INTRACEREBRAL EPIDERMOID TUMOR:
PATHOGENESIS OF INTRAPARENCHYMAL
LOCATION AND MAGNETIC RESONANCE IMAGING
FINDINGS

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Epidermoid tumors are rare slow-growing lesions, accounting for 0.3% to 1.8% of all primary intracranial neoplasms [3]. Common locations include the cerebellum and parietal region (30%), and more rarely the middle cranial fossa, diploic or spinal canal [3, 5]. Purely intraparenchymal epidermoid tumors are rarely described. The pathogenesis of such a location is still not clear.

A 40-year-old man was admitted after a sudden episode of convulsive seizures followed by a prolonged loss of consciousness. At examination, he was perfectly awake without any neurologic deficit.

Enhanced computed tomography (CT) showed a rounded mass on T2 weighted MR imaging (figures 2a and 2b). Gadolinium enhanced MR images showed slight ring enhancement. Analyzing the internal structure of the lesion, we found a ring-like structure isointense to the brain parenchyma on T1 and T2 sequences. Coronal images showed the close relationship of the cyst to the inner table of the anterior skull base. At this point, 4 main diagnoses were suggested pre-operatively: cystic low grade glioma, inclusion tumor, arachnoid cyst, and hydatid cyst.

The patient underwent a left fronto-pterional craniotomy. After opening the dura, microscopic dissection discovered an inferior frontal yellowish tumor covered by a thick arachnoid membrane, within the left rectus gyrus. This arachnoid membrane was closely adherent to the dura of the anterior skull base. After opening of this arachnoid membrane, white keratinized material was progressively removed and the cyst capsule was carefully separated from the surrounding gliotic brain parenchyma. The tumor and its capsule were totally removed. The histological examination confirmed a diagnosis of epidermoid cyst. The postoperative course was uneventful. The one year follow-up examination revealed no deficit and the patient is anti-seizure medication-free.

Epidermoid cysts, also known as pearly tumors or cholesteatomas, are uncommon congenital lesions, representing between 0.3% and 1.8% of all intracranial tumors [3]. These histologically benign tumors arise from the sequestration of neuroectodermal epithelial cells during neural tube closure. Intraparenchymal location is very rare, accounting for less than 2% of all intracranial epidermoids [1, 3, 4].

In reviewing the literature, we found no more than 40 reported cases of intracerebral epidermoid cysts [4]. Most of these cases were reported before the MRI era. Involvement of the frontal and temporal regions was most frequently reported (about 80% of cases).

Concerning the pathogenesis of purely intraparenchymal epidermoid tumors, many theories have been suggested. In 1978, Rengachary et al. [5] stated that epidermoids resulted from congenital sequestration of neuroectodermal elements occurring between the third and fifth weeks of intrauterine life. Kaido et al. [4] summarized their hypothesis explaining the development of intracranial epidermoid cysts. Thus, as the sequestration of ectodermal elements would occur during the development of primary cerebral vesicle (3rd week of embryogenesis) or secondary cerebral vesicle (4th and 5th week of intrauterine life), epidermoid cysts can involve intracerebral and intraventricular regions or the cerebellopontine angle respectively. Finally, during the closure of the neural tube, rests of ectodermal cells could be left either on the inner, the outer surface or within the neural tube, leading to this variability in epidermoid tumor location [2].

Usually, intraparenchymal epidermoid cysts are homogeneous nonenhancing hypodense lesions on CT, often without surrounding edema. Calcifications or slight ring enhancement may be found. Differential diagnosis includes cystic gliomas, dermoid and arachnoid cysts, and hydatid cysts. MR Imaging allows more accurate diagnosis, particularly with the use of fluid attenuated inversion recovery (FLAIR), constructive interference in steady state (CISS) sequences and diffusion-weighted imaging (DWI). A majority of epidermoids display hypointensity or isointensity on T1 weighted sequences and hyperintensity on T2 images. Slight ring enhancement may also be found. Nevertheless, most intraparenchymal epidermoid cysts have nonspecific findings on MRI [1, 4].

Surgical management is not different from that used for epidermoids in other locations. Indeed, those tumors may have a thick tumor capsule very adherent to the surrounding brain tissue. Total removal is preferred when possible in order to prevent late recurrences; however, it is acceptable to leave a part of the tumor capsule especially in eloquent locations.
REFERENCES


MALIGNANT OCULOMOTOR SCHWANNOMA: DIFFUSION MR IMAGING

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Malignant peripheral nerve sheath tumors involving the cranial nerves are very rare [7-11]. These malignant schwannomas have been studied by MR imaging, however, in the literature there is no previous report on their diffusion MR imaging features, except for a study on benign vestibular schwannomas [12].

The patient, a 15 month-old boy, presented with left-sided ptosis. Neurologic examination revealed left oculomotor nerve paralysis. Laboratory testing did not reveal any abnormality. There was no evidence of neurofibromatosis. On MR imaging, a large mass was seen originating from the left oculomotor nucleus and extending along the oculomotor nerve. The nerve was thickened up to 3 cm (figure 1a). On the b=1000 sec/mm^2 images diffusion MR imaging sequence (TR=5700; TE=139 msec) it was noted that the tumor had hyperintense signal (figure 1b). ADC
values obtained from the tumor ranged between 0.45 and 0.60 × 10^{-3} \text{mm}^2/\text{sec}. These values were lower than the normal brain parenchyma regions (ranging between 0.76-0.98 × 10^{-3} \text{mm}^2/\text{sec}) (figure 1c). Stereotactic biopsy was done, and histopathologic examination of the biopsy material revealed the features of malignant nerve sheath tumor. The tumor was removed in a subsequent operation. Chemotherapy was given. There was no evidence of recurrence, 4.5 years postoperatively, except for meningeal thickening at the left side of the brainstem (figure 2).

Previously reported malignant cranial nerve schwannomas have included vestibular, trigeminal, optic, trochlear, abducens, facial, and oculomotor schwannomas [7-11]. Isolated oculomotor nerve involvement in the absence of neurofibromatosis appears to be extremely rare, as seen in the current patient. A recent report dealt with diffusion MR imaging of vestibular schwannomas [12]. That study included six patients with benign solid vestibular schwannomas. It was reported that those benign schwannomas appeared isointense on b=1000 sec/mm² images, compared with the normal brain parenchyma. At the same time ADC values were high (1.42±0.17 × 10^{-3} \text{mm}^2/\text{sec}) compared with that of normal brain parenchyma (0.80±0.11 × 10^{-3} \text{mm}^2/\text{sec}). It was concluded that high ADC values were consistent with increased amounts of extracellular water, representing presence of relatively loose tumor tissue, and it was suggested that diffusion MR imaging findings could aid in differentiating vestibular schwannomas at least from other tumors with a high nuclear-to-cytoplasmic ratio [12]. In the present patient with malignant oculomotor schwannoma, the diffusion MR imaging pattern was the opposite of the pattern reported in benign vestibular schwannomas. In this malignant oculomotor schwannoma the tumor was hyperintense on b=1000 sec/mm² images, and the ADC values were low (ranging between 0.45 and 0.60 × 10^{-3} \text{mm}^2/\text{sec}) compared to the isointensity, and high ADC values of benign schwannomas. This suggested that diffusion MR imaging could aid in differentiating malignant from benign schwannomas.
REFERENCES


ENDOVASCULAR TREATMENT OF RUPTURED INTRACRANIAL ANEURYSMS: IMMEDIATE RESULT AND LONG TERM FOLLOW UP

An indian experience

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Intracranial aneurysms have a prevalence of 0.5% to 6% in adults and the most dreaded complication of intracranial aneurysm is rupture causing subarachnoid hemorrhage with mortality rate of 32-67%. Endovascular treatment of ruptured intracranial aneurysms is being used increasingly and has evolved as an alternative to surgical clipping [1-3]. Rebleeding occurred in 2.6% of patients who underwent coiling or attempted coiling and in 1.0% of those who underwent surgery or attempted surgery [4].

We tried to assess retrospectively the immediate angiographic result and a long-term follow up to assess the rebleed rate. A total of 27 ruptured intracranial aneurysms were coiled. On post procedure angiograms, small sized aneurysms (<10 mm dome diameter) showed complete occlusion in 15/19 (78.94%) cases with stable occlusion noted in 17/19 (89.47%) of cases and progressive thrombosis seen in 1/19 (5.26%) case on follow up angiograms at 6 months and 1 year. In large sized aneurysms (10-25 mm), complete occlusion was noted in only 4/8 (50%) cases on immediate post procedure angiograms at 6 months and 1 year. In large sized aneurysms (10-25 mm), complete occlusion was noted in only 4/8 (50%) cases on immediate post procedure angiograms with near complete occlusion seen in 1/8 (12.5%) case (figure 1). On follow up angiogram at 1 year, stable unchanged result was seen in only 4/8 (50%) cases with recanalization seen in 4/8 (50%) cases. Thus, the recanalization rate was much higher for large sized aneurysm (50.0%) than for small sized aneurysm (15.78%). In case of narrow necked aneurysms (neck <4 mm), complete and near complete occlusion was seen in 15/18 (83.33%) cases (figure 2) with 16 cases (88.88%) showing stable status and 2 (11.11%) showing recanalization on follow up angiogram at 1 year. Wide necked aneurysms showed complete and near complete occlusion in 4/9 (44.44%) cases with progressive thrombosis in 2 cases (22.22%) (figure 3) and recanalizations in 3 cases (33.33%) on follow up imaging. Aneurysm with wide neck showed slightly higher rate of recanalization (33.33%) compared to narrow necked aneurysms (11.11%). Of the 27 cases of intracranial aneurysms that underwent selective aneurysm coiling, good immediate outcome (Glasgow outcome scale 1 and 2) was seen in 24/27 (88.88%) cases with poor clinical outcome in the remaining 3/27 (11.11%) cases. The patients presenting with Grade 1 SAH had much better immediate clinical outcome (14/15) as compared to those presenting in Grade 2 or Grade 3 SAH (10/12) with good outcome in 93.33% and 83.33% cases respectively. Long-term follow up was available in 19/25 cases with good outcome (Glasgow outcome scale 1 and 2) was seen in 24/27 (88.88%) cases with poor clinical outcome in the remaining 3/27 (11.11%) cases. The patients presenting with Grade 1 SAH had much better immediate clinical outcome (14/15) as compared to those presenting in Grade 2 or Grade 3 SAH (10/12) with good outcome in 93.33% and 83.33% cases respectively. Long-term follow up was available in 19/25 cases with good outcome (Glasgow outcome scale 1 and 2) was seen in 24/27 (88.88%) cases and poor outcome in 2 cases. No rebleed was seen in any of the treated cases.

Endovascular treatment of intracranial aneurysms is a safe and effective treatment modality that offers protection from recurrent subarachnoid hemorrhage. With expertise, rebleed can be prevented contrary to what has been stated in other series [4].
FIG. 1. — (a) Left internal carotid artery injection shows a giant aneurysm with wide neck. (b) Post coiling angiogram shows complete packing and total obliteration of the aneurysm.

FIG. 1. – Anévrisme géant à large collet de l’artère carotide interne gauche : aspect avant (a) et après (b) embolisation.

FIG. 2. – Right internal carotid artery injection shows a small narrow necked middle cerebral artery bifurcation aneurysm. (b) Post coiling angiogram shows total obliteration of the aneurysm.

FIG. 2. – Anévrisme à collet étroit de l’artère cérébrale moyenne droite. Aspect avant (a) et après (b) embolisation.

FIG. 3. – (a, b) Left vertebral artery injection anteroposterior and lateral views show a wide necked basilar top aneurysm. (c, d) Post coiling angiogram shows complete packing and total obliteration of the aneurysm. This was done with balloon remodeling technique. Note total luminal preservation of both posterior cerebral arteries.

FIG. 3. – Anévrisme géant à large collet de la terminaison de l’artère basilaire : aspect avant (a, b) et après (c, d) embolisation. Utilisation de la technique de remodelling.
NEUROSARCOIDOSE RÉVÉLATRICE D’UNE SARCOIDOSE SYSTÉMIQUE

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Une femme de 49 ans présentait, six mois après une neuropathie ischémique antérieure aiguë de l’œil droit, des troubles sensitifs et moteurs des quatre membres rapidement progressifs, associés à des troubles sphinctériens ayant abouti à une paraplégie flasque. L’IRM cérébromédullaire révélait une moelle cervicale élargie et siège d’hypersignaux T2 et de rehaussements médullaire et pie-mérien étendus du foramen Magnum jusqu’en regard de C5 (figure 1). À l’étage cérébral étaient notées des zones en hypersignal T2 insulaire droite et bulbaire, une tuméfaction et un rehaussement anormal de la tige pituitaire et de la région unfundibulo-chiasmatique (figure 2). Radiographie standard, TDM thoracique et explorations respiratoires fonctionnelles étaient normales. La démonstration d’une inflammation bronchique avec au lavage bronchioloalvéolaire une hypercellularité à prédominance macrophagique et des granulomes épithéloïdes sans nécrose caseuse à la biopsie bronchique ont confirmé le diagnostic de sarcoïdose. Sous corticothérapie était notée une régression progressive des troubles neurologiques : reprise de la marche mais persistance de la cécité de l’œil droit. Le contrôle IRM à 8 mois démontrait la disparition quasi-totale du rehaussement unfundibulo-chiasmatique et médullaire, la reprise d’un volume médullaire normal et la persistance des anomalies de signal de la substance blanche.


Les aspects IRM de la neurosarcoïdose sont polymorphes posant le diagnostic différentiel avec d’autres affections. L’association d’anomalies de rehaussement leptoméningé et d’atteinte hypothalamo-hypophysaire doit faire évoquer le diagnostic. La fibroscopie bronchique, même à imagerie thoracique normale, permet de poser le diagnostic et d’éviter un abord neurochirurgical.

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THE ABNORMALLY DILATED INTERNAL AUDITORY CANAL: A NON-SPECIFIC FINDING OR A DISTINCTIVE PATHOLOGIC ENTITY¹

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Congenital malformation of the inner ear has been improved with the more widespread use of computed tomography (CT) and magnetic resonance imaging (MRI) for the evaluation of cochlear implant candidates. The prevalence of internal auditory canal (IAC) anomalies among the inner ear malformations is variable with the overall severity of the anomaly. The abnormally dilated IAC, in conjunction with known syndromes or unknown nosologic entities, is

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rarely observed in the literature [4, 5]. Eight cases with abnormally dilated IAC, associated with various vestibulocochlear anomalies are reported. We examine a possible association with specific nosologic conditions in combination with the results in the literature.

Temporal bone studies of 68 patients with profound sensorineural or mixed hearing loss and radiographic evidence demonstrating inner ear malformations were retrospectively identified. All patients underwent high-resolution CT and 47 of them underwent MRI temporal bone studies. Measurements of the IAC were obtained in millimetres with a bony algorithm CT and magnified computer images. The criterion for abnormally dilated IAC was based on a diameter greater than the established normal mean IAC width (5.31±1.11) mm [2]. Individual measurements of the abnormally dilated IACs were recorded and compared with temporal bones from patients of the same age group without inner ear deformities.

Data analysis was performed using the Student’s t-test (P<0.05).

Eight patients (3 females, 5 males) were identified with the anomaly under study. In 2 patients the IAC was abnormally dilated bilaterally, while 6 patients demonstrated the anomaly unilaterally. The mean age of the patient population at the time of evaluation was 4.0±3.1 years, (range; 7 months to 20 years). The mean width of IAC in temporal bones with IAC abnormality was 7.1±0.3 mm and in temporal bones of the control group 3.9±1.3 mm (P=0.02). All patients demonstrated an ipsilateral sensorineural hearing loss and four of them received cochlear implants. Two representative imaging illustrations of the patients with abnormally dilated IACs are shown in figures 1 and 2. Two of the three patients with CHARGE association had a dilated IAC, which was always accompanied by a Mondini type B malformation of the cochlea and vestibular abnormalities. Furthermore, the dilated IAC (in conjunction with vestibular anomalies) was found in two of fifteen cases with Goldenhar syndrome. Finally, the four patients without a recognizable diagnosis demonstrated abnormally dilated IACs in keeping with other related inner ear findings, the most striking of which was the presence of a common cavity.

The intraindividual variability in size, shape, and orientation of the IAC is small, implying a relative constancy of the volume of the contained neurovascular bundle for a particular individual. The malformed IAC may appear as follows: abnormally narrowed, containing only the VIIth nerve; “double”, with a prominent falciform crest; tapered towards the lamina cribrosa at its lateral end (often associated with common cavity and spontaneous cerebrospinal fluid fistula); and widened or balloned [3]. Moreover, a widened IAC at its lateral end can be associated with a tumour-like acoustic neuroma, a dural ectasia (neurofibromatosis), or a chronic hydrocephalus. In these cases, the dilation is secondary to the increased local pressure and not a congenital defect. Goldenhar syndrome, Apert syndrome, Patau syndrome and CHARGE association as well as non-classifiable congenital malformations (i.e. after congenital cytomegalovirus infection) can appear with dilatation of the IAC and defective fundus (resulting in a high incidence of a profuse anomalous flow of perilymph (gusher) upon intraoperative stapes manipulation) [1]. The IAC may be normal in the presence of other inner ear malformations (frequently Mondini malformations) and vice versa. The distinctive embryological development of the IAC from that of the labyrinth may be a possible reason. Although a narrow IAC is assumed to be caused by an abnormality in the end organs of the labyrinth, which result in aplasia or hypoplasia of the eighth nerve, there is no adequate embryological explana-

Fig. 1. – Axial high-resolution CT scan of the temporal bone of a two-year-old male patient with Goldenhar syndrome and sensorineural hearing loss on the left side. Note the tapered internal auditory canal (white arrow) and the enlarged vestibule on the left side (black arrow).

Fig. 1. – Coupe scanographique axiale, haute résolution du rocher gauche. Enfant de 2 ans atteint d’un syndrome de Goldenhar avec surdité gauche. Aspect effilé du CAI (flèche blanche) et vestibule élargi (flèche noire).

Fig. 2. – Axial MRI scan of the temporal bone of an one-year old male patient with sensorineural hearing loss bilaterally. A specific syndrome was not diagnosed after radiological, clinical, and genetic studies. Note the ballooned internal auditory canals bilaterally. The cochleovestibular nerve bundle is bilaterally intact.

Fig. 2. – IRM, coupe axiale. Enfant âgé d’un an, surdité bilatérale. Aucun diagnostic retrouvé après les études radiologique, clinique et génétique. Noter l’aspect ballonné des CAI. Les nerfs cochleovestibulaires sont ints.
tion for the abnormally dilated IAC. The abnormally dilated IAC in our patients did not seem to be specific for a unique nosologic condition, either. Remarkably, in all of our patients the abnormal IAC was present with cochleovestibular malformations, while a stapes gusher was presented in 3 cases during the operation. Our findings are consistent with reported observations, where the dilated IAC is encountered in the common cavity and cochleovestibular hypoplasia [4]. According to this report, a fundus defect is a fairly common finding, which is not always present with stapes gusher. Radiologists and otolaryngologists should be aware of the IAC malformations and their possible complications (spontaneous CSF fistula, CSF leak upon cochlotoomy, recurrent meningitis). Rarely, the enlarged IAC can appear as an isolated radiographic feature without other ear malformations. Thus, in the presence of an abnormally dilated IAC, a careful study of its fundus and documentation of the intact bony partition between the IAC and the cochlea is crucial.

REFERENCES


COMPARISON OF MULTIDETECTOR ROW CT CROSS-SECTIONAL SOURCE IMAGES WITH MULTIPLANAR 2D-, 3D- RECONSTRUCTIONS AND VIRTUAL ENDOSCOPY IN ASSESSMENT OF THE MIDDLE EAR

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Owing to the miniature size and complexity, the anatomy of the middle ear is difficult to comprehend by two dimensional images alone. A large number of slices generated in MDCT examination has to be mentally integrated to reach a three-dimensional understanding of the anatomy. Therefore multiplanar and 3D-rendering of MDCT data of the petrous bone may be expected to provide detailed and confident information concerning the middle ear cavity and its structures together with endoluminal views of satisfying image quality, competing with transtympanic endoscopy.

Evaluation and comparison of CT axial images with 3D reformatations (virtual endoscopy and volume rendering) (figure 1) and 2D multiplanar reconstructions (figure 2) in the assessment of the middle ear in different pathological conditions.

80 patients were examined over a 20-months period (October 2002 – May 2004) with the initial diagnosis of chronic otitis media, otosclerosis, trauma and tumors. CT examination (GE Light Speed Ultra Advantage 8-row) of the petrous bone was obtained using 1.25 mm thick slices with 0.62 mm slice overlap, 120 kV, 140-170 mAs and 0.8 s scan time. Images were reconstructed with bone algorithm and a small field of view FOV=9.6 cm, separately for the right and left petrous bones. Images were then transferred on a separate workstation and processed with the Reformat tools and Navigator virtual endoscopic software (General Electric, Advantage Workstation 4.2). A threshold value ranging -350 to -600 HU was applied.

Evaluation of eight different groups of features was performed, separately for axial images, two-dimensional (multiplanar and curved reformatations) and three-dimensional reconstructions with virtual endoscopy.

In patients with middle ear inflammation, pathology was best visible on multiplanar and curved reconstructions (two-dimensional).

In otosclerosis, foci of osteopetrosis were best visualized on axial images. Stapes prosthesis fixation and exact positioning were evaluated with virtual endoscopy.

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In case of temporal bone trauma, all lesions were best assessed on multiplanar reconstructions. The only exception was disruption of the ossicular chain, visualized with best accuracy on virtual endoscopy.

In case of tumors, multiplanar and curved reconstructions were most useful. Volume rendering was of value for highly vascularized tumors (angio-CT evaluation).

Middle ear virtual endoscopy is useful in evaluating post-traumatic, post-operative changes and congenital malformations of the ossicular chain.

Multiplanar reformations show anatomical features and pathologic conditions more precisely than axial images in a majority of cases.

The otospongiotic foci in otosclerosis are best visualized on axial images.

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REFERENCES


La localisation atypique d’une tumeur fibreuse solitaire dans le quatrième ventricule

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La tumeur fibreuse solitaire (TFS) est une tumeur bénigne fibreuse d’origine mésenchymateuse. Initialement rapportée au niveau de la plèvre [7], elle peut toucher de nombreux autres organes [2]. Les localisations cérébrales, généralement extra-axiales à base d’implantation durale, sont rares [1]. Nous rapportons une localisation très atypique de TFS dans le quatrième ventricule (V4).

Une patiente de 32 ans, sans antécédent particulier, a été hospitalisée dans notre institution pour la prise en charge d’une lésion du V4 découverte devant des céphalées. La patiente décrivait depuis 6 mois des céphalées matinales, bitemporales, associée à des sensations vertigineuses positionnelles, des acouphènes, un flou visuel et des nucalgies.

L’examen clinique montrait une diplopie dans le regard horizontal gauche. Le reste de l’examen neurologique était normal. Un scanner (figure 1), et une IRM (figure 2) révélaient une masse arrondie, bien limitée, de 2,5 cm de diamètre, située dans le V4, prenant le contraste de manière intense et homogène.

L’intervention neurochirurgicale permit d’accéder à une tumeur jaunâtre, ferme, non adhérente aux structures adjacentes. L’examen anatomopathologique montrait des cellules fusiformes à noyau ovaire allongé, avec la présence de collagène abondant, distribué en rubans intercellulaires et une absence de mitose, d’atypie cellulaire ou de zone de nécrose. L’examen immuno-histochimique mettait en évidence la présence de marqueurs CD 34 +, BCl 2 + et de vimentine + (figure 3). Les cellules tumorales étaient en revanche EMA-négatives, cytokeratine-négatives. Ces données anatomopathologiques et immuno-histochimiques étaient évocatrices d’une TFS.

La nature exacte de la TFS reste encore sujette à débat parmi les anatomopathologistes. Récemment, une nouvelle classification a été élaborée pour subdiviser en trois catégories le groupe hétérogène des lésions qui étaient avant étiquetées hémangiopéricytome (HPC) [3]. La première comprend les tumeurs non-HPC présentant des caractéristiques histologiques aspécifiques d’HPC. La seconde correspond aux HPC vraies. Enfin, la dernière regroupe la TFS ainsi que les lésions apparentées.

Seules l’anatomopathologie, avec visualisation de cellules fusiformes à noyau allongé, et l’immuno-histochimie, avec la présence de cellules CD 34 +, Vimentine +, S-100 B - et négatives pour l’antigène de membrane épithéiale (EMA), permettent un diagnostic de certitude de TFS. La forte positivité des cellules à l’antigène CD 34 permet de distinguer la TFS de l’HPC, où celle-ci est plus faible [3].

La localisation intraventriculaire des TFS cérébrales est exceptionnelle avec seulement trois cas rapportés dans le V4 [9, 15, [6]. L’hypothèse avancée pour expliquer cette topographie inhabituelle, ainsi que d’exceptionnelles localisations intraparenchymateuses, serait que les cellules tumorales auraient pour origine le mésenchyme satellite de la microvascularisation cérébrale et non des méninges [6].

En scanner, la TFS apparaît généralement isodense au parenchyme cérébral. Elle présente un hyposignal T1 et un hypersignal T2 hétérogène en IRM. Au sein de la tumeur, des zones en hyposignal T2, évocatrices du diagnostic, correspondent à des bandes de collagène. En scanner comme en IRM, elle présente un rehaussement intense et homogène. Cet aspect en imagerie est toutefois aspécifique et ne permet pas de distinguer formellement la TFS d’un méningiome ou d’un HPC [5]. Dans le V4, les principaux diagnostics différentiels de la TFS sont le méulloblastome, mais l’âge de survenue est en général plus précoce, et l’épendymome, qui présente habituellement un aspect plus hétérogène avec des calcifications et des portions hémorragiques.

Notre observation présente une localisation très rare de TFS intra-crânienne. Il incite, en dépit de la rareté de cette tumeur, à inclure cette lésion dans la gamme diagnostique des tumeurs du V4.

**FIG. 1.** Scanner cérébral montrant une masse arrondie du quatrième ventricule présentant une prise de contraste intense et homogène.
RÉFÉRENCES


