Clinical Neurology and Neurosurgery 111 (2009) 768–773

Contents lists available at ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

Review

Primary intraventricular schwannomas

Markus F. Oertel[a,*], Kay W. Nolte[b], Marcus Blaum[c], Joachim Weis[b], Joachim M. Gilsbach[a], Marcus C. Korinth[a]

[a] Department of Neurosurgery, University Hospital, RWTH Aachen University, Pauwelsstrasse 30, 52074 Aachen, Germany
[b] Department of Neuropathology, University Hospital, RWTH Aachen University, Aachen, Germany
[c] Department of Neuroradiology, University Hospital, RWTH Aachen University, Aachen, Germany

ARTICLE INFO

Article history:
Received 27 July 2008
Received in revised form 5 May 2009
Accepted 22 June 2009
Available online 25 July 2009

Keywords:
Fourth ventricle
Intraventricular
Nerve sheath tumor
Review
Schwann cell
Schwannoma
Scintillating scotoma

ABSTRACT

Schwann cell tumors arising within the neuraxis and in an intraventricular location are an exceedingly rare tumor entity of the brain. The authors present the first case of a cellular intraventricular schwannoma occurring in the fourth ventricle. The pertinent literature is reviewed.

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.

1. Introduction

Intracranial schwannomas usually arise from the neurilemmal sheaths with the vestibular division of the eighth cranial nerve being most commonly involved and present as extraaxial cerebellopontine angle tumors [1–8]. Sporadic intraaxial schwannomas have been reported, including those that occur along the ventricular system [1–15]. The authors present an illustrative case of a cellular schwannoma manifesting in the fourth ventricle and review previous cases of primary intraventricular schwannomas reported in the literature.

2. Case report

A previously healthy 71-year-old female presented with a 1-week episode of transient scintillating scotomas 2 months prior to admission. Her past medical and family history was unremarkable and the general physical and neurological examination was normal.

Non-contrast enhanced cranial computed tomography (CT) scanning (Fig. 1A) disclosed a slightly hyperdense mass lesion filling the fourth ventricle with homogeneous enhancement after contrast application (Fig. 1B). Additional magnetic resonance imaging (MRI) (Fig. 2) revealed a solitary gadolinium enhancing 1.5 cm exophytic mass within the fourth ventricle.

Microsurgical excision was performed via a midline suboccipital craniotomy and tonsillo-nodular approach. At surgery, the tumor appeared as a whitish, well demarcated, moderately vascular, firm and spherical mass that could be totally removed with preservation of the surrounding neurovascular structures. The lesion originated from the roof of the fourth ventricle and extended intraventricularly.

Histopathological examination showed predominantly compact and in part hypercellular tumor tissue of Antoni A type with an incomplete fibrous capsule (Fig. 3A) and densely packed schwannoma cells often forming nuclear palisades reminiscent of classical Verocay bodies (Fig. 3B). Semithin sections revealed neoplastic Schwann cells with elongated and closely aligned cell processes lying in bundles (Fig. 3C). Reticulin pattern was pericellular especially in these areas with typical features of schwannian differentiation (Fig. 3D). Occasionally, whorls were seen consisting of tumor cells with scant cytoplasm. Psammoma bodies were absent. In between large cellular areas with compact tumor growth some loose textured tumor tissue occurred. In this minor tumor component thick-walled hyalinized vessels as well as scat-

* Corresponding author. Tel.: +49 241 8088480; fax: +49 241 8082420.
E-mail address: mfoertel@ukaachen.de (M.F. Oertel).

0303-8467/$ – see front matter © 2009 Elsevier B.V. All rights reserved.
doi:10.1016/j.clineuro.2009.06.009

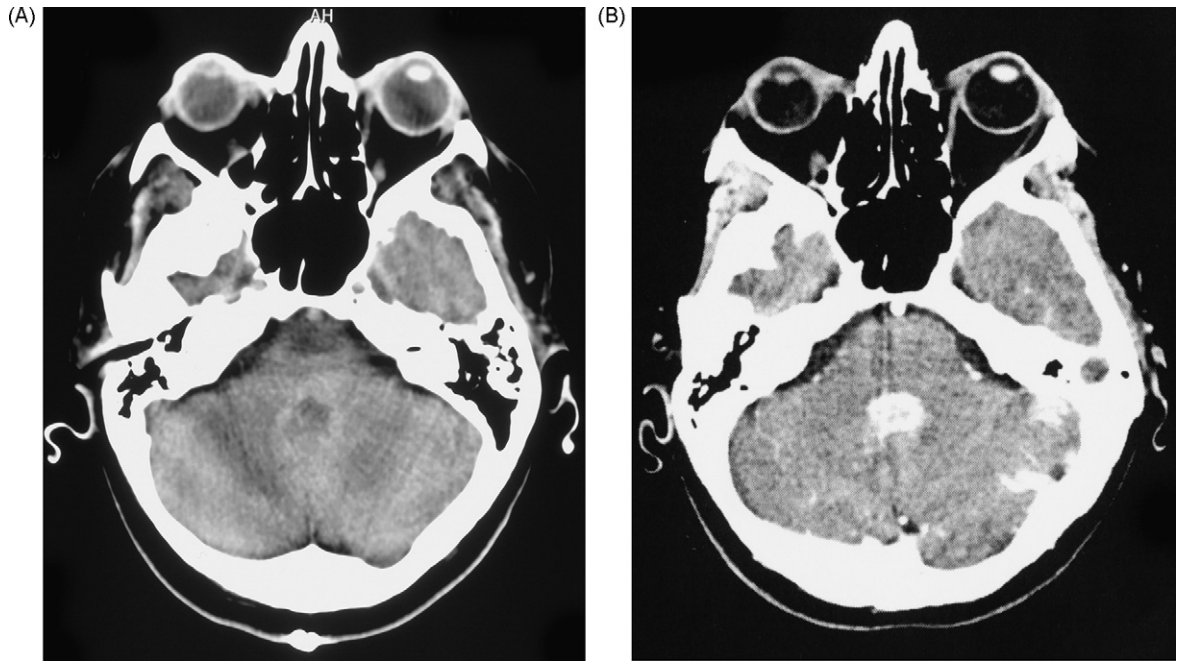

Fig. 1. Preoperative non-contrast enhanced (A) and contrast enhanced (B) axial CT scan showing a hyperdense mass lesion with marked homogeneous contrast enhancement in the fourth ventricle.

tered lymphocytes could be seen. No aggregates of foamy cells, cholesterol clefts or hemosiderin deposits were noted. Immunohistochemically, S-100 protein expression could be detected in the majority of tumor cells (Fig. 3E). GFAP reactivity was observed only at the margins of the specimen, but not within tumor tissue. Neurofilament protein-positive axons were focally encountered being intermingled with the overlying collagenous fibre bundles or subcapsular in location. Tumor cell nuclei were round to oval or spindle-shaped showing no distinct nucleoli, hyperchromasia or considerable pleomorphism. However, in few small tumor areas mitotic figures were readily identifiable and came up to 20 per 10 high-power fields (HPF). The labelling index (LI) for the proliferation marker MIB-1 was also focally increased with up to 25% of tumor cell nuclei being strongly Ki67-immunoreactive (Fig. 3F). In large parts of the tumor tissue mitotic count and MIB-1/Ki67-LI were less than 1 per 10 HPF and 3–5%, respectively. A low to moderate level of p53 protein immunoreactivity was seen in a subpopulation of tumor cells. No foci of necrosis were noted. Thus, neuropathological findings of a well differentiated, partly hypercellular peripheral nerve sheet tumor of predominantly Antoni A type exhibiting focal increase in mitotic activity led to the diagnosis of a primary intraventricular cellular schwannoma (WHO grade I).

The postoperative course was uneventful. Two and a half years later there was no evidence of recurrence.

3. Discussion

In 1957 Marchand et al. published the first case of an intraventricular Schwann cell tumor. This malignant schwannoma developed in the fourth ventricle in a 43-year-old man with schizophrenia [14]. Only 15 further cases of intraventricular schwannoma have been reported in the literature to date [1–13,15]. The current case is presented as the seventeenth one and the fifth one located in the fourth ventricle. All cases are summarized in Tables 1 and 2.

When reviewing the literature including the present case, the age distribution of patients with intraventricular schwannoma ranged from 7 [7] to 78 years [8] (average age 33.4 years). Eleven patients were males [1,2,4,6–8,10,11,13–15] and six females [3,5,8,9,12]. It is interesting that in contrast to more typically located Schwann cell tumors, in the majority of instances intraventricular situated schwannomas tend to occur earlier in life and primarily in male patients. However, the present case itself differs from the majority of cases of intraventricular schwannoma reported in that our patient was a 71-year-old female. Intraventricular schwannomas frequently present with short duration of signs and symptoms of increased intracranial pressure, focal neurological deficits and seizures [1–5,7–10,12,13,15]. The duration of symptoms varied from 2 weeks [1] to 40 years [12]. In three cases reported a schwannoma was an occasional finding [6,11,14]. The cause of the abnormal visual sensations that occurred in our case remains uncertain, but may have been related to the prodromal phase of migraine. In particular, with regard to the patient reported of preceding incidental migraine-like headache episodes, the clinical evidence strongly supports a non-tumorous origin of the transient symptomatology and it appears most likely that the scintillating scotomas as described would be consistent with visual aura of migraine without accompanied cephalgia.

The treatment of choice for intraventricular schwannoma is microsurgical excision. With the exception of the case reported by Marchand et al. describing a chance finding of a malignant schwannoma upon autopsy in a patient who died during electroshock therapy for schizophrenia [14], all patients underwent surgery usually with complete removal of the tumor. Gradual or complete relief of clinical symptomatology without surgery-associated complications and recurrence was mostly achieved after surgical resection [1–13,15]. Because the majority of schwannomas was located in the lateral ventricle, usually a transcortical approach after osteoplastic craniotomy was performed for tumor removal [1,2,4,6,9–13,15]. In cases of third or fourth ventricle schwannoma, a midline suboccipital access was applied [3,5,7,8]. With the exception of a patient with malignant schwannoma and tumor recurrence with drop metastasis treated by whole-brain radiation and second operation for recurrence and metastasis [13], surgical removal was curative with no need for further adjuvant therapy. Rarely, occlusive hydrocephalus and sequestration of ventricular compartments may