A 21-YEAR-OLD FEMALE WITH A THIRD VENTRICULAR TUMOR

Contributed by: A. Martina Messing-Jünger, MD[1]; Markus J. Riemenschneider, MD[2]; Guido Reifenberger, MD, PhD[2]

[1]Neurosurgical Clinic, and [2]Department of Neuropathology, Heinrich-Heine-University, Düsseldorf, Germany.

CLINICAL HISTORY

A previously healthy 21-year-old female presented with permanent tinnitus, vertigo and nausea lasting for 2 months. Physical examination did not reveal any neurological deficits. Her family history was negative. All blood tests, including serum levels of the germ cell tumor markers alpha-fetoprotein (1.1 µg/l) and beta-human chorion gonadotropin (<2 mIU/ml), were normal. Magnetic resonance imaging (MRI) of the brain demonstrated a small, partially cystic, contrast-enhancing mass in the posterior part of the third ventricle. Figure 1 shows the tumor (arrow) on a contrast-enhanced T1-weighted sagittal MRI scan. Figure 2 demonstrates a coronal section through the tumor. The tumor did not result in an obstruction of the cerebrospinal fluid flow. No further intracranial lesions were present. We additionally performed a digital subtraction angiography, which revealed a variant of the vein of Galen but no pathological vascularization of the intraventricular mass. The tumor was resected via an infratentorial supracerebellar approach. The postoperative course was uneventful.

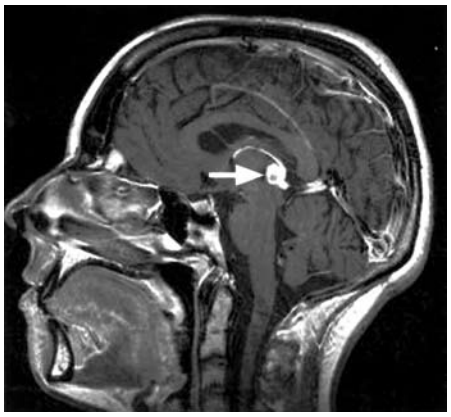

Figure 1.

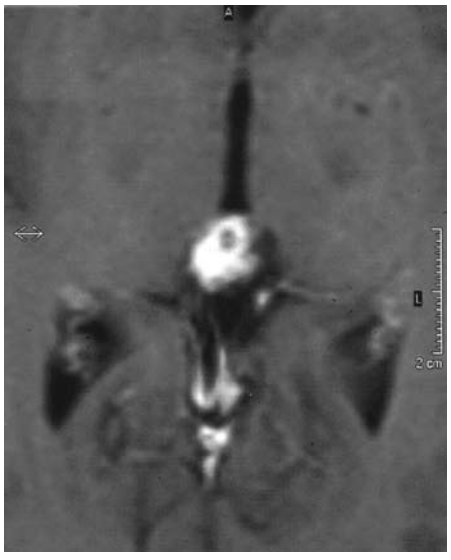

Figure 2.

MICROSCOPIC DESCRIPTION

Histological investigation showed a moderately cellular tumor that consisted of spindle-shaped tumor cells with elongated-ovoid nuclei and long, wavy processes (Figure 3). Mitoses and necroses were absent. The tumor matrix was densely reticulin rich (Figure 4). Myelinated nerve fibers were not detected. Focally, the tumor tissue involved the plexus choroideus and contained conchoidal calcifications of the type seen in plexus. Immunohistochemistry revealed a strong and diffuse expression of protein S-100 (Figure 5). Immunostains for glial fibrillary acidic protein and epithelial membrane antigen were negative. The Ki-67 (MIB-1) labeling index was low (<1%).

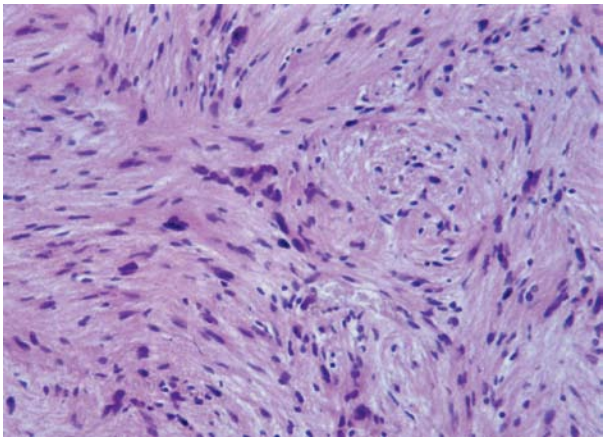

Figure 3.

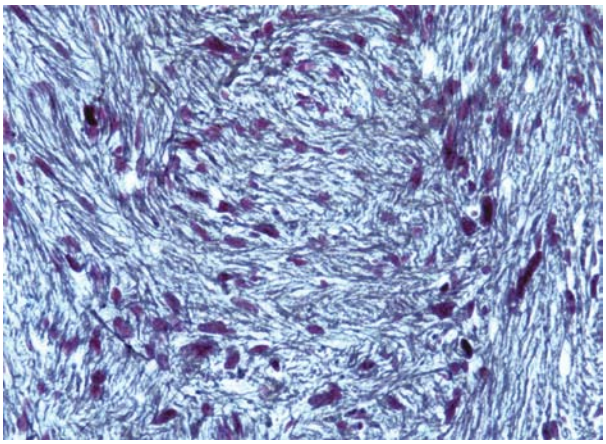

Figure 4.

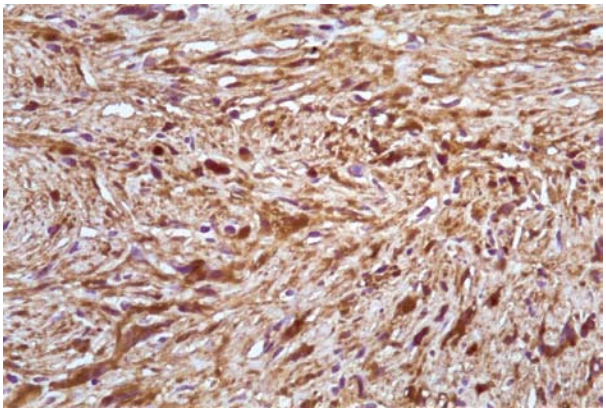

Figure 5.

DIAGNOSIS

Intraventricular schwannoma (WHO grade I).

DISCUSSION

Schwannomas account for up to 8% of intracranial neoplasms (14). The vast majority of cases arise from the cranial nerves, with the eighth nerve being most commonly involved (14). Rare cases of schwannomas may develop as primarily meningeal, intracerebral or intraventricular tumors (14). In a review of the literature, we found that only few cases of intraventricular Schwann cell tumors have been reported so far (1, 4-9, 12, 13). These tumors were generally located in the lateral or fourth ventricles. To our knowledge, the tumor of our patient is the first reported schwannoma of the third ventricle.

The origin of intraventricular schwannomas is unknown. However, similar to the hypotheses about the histogenesis of spinal intramedullary (3) and intracerebral schwannomas (11), which are slightly more common than intraventricular schwannomas, different possibilities may be considered. Schwannomas may arise from the sympathetic nerve plexus surrounding blood vessels. These vascular nerves can be found around medullary vessels, in the choroid plexus and in the meninges (10). An alternative hypothesis is the origin from ectopic neural crest-derived cells, which had been displaced during embryogenesis. This hypothesis may point to a link between intraventricular schwannomas and the so-called neurocristopathies, which comprise a spectrum of dysgenetic and neoplastic disorders associated with alterations in the migration, growth and differentiation of neural crest tissue during embryogenesis (2). Prominent examples include the neurofibromatoses, Hirschsprung disease, and Waardenburg syndrome (2). However, our patient did not demonstrate any clinical signs or symptoms indicative of these diseases.

Neuroradiologically, intraventricular schwannomas cannot be differentiated with certainty from other, more common intraventricular neoplasms such as ependymal and choroid plexus tumors. Histologically, the diagnosis is straight forward, with the main differential diagnoses being pilocytic astrocytoma and fibroblastic meningioma. However, the abundance of reticulin fibers and the lack of GFAP immunoreactivity rules out a pilocytic astrocytoma, while the strong expression of protein S-100 and absence of immunoreactivity for epithelial membrane antigen excludes a fibroblastic meningioma.

In summary, we have reported on a benign schwannoma in the posterior part of the third ventricle. The origin of this tumor is unclear. Schwann cell tumors should be included as a rare differential diagnosis in patients with intraventricular tumors.

REFERENCES

1. Barbosa MD, Rebelo O, Barbosa P, Goncalves J, Fernandes R (2001) Cystic intraventricular schwannoma: case report and review of the literature. *Neurocirugia (Astur)* 12:56-60.

2. Bolande RP (1997) Neurocristopathy: its growth and development in 20 years. *Pediatr Pathol Lab Med* 17:1-25.

3. Brown KM, Dean A, Sharr MM (2002) Thoracic intramedullary schwannoma. *Neuropathol Appl Neurobiol* 28:421-424.

4. Dow GR, Hussein A, Robertson IJ (2004) Supratentorial intraventricular schwannoma. *Br J Neurosurg* 18: 561-562.

5. Erdogan E, Öngürü Ö, Bulakbasi N, Baysefer A, Gezen F, Timurkaynak E (2003) Schwannoma of the lateral ventricle: eight-year follow-up and literature review. *Minim Invas Neurosurg* 46:50-53.

6. Estrada Mastache J, Sanchez Rodriguez R, Farias Garcia R, Calderon Garciduenas AL (2002) Schwannoma del cuarto ventriculo. *Rev Med IMSS* 40:405-408.

7. Jung JM, Shin HJ, Chi JG, Park IS, Kim ES, Han JW (1995) Malignant intraventricular schwannoma.: Case report. *J Neurosurg* 82 :121-124.

8. Ost AH, Meyer R (1990) Cystic intraventricular schwannoma: a case report. *Am J Neuroradiol* 11:1262-1264.

9. Pimentel J, Tavora L, Cristina ML, Antunes JA (1988) Intraventricular schwannoma. *Childs Nerv Syst* 4:373-375.

10. Riggs HE, Clary WU (1957) A case of intramedullary sheath cell tumor of the spinal cord. Consideration of vascular nerves as a source of origin. *J Neuropathol Exp Neurol* 16:332-336.

11. Sharma MC, Karak AK, Gaikwad SB, Mahapatra AK, Mehta VS, Sudhka K (1996) Intracranial intraparenchymal schwannomas: a series of eight cases. *J Neurol Neurosurg Psychiatry* 60:200-223.

12. Takahashi Y, Sugita Y, Abe T, Yuge T, Tokutomi T, Shigemori M. (2000) Intraventricular malignant triton tumour. *Acta Neurochir (Wien)* 142:473-477.

13. Weiner HL, Zagzag D, Babu R, Weinreb HJ, Ransohoff, J (1993) Schwannoma of the fourth ventricle presenting with hemifacial spasm. A report of two cases. *J Neurooncol* 15:37-43.

14. Woodruff JM, Kourea HP, Louis DN, Scheithauer BW (2000) Schwannoma in Kleihues P, Cavanee WK (eds). *Pathology and Genetics of Tumors of the Nervous System (WHO-Classification)*, 2nd edn, ACS/Oxford University Press, USA, pp. 164-168.