

# Fourth ventricular schwannoma

[Intracranial schwannomas](#) have been reported in several unusual locations. [Intraventricular schwannomas](#) are rare primary brain tumors, with fewer than 25 cases reported in the literature.

In 1957, Marchand et al. published the first case of an intraventricular schwannoma <sup>1)</sup>.

Schwannomas of the fourth ventricle are infrequent but should be accounted in the differential diagnosis of space-occupying lesions in this location. Gross total resection might be the definite treatment of these tumors if deemed possible <sup>2)</sup>.

Although the embryologic origins may be different from nerve sheath-derived schwannomas, the histologic, clinical, and natural history appear identical and thus should be managed similarly <sup>3)</sup>.

The hypothesis of intraventricular schwannoma is postulated to be aberrantly placed multipotent cell during embryogenesis and later transforming into [Schwann cell](#) and producing schwannoma. Pertinent literature is reviewed along with diagnosis, and management of such rare case is discussed briefly <sup>4)</sup>.

In 2018 Zhu et al., reported two extremely rare cases of schwannomas originating in the [fourth ventricle](#), where it did not have any attachment to the surrounding structures. The clinical course, radiological and pathological features, treatment, and follow-up are described <sup>5)</sup>.

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In 2017 Moscote-Salazar et al., could find only seven cases reported in the literature in the form of isolated cases report, who were managed surgically, with only one being a pediatric case. They report the second case of intra-fourth ventricular schwannoma occurring in the pediatric age group. The hypothesis of intraventricular schwannoma is postulated to be aberrantly placed multipotent cell during embryogenesis and later transforming into Schwann cell and producing schwannoma. Pertinent literature is reviewed along with diagnosis, and management of such rare case is discussed briefly <sup>6)</sup>.

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In 2015 Santos et al., reported a 53-year-old man presented with a 1.5-year history of gait unsteadiness and vertigo and a few-week history of headache, emesis, and neurogenic dysphagia. A brain magnetic resonance imaging revealed a large, heterogeneously contrast enhancing mass located within the fourth ventricle, compressing the brainstem and causing supratentorial ventricle enlargement. A suboccipital craniotomy and a telovelar approach were performed to resect the tumor. The ventricular system was repermeabilized at the end of the operation.

A postoperative magnetic resonance imaging confirmed complete tumor removal. There was an initial worsening of the preoperative deficits, which progressively improved. The tumor was classified as a fourth ventricle schwannoma. There has been no evidence of tumor recurrence during the 6 years of follow-up. At present, the patient is ambulatory and reports an intermittent diplopia on conjugated gaze.

This case report intends to reveal the eighth case of a fourth ventricle schwannoma since 1957. Schwannomas of the fourth ventricle are infrequent but should be accounted in the differential

diagnosis of space-occupying lesions in this location. Gross total resection might be the definite treatment of these tumors if deemed possible <sup>7)</sup>.

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In 2013 Chen et al., reported a primary fourth ventricular solitary schwannoma case report and review of the literature <sup>8)</sup>.

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In 2009 a 71-year-old female was admitted to the hospital with an incidental finding of a ventricular tumor. Computed tomography scanning and magnetic resonance imaging revealed a solitary contrast enhancing exophytic mass lesion within the fourth ventricle. Microsurgical excision via a midline suboccipital craniotomy and tonsillo-nodular approach led to complete tumor removal. Subsequent histopathological examination confirmed the diagnosis of an unsuspected primary intraventricular cellular schwannoma. A unique case of an initially unexpected benign schwannoma arising from the fourth ventricle that could be successfully treated by microsurgery and finally confirmed by histopathological analysis with excellent patient outcome is presented. Although highly uncommon, Schwann cell tumors of both benign and malignant nature may present as ventricular lesions and should be included as a differential diagnosis in patients with either solely intraventricular masses or intra- and extraaxial tumors with extension to the ventricles <sup>9)</sup>.

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In 2012 Kachhara et al., described a very unusual case of schwannomas originating in lateral recess of the fourth ventricle. Tumor was completely excised micro-surgically via midline suboccipital craniectomy and C1 laminectomy. Dissection of the surgical specimen revealed that the tumor was completely free from surrounding structures and just hanging in the fourth ventricle. It was not attached to any cranial nerves, brain parenchyma, and blood vessel or to the dura mater. Histopathological examination confirmed the diagnosis of schwannoma. No such case has been reported so far from this extremely rare location. Relevant literature is reviewed and hypothesis for ectopic location of these tumors has been highlighted <sup>10)</sup>.

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Hodges et al., reported in 2011 the sixth reported case in the literature of fourth ventricular schwannoma. The etiology and natural history of intraventricular schwannomas is not well understood. A thorough review of potential etiopathogenic mechanisms is provided in this case report.

A 69-year-old man presented with an incidentally found fourth ventricular tumor during an evaluation for generalized weakness, gait instability, and memory disturbance. Magnetic resonance imaging (MRI) revealed a heterogeneously enhancing lesion in the fourth ventricle. A suboccipital craniotomy was performed to resect the lesion. Histopathological examination confirmed the diagnosis of schwannoma (WHO grade I).

Schwannomas should be considered in the differential diagnosis of intraventricular tumors. Although the embryologic origins may be different from nerve sheath-derived schwannomas, the histologic, clinical, and natural history appear identical and thus should be managed similarly <sup>11)</sup>.

In 2002 Mastache et al., published a 36/F Headache, ataxic gait 9 months operated by a Midline suboccipital craniectomy 4x3.8 cm Unimproved, follow-up not available <sup>12)</sup>.

In 1993 two cases of cystic brainstem schwannomas protruding into the fourth ventricle were described. Both patients presented with hemifacial spasm. While there is only one previous case report of an intraventricular brainstem schwannoma, there has been no prior description in the literature of hemifacial spasm associated with such a tumor. The clinical, radiographic, surgical, and histopathological features of these tumors are elaborated. The relationship of hemifacial spasm to the various putative theories of origin proposed for intraparenchymal schwannomas is discussed <sup>13)</sup>.

In 1990 Redekop et al., described a schwannoma arising from the dorsum of the pontomedullary junction and presenting as an exophytic mass in the fourth ventricle. The presenting clinical and radiographic features and the pathology of this tumor are summarized, and an explanation is sought for its unusual location <sup>14)</sup>.

<sup>1)</sup>

MARCHAND L, KOECHLIN P, RACINE Y. [Malignant neurinoma of the fourth ventricle with intrabulbar propagation in a schizophrenic; death during electroshock]. *Ann Med Psychol (Paris)*. 1957 Jun;115(1):108-13. French. PubMed PMID: 13444909.

<sup>2)</sup> <sup>7)</sup>

Santos MM, Timóteo Â, Coiteiro D, Pimentel J. Schwannoma of the Fourth Ventricle: The Eighth Case Report. *World Neurosurg*. 2015 Nov;84(5):1493.e9-13. doi: 10.1016/j.wneu.2015.04.036. Epub 2015 Apr 25. PubMed PMID: 25920574.

<sup>3)</sup> <sup>11)</sup>

Hodges TR, Karikari IO, Nimjee SM, Tibaleka J, Cummings TJ, Radhakrishnan S, Friedman AH. Fourth ventricular schwannoma: identical clinicopathologic features as schwann cell-derived schwannoma with unique etiopathologic origins. *Case Rep Med*. 2011;2011:165954. doi: 10.1155/2011/165954. Epub 2011 Dec 13. PubMed PMID: 22194753; PubMed Central PMCID: PMC3238494.

<sup>4)</sup> <sup>6)</sup>

Moscote-Salazar LR, Satyarthee GD, Farid-Escorcía H, Calderon-Miranda WG, Padilla-Zambrano HS, Lee A, Pacheco-Hernandez A, Agrawal A. Intra-fourth Ventricular Schwannoma in Pediatric Age Group: Report of Second Case in the Western Literature with Review of Literature. *J Pediatr Neurosci*. 2017 Oct-Dec;12(4):371-373. doi: 10.4103/jpn.JPN\_106\_17. PubMed PMID: 29675081; PubMed Central PMCID: PMC5890562.

<sup>5)</sup>

Zhu T, Chen M, Xu M, Chen D, Xu J, Yang L, Zhong P. Schwannoma of the Fourth Ventricle: Report of two cases and Review of Literature. *World Neurosurg*. 2018 Jun 12. pii: S1878-8750(18)31240-3. doi: 10.1016/j.wneu.2018.06.025. [Epub ahead of print] PubMed PMID: 29906577.

<sup>8)</sup>

Chen LH, Zhang HT, Xu RX, Wei Q, Li YJ, Li WD, Zhao H. Primary fourth ventricular solitary schwannoma: case report and review of the literature. *Neurol India*. 2013 May-Jun;61(3):330-2. doi: 10.4103/0028-3886.115099. Review. PubMed PMID: 23860172.

<sup>9)</sup>

Oertel MF, Nolte KW, Blaum M, Weis J, Gillsbach JM, Korinth MC. Primary intraventricular schwannomas. *Clin Neurol Neurosurg*. 2009 Nov;111(9):768-73. doi: 10.1016/j.clineuro.2009.06.009. Epub 2009 Jul 25. PubMed PMID: 19632768.

10)

Kachhara R, Raje P, Pauranik A. Schwannoma originating in lateral recess of the fourth ventricle. Asian J Neurosurg. 2012 Jul;7(3):151-3. doi: 10.4103/1793-5482.103728. PubMed PMID: 23293673; PubMed Central PMCID: PMC3532764.

12)

Mastache J, Rodriguez G, Garcia R, et al. Schwannoma of the fourth ventricle. Case description and literature review. Rev Med IMSS 2002;40: 405-408.

13)

Weiner HL, Zagzag D, Babu R, Weinreb HJ, Ransohoff J. Schwannoma of the fourth ventricle presenting with hemifacial spasm. A report of two cases. J Neurooncol. 1993 Jan;15(1):37-43. Review. PubMed PMID: 8455061.

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Redekop G, Elisevich K, Gilbert J. Fourth ventricular schwannoma. Case report. J Neurosurg. 1990 Nov;73(5):777-81. PubMed PMID: 2213169.

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