horn of the right lateral ventricle, and there were no neurological sequelae. With a follow-up of 26 months, there was no recurrence and the patient had clinical stability. Intraventricular tumors usually have slow growth and reach a considerable size until they cause symptoms and then are diagnosed. In addition, the deep location of the tumor and its proximity to eloquent areas make these tumors a neurosurgical challenge. The angiomatous subtype, due to the presence of hypervascularization (consisting of > 50% of vascular components), may, in some cases, hinder surgical resection as well as be erroneously diagnosed. However, surgical treatment aimed at total resection of the lesion remains the conduct of choice in the case reported here, especially in patients in the first two decades of life, in which the use of radiation is avoided. Specifically when it comes to the surgery, we chose a transcallosal approach that allows a good transoperative visualization of the lesion when located in the body and frontal horn of the lateral ventricle.

Keywords

- ▶ meningioma
- ▶ intraventricular neoplasms
- ▶ pediatrics
- ▶ lateral ventricles

Resumo

Meningiomas pediátricos são raros, representando apenas 2,2% dos tumores do sistema nervoso central (SNC). Em tal faixa etária, localizam-se com maior frequência em sítios atípicos, como, principalmente, o sistema ventricular – com frequência de 8,8

received
February 20, 2017
accepted
May 31, 2017
published online
June 23, 2017

DOI <https://doi.org/10.1055/s-0037-1604037>.
ISSN 0103-5355.

© 2017. Sociedade Brasileira de Neurocirurgia. All rights reserved. This is an open access article published by Thieme under the terms of the Creative Commons Attribution-NonDerivative-NonCommercial-License, permitting copying and reproduction so long as the original work is given appropriate credit. Contents may not be used for commercial purposes, or adapted, remixed, transformed or built upon. (<https://creativecommons.org/licenses/by-nc-nd/4.0/>)
Thieme Revinter Publicações Ltda., Rua do Matoso 170, Rio de Janeiro, RJ, CEP 20270-135, Brazil

a 13,6%. Em vista disso, e de que o subtipo angiomatoso constitui apenas 2,1% de todos meningiomas, corrobora-se a raridade do caso aqui relatado: paciente feminina de 17 anos diagnosticada com meningioma angiomatoso intraventricular. Foi submetida à ressecção cirúrgica de tumor em corpo e corno frontal do ventrículo lateral direito, não havendo seqüela neurológica. Com acompanhamento de 26 meses, não apresentou recidiva e demonstra estabilidade clínica. Tumores intraventriculares geralmente possuem crescimento lento e atingem tamanho considerável até ocasionarem sintomas e serem, então, diagnosticados. Somado a isso, a localização profunda do tumor e a proximidade com áreas eloquentes tornam tais tumores um evidente desafio neurocirúrgico. O subtipo angiomatoso, por apresentar hipervascularização (é constituído por > 50% de componentes vasculares), pode tanto dificultar, em alguns casos, a ressecção cirúrgica quanto ser erroneamente diagnosticado. Porém, o tratamento cirúrgico visando à ressecção da lesão mantém-se como a conduta de escolha no caso aqui relatado, especialmente em pacientes nas duas primeiras décadas de vida, nos quais se evita o uso de radiação. Especificamente em relação à cirurgia, optou-se por uma abordagem transcalosa que possibilita uma boa visualização transoperatória da lesão quando localizada em corpo e corno frontal do ventrículo lateral.

Palavras-chave

- ▶ meningioma
- ▶ neoplasia intraventricular
- ▶ pediatria
- ▶ ventrículos laterais

Introduction

Meningiomas have a progressively higher incidence with increasing age, with a mean age of presentation of 65 years old. Thus, they constitute the most frequently reported tumors of the central nervous system (CNS) in adulthood.¹ Children and adolescents cases are rare, representing 2.2 to 2.6% of CNS tumors.^{1,2} In this age group, they are more frequently located in unusual sites, such as in the ventricular system.³

Such intraventricular meningiomas have the particularities of being slow-growing and reaching a considerable size until they become symptomatic.⁴ In addition, the deep localization and the relationship with the underlying eloquent areas make tumor resection a neurosurgical challenge.^{5,6} In view of this, and that the angiomatous subtype – defined for presenting > 50% of the vascular components at the microscopic analysis – constitutes only 2.1% of all meningiomas, it is credited to the case reported herein.⁷ We are conscious of the fact that this is possibly the first report of a patient in the first 2 decades of life with angiomatous meningioma in the intraventricular site.

In this report, we aim to expose our neurosurgical experience in a case with rare variants and to conduct a review of the literature on the main aspects that we deem necessary to support our conduct.

Case Description

A 17-year-old female patient, previously healthy, presented orbital headache for 3 months, followed by blurred and double vision. On physical examination, convergent strabismus by right lateral rectus muscle paresis was found. In the other cranial pairs, sensitivity, strength and reflexes were preserved and there were no meningeal signs. It was then performed an investigation with cranial magnetic resonance imaging (MRI),

which evidenced an expansive lesion in the frontal horn of the right ventricle, directly ahead of the foramen of Monro, with dimensions of 2.0 × 1.3 × 1.8 cm (AP x L x H) in the larger diameter sections. The lesion presented moderate hyperintensity with small hypointense foci in the T2 weighted images. On T1, it was isointense, and after the contrast showed intense and homogeneous impregnation, except for the same hypointense T2-weighted spots. There were no signs of dilatation of the supratentorial ventricular system (▶Fig. 1 A-D). Such radiological features suggested the diagnosis of intraventricular

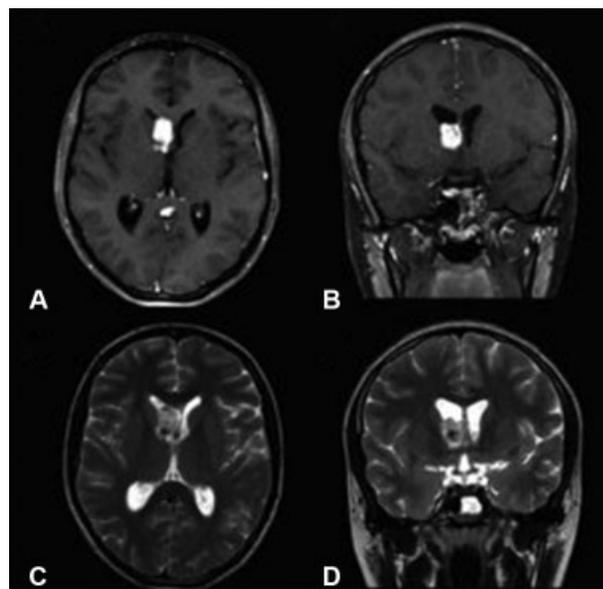


Fig. 1 Preoperative magnetic resonance imaging: A and B – T1 with gadolinium, axial (a) and coronal (b). Lesion with intense uptake at the level of the right frontal horn, close to the foramen of Monro. C and D – T2 axial (C) and coronal (D). Hyperintense lesion, with a focus of hypointense calcification (also visualized in A).

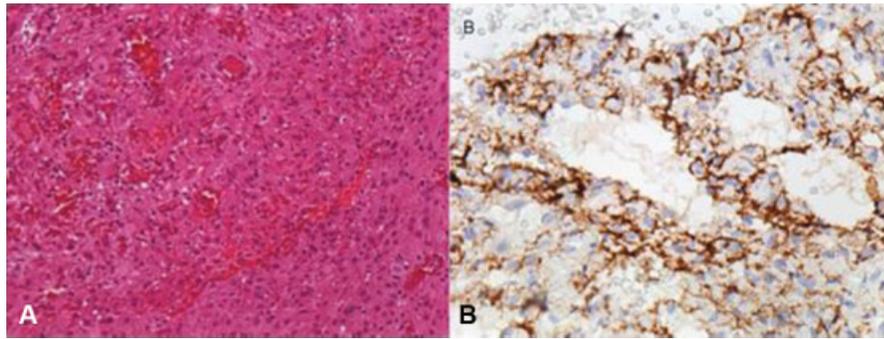


Fig. 2 (A) Hematoxylin and eosin staining, 200 x magnification: Image evidencing histopathological features of meningioma and significant vascular component. (B) Immunohistochemistry, Magnification - Analysis of 400 times: tumor cells presenting positive for epithelial membrane antigen.

meningioma, with calcification areas. It was considered an occasional finding, since the topography was not compatible with the presentation symptoms.

Surgical treatment was indicated, and this was performed at the same hospitalization, due to the risk of acute hydrocephaly. The approach was performed by the transcalous approach, with the following steps: dorsal decubitus and head in neutral position; bicorony incision; right frontal paramedian craniotomy, with lateral extension of 5.5 cm from the midline and 5.5 cm from the coronary suture to the front; opening of the dura mater in “Ç” format based on the midline; under microscopy, dissection of the interhemispheric fissure and removal of the frontal lobe with positioning of the fixed spatula in Leyla support on the medial surface; identification of the rotating of the cingulate and pericalous arteries; 1.2 cm callosotomy starting from the transition between the knee and the corpus callosum body; identified the vegetating greyish lesion inside the right frontal horn, which presented softened consistency, aspirable after coagulation, and vascularized; resection of the

lesion through coagulation and aspiration and by fragments, providing small residue adhered to the ependyma of the thalamocaudal droppings. Postoperative without interferences and absence of new deficits, remaining diplopia and strabismus. The histopathologic analysis and the immunohistochemical profile demonstrated extensive vascularization and low mitotic index (Ki67 < 2%). In addition, the results verified are: epithelial membrane antigen (EMA), positive; cytokeratin (CK), negative; progesterone receptor, negative; and glial fibrillar acid protein (GFAP), positive (►Fig. 2 A-B). Thus, the diagnosis of angiomatous meningioma (World Health Organization [WHO] grade I) is confirmed. The postoperative MRI showed small residual focus along the striated thalamus (►Figs. 3 A-D and 4 A-D). After 6 months, the patient underwent strabismus correction with an ophthalmologist at another institution. Currently, with ~ 26 months of follow-up, she is asymptomatic and without evidence of recurrence of the residual lesion.

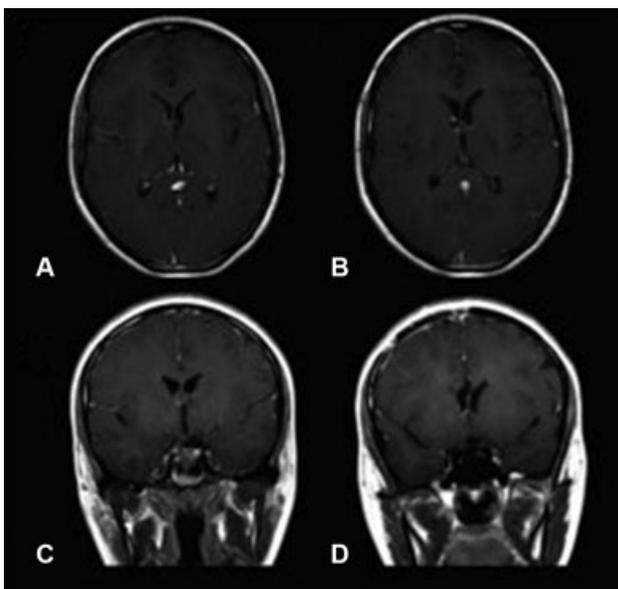


Fig. 3 Postoperative magnetic resonance imaging: T1 with gadolinium, axial planes (A and B) and Coronals (C and D). Small residual focus near the foramen of Monro, adhered to the striate thalamus vein (identified in the transoperatory).

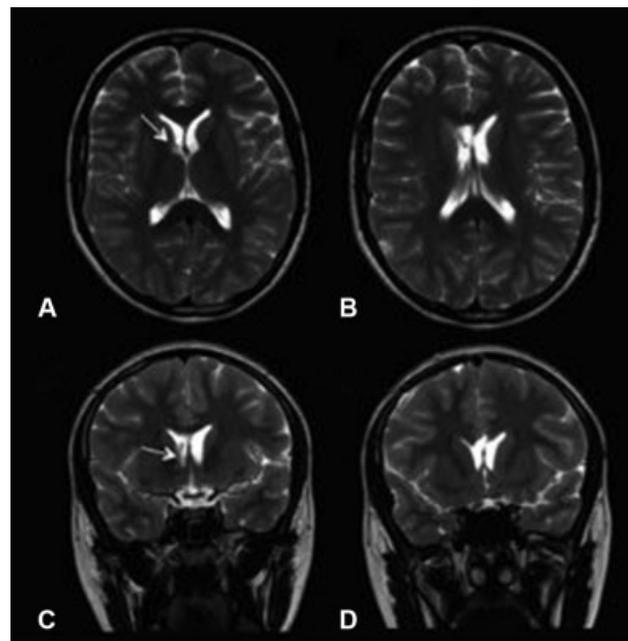


Fig. 4 Postoperative magnetic resonance imaging: T2 axial (A and B) and coronal (C and D). Small residual focus at the level of the foramen of Monro (arrows).

Discussion

Meningiomas are tumors that predominate in the 5th and 6th decades of life, having an average age of presentation of 65 years old.¹ In general, they represent 36.4% of the primary CNS tumors and ~ between 24 and 30% in adults.^{1,8} On the other hand, in the pediatric population, the prevalence of CNS tumors varies between 0.4 and 4.6%.^{2,3,9-13} Gender equivalence also contrasts with what occurs in the adult population, which has a ratio between female and male gender of 2:1.^{1,14} It is believed that this difference is due, especially in the prepubertal period, to the absence of the effect of hormones in the corticosteroid receptors of the cells of the meningioma.^{10,15-17}

In the first two decades of life, there is a higher incidence of grade II (atypical) and grade III (anaplastic) meningiomas, according to the WHO: 9.9 and 8.9%, respectively.^{2,18} They are characterized by being genetically and phenotypically more aggressive, with a high frequency of cerebral invasion.^{9,14} Among the most frequent grade I meningiomas, the angiomatous subtype occurs in 2.8% of the cases,² and in 2.1% of all meningiomas at any age.¹⁹ This subtype is defined when the vascular component exceeds 50% of the total tumor area.^{7,19} However, differential diagnosis is necessary with hemangioblastoma and hemangiopericytoma, with essential immunohistochemical and morphology roles in the diagnostic confirmation: MIB-1/Ki67 low index and positivity for progesterone receptor, EMA, vimetin, cytokeratin and desmoplatelet.^{7,19-23}

Since these tumors are uncommon, the characteristics of angiomatous meningiomas are considered in few studies.^{7,19,24,25} They may present moderate to severe cerebral edema with a frequency of 74 to 88.9%,^{7,19} due to hypervascularization, increased capillary permeability and vascular endothelial growth factor (VEGF) secretion.¹⁹ In magnetic resonance imaging, they may present more signs of flow voids, rarely present necrosis, and they tend to have homogeneous enhancement to paramagnetic contrast.^{19,23}

Meningiomas in pediatric patients present in atypical sites more frequently than in adults: in the lateral ventricles, in the skull base, and in the posterior fossa.^{2,3,10,15,26} The intraventricular localization occurs in 11%, compared with between 0.3 and 3% in all ages and between 0.5 and 4.5% in adults.^{2,10,27}

Intraventricular meningiomas (IVMs) are in the lateral ventricles (more common on the left side) in 76% of the cases; 16% in the 3rd ventricle; and 7%, in the 4th ventricle.^{5,27} There are studies suggesting that lateral ventricles are the favorite site of pediatric IVMs.^{26,28} These originate from the choroid plexus, growing on the choroid screen.⁴ The vascularization of the tumor depends on its location in the ventricle, and, in general, the main nutrient vessels depart from the choroidal arteries and are of small caliber.²⁷

Clinically, pediatric IVMs are usually asymptomatic, until they reach large dimensions in the lateral ventricles, where the risk of hydrocephaly is lower. On the other hand, when located in the 3rd or 4th ventricle, the obstruction of the cerebrospinal fluid (CSF) flow may result in manifestations in early stages of

the tumoral growth.^{5,27,29,30} Therefore, symptoms – headache, nausea, vomiting, and visual disturbances^{5,27,31} – are more frequently related to tumor compression and to an insidious increase in intracranial pressure. Indolent cognitive deficits compromising memory and attention can also occur.^{32,33} Typical symptoms of acute intracranial pressure increase are uncommon.²⁹ The clinic thus correlates with the location of the tumor within the ventricle, the size of the tumor and the direction of its growth.²⁷ Finally, we emphasize that the clinical presentation of the patient reported – convergent strabismus by paresis of the right lateral rectus muscle – showed no correlation with the tumor, which still had a relatively small size and its location did not justify the signs and symptoms.

Intraventricular meningiomas usually present the classic radiological appearance of other meningiomas: well-defined globular form, but without dural tail. They are usually isointense to hypointense in T1-weighted images, hyperintense in T2-weighted image and undergo strong contrast enhancement.^{16,27} In particular, in the pediatric population, other more frequent intraventricular tumors may difficult the differential diagnosis: choroid plexus tumors, ependymoma, primitive neuroectoplasmic tumor, teratoma and astrocytoma.^{30,34} Choroid plexus tumors usually affect children < 10 years old, and, at MRI, they have a multilobulated mass with intense contrast enhancement and fronds appearance. Ependymomas represent approximately one-third of CNS tumors in children < 3 years old and are characterized by necrosis, hemorrhage, cyst formation, and for presenting, in MRI, hypointense in T1 and hyperintense and heterogeneous in T2.³⁵

The surgical approach of a benign IVM is a neurosurgical challenge, in view of its deep location and its proximity to eloquent areas and vessels of the ventricles walls.^{5,34} The extension of the initial resection is an independent prognostic factor, with significant association with recurrence and malignization.² The patient reported here did not present recurrence in the 26 months of follow-up, which is a result consistent with the literature. In a 2012 review, with 201 cases of several series, there were only 8 recurrences.²⁷ However, in a meta-analysis with 677 cases of meningiomas in the first 2 decades, the numbers are more meaningful. There were 141 recurrences, with an average presentation of 3.6 years and with mortality for this event in 46 cases.² Recurrence in this age group occurs basically in cases of atypical and anaplastic meningiomas, or after partial resection.³⁶ Mortality and postsurgical morbidity in postpubertal patients, as is the case of the patient reported here, are approaching the ones observed in meningioma cases in adults.² The use of adjuvant radiotherapy should be avoided in young patients, and it may be possible to opt for serial evaluation and reoperation in case of recurrence.^{2,3,8-11,14,18,37}

In the literature, there are several surgical approaches for IVM resection: temporoparietal approach, transfrontal, medial posterior temporal gyrus, posterior inferior temporal gyrus, parieto-occipital, and transcalous.^{5,34} The choice is individualized and based on the location of the tumor within the ventricle, the tumor size and its vascular network, to always preserve the adjacent cerebral tissue, performing small

corticoectomies and retracting as little as possible.^{5,6,27,30,33,34} The rationality behind the choice of the gem approach is determined by the option that allows the best access to the largest axis of the lesion, to minimize transcortical transgression, by the spectrum of neurological deficits Pre-operative, proximity with the aforementioned eloquent structures, besides the anatomical knowledge of the cortical and white substance.²⁹ In the reported case, we chose the transcallosal approach to be the one that allows the best access to the frontal horn and lateral ventricle body. This approach avoids cortical injury; However, certain care is needed with the possible presence of tributary cortical veins of the superior sagittal sinus, which can be anticipated in the preoperative examinations, and with the corpus callosum, which should be distinguished from the gyrus rotation by the color change.⁵ The posterior transcallosal approach has disconnection syndrome as one of its possible complications; however, the experience in our neurosurgery service demonstrates that the risk is minimal, since we have performed surgeries with the resection of the two thirds Posterior callosotomy of the corpus callosum with very low rate of this postoperative syndrome.^{5,30,33,38} Despite the degree of difficulty, IVM surgery has shown low rates of morbidity and mortality in the last decades, and most postoperative complications – visual deficits and apraxias – are temporary.^{27,29,34} These low rates are consistent with the case reported here, which did not present postoperative complications or transoperative sequelae.

Conclusion

Intraventricular angiomatous meningioma is a rare entity, even more in patients in the first 2 decades of life. The clinic is unspecific in most cases, making it necessary to assess MRI for diagnosis and definition of the surgical approach, and the histopathological analysis is what defines the diagnosis of the angiomatous subgroup. Surgical resection is the treatment of choice. However, the objective of total resection should not be above the objective of preserving the functions and the quality of life of the patient.

Conflicts of Interests

The authors declare that there are no conflicts of interests.

References

- Ostrom QT, Gittleman H, Fulop J, et al. CBTRUS Statistical Report: Primary Brain and Central Nervous System Tumors Diagnosed in the United States in 2008-2012. *Neuro-oncol* 2015;17(Suppl 4): iv1-iv62
- Kotecha RS, Pascoe EM, Rushing EJ, et al. Meningiomas in children and adolescents: a meta-analysis of individual patient data. *Lancet Oncol* 2011;12(13):1229-1239
- Menon G, Nair S, Sudhir J, Rao BR, Mathew A, Bahuleyan B. Childhood and adolescent meningiomas: a report of 38 cases and review of literature. *Acta Neurochir (Wien)* 2009;151(03): 239-244, discussion 244
- Bhatoe HS, Singh P, Dutta V. Intraventricular meningiomas: a clinicopathological study and review. *Neurosurg Focus* 2006;20(03):E9
- Nanda A, Bir SÇ, Maiti T, Konar S. Intraventricular Meningioma: Technical Nuances in Surgical Management. *World Neurosurg* 2016;88:526-537
- Lyngdoh BT, Giri PJ, Behari S, Banerji D, Chhabra DK, Jain VK. Intraventricular meningiomas: a surgical challenge. *J Clin Neurosci* 2007;14(05):442-448
- Hasselblatt M, Nolte KW, Paulus W. Angiomatous meningioma: a clinicopathologic study of 38 cases. *Am J Surg Pathol* 2004;28(03): 390-393
- Kwee LE, van Veelen-Vincent ML, Michiels EM, Kros JM, Dammers R. The importance of microsurgery in childhood meningioma: a case report. *Childs Nerv Syst* 2015;31(01):161-165
- Liu Y, Li F, Zhu S, Liu M, Wu C. Clinical features and treatment of meningiomas in children: report of 12 cases and literature review. *Pediatr Neurosurg* 2008;44(02):112-117
- Ravindranath K, Vasudevan MÇ, Pande A, Symss N. Management of pediatric intracranial meningiomas: an analysis of 31 cases and review of literature. *Childs Nerv Syst* 2013;29(04):573-582
- Santos MV, Furlanetti L, Valera ET, Brassesco MS, Tone LG, de Oliveira RS. Pediatric meningiomas: a single-center experience with 15 consecutive cases and review of the literature. *Childs Nerv Syst* 2012;28(11):1887-1896
- Cushing HL, Eisenhardt L. Meningiomas: their classification, regional behavior, life history and surgical end results. Charles C Thomas Springfield, Ill; 1938
- Mehta N, Bhagwati S, Parulekar G. Meningiomas in children: A study of 18 cases. *J Pediatr Neurosci* 2009;4(02):61-65
- Arivazhagan A, Devi BI, Kolluri SV, Abraham RG, Sampath S, Chandramouli BA. Pediatric intracranial meningiomas—do they differ from their counterparts in adults? *Pediatr Neurosurg* 2008; 44(01):43-48
- Jaiswal S, Vij M, Mehrotra A, Jaiswal AK, Srivastava AK, Behari S. A clinicopathological and neuroradiological study of paediatric meningioma from a single centre. *J Clin Neurosci* 2011;18(08): 1084-1089
- Whittle IR, Smith C, Navoo P, Collie D. Meningiomas. *Lancet* 2004; 363(9420):1535-1543
- Donnell MS, Meyer GA, Donegan WL. Estrogen-receptor protein in intracranial meningiomas. *J Neurosurg* 1979;50(04):499-502
- Kotecha RS, Junckerstorff RÇ, Lee S, Cole CH, Gottardo NG. Pediatric meningioma: current approaches and future direction. *J Neurooncol* 2011;104(01):1-10
- Liu Z, Wang C, Wang H, Wang Y, Li JY, Liu Y. Clinical characteristics and treatment of angiomatous meningiomas: a report of 27 cases. *Int J Clin Exp Pathol* 2013;6(04):695-702
- Rao S, Rajkumar A, Kuruvilla S. Angiomatous meningioma: a diagnostic dilemma. *Indian J Pathol Microbiol* 2008;51(01):53-55
- Bansal D, Diwaker P, Gogoi P, Nazir W, Tandon A. Intraparenchymal Angiomatous Meningioma: A Diagnostic Dilemma. *J Clin Diagn Res* 2015;9(10):ED07-ED08
- Rathod GB, Vyas K, Shinde P, Goswami SS, Tandan RK. Angiomatous meningioma in 49 years old male A rare case report. *Int J Curr Microbiol Appl Sci* 2014;3(11):256-260
- Meng Y, Chaohu W, Yi L, Jun P, Songtao Q. Preoperative radiologic characters to predict hemangiopericytoma from angiomatous meningioma. *Clin Neurol Neurosurg* 2015;138:78-82
- Hwang J, Kong D-S, Seol HJ, Nam D-H, Lee J-I, Choi JW. Clinical and Radiological Characteristics of Angiomatous Meningiomas. *Brain Tumor Res Treat* 2016;4(02):94-99
- Ben Nsir A, Chabaane M, Krifa H, Jeme H, Hattab N. Intracranial angiomatous meningiomas: A 15-year, multicenter study. *Clin Neurol Neurosurg* 2016;149:111-117
- Rushing EJ, Olsen C, Mena H, et al. Central nervous system meningiomas in the first two decades of life: a clinicopathological analysis of 87 patients. *J Neurosurg* 2005;103(6, Suppl):489-495
- Ødegaard KM, Helseth E, Meling TR. Intraventricular meningiomas: a consecutive series of 22 patients and literature review. *Neurosurg Rev* 2013;36(01):57-64, discussion 64

- 28 Deen HG Jr, Scheithauer BW, Ebersold MJ. Clinical and pathological study of meningiomas of the first two decades of life. *J Neurosurg* 1982;56(03):317–322
- 29 Fusco DJ, Spetzler RF. Surgical considerations for intraventricular meningiomas. *World Neurosurg* 2015;83(04):460–461
- 30 Liu M, Wei Y, Liu Y, Zhu S, Li X. Intraventricular meningiomas: a report of 25 cases. *Neurosurg Rev* 2006;29(01):36–40
- 31 Rochat P, Johannesen HH, Gjerris F. Long-term follow up of children with meningiomas in Denmark: 1935 to 1984. *J Neuro Surg* 2004;100(2, Suppl Pediatrics):179–182
- 32 Kashiwazaki D, Takaiwa A, Nagai S, et al. Reversal of cognitive dysfunction by total removal of a large lateral ventricle meningioma: a case report with neuropsychological assessments. *Case Rep Neurol* 2014;6(01):44–49
- 33 Nakamura M, Roser F, Bundschuh O, Vorkapic P, Samii M. Intraventricular meningiomas: a review of 16 cases with reference to the literature. *Surg Neurol* 2003;59(06):491–503, discussion 503–504
- 34 Grujicic D, Cavallo LM, Somma T, et al. Intraventricular Meningiomas: A Series of 42 Patients at a Single Institution and Literature Review. *World Neurosurg* 2017;97:178–188
- 35 Agarwal A, Kanekar S. Intraventricular Tumors. *Semin Ultrasound CT MR* 2016;37(02):150–158
- 36 Caroli E, Russillo M, Ferrante L. Intracranial meningiomas in children: report of 27 new cases and critical analysis of 440 cases reported in the literature. *J Child Neurol* 2006;21(01):31–36
- 37 Okechi H, Albright AL. Intraventricular meningioma: case report and literature review. *Pediatr Neurosurg* 2012;48(01):30–34
- 38 Paglioli E, Martins WA, Azambuja N, et al. Selective posterior callosotomy for drop attacks: A new approach sparing prefrontal connectivity. *Neurology* 2016;87(19):1968–1974