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# Intraventricular Noncolloid Neuroepithelial Cysts

Paul F. J. New<sup>1</sup> Kenneth R. Davis<sup>1</sup> Symptomatic intraventricular developmental cysts of neuroepithelial origin occurring in the lateral and fourth ventricles are extremely rare. We report the clinical, radiologic, and computed tomographic (CT) findings in three such cases, two occurring in the lateral ventricles and one in the fourth ventricle. Within the lateral ventricles, these cysts are distinguished from colloid cysts by their large size, more posterior locations, and CSF-equivalent CT attenuation of their contents. Lack of contrast enhancement on CT and angiographic avascularity are also features. Fenestration in two cases and partial cyst excision in the third case relieved the predominant symptoms. The symptomatology and theories of histogenesis are reviewed and the differential diagnosis discussed.

The radiologic findings in three exceptional symptomatic intraventricular thin-walled cysts were presented by us in 1978 [1, 2]. In two instances, the cysts occupied the posterior part of a lateral ventricle. One of these was proven to be a neuroepithelial cyst. The second, in which surgical proof was not then available, was presumed also to be a neuroepithelial cyst, owing to the remarkable resemblance of the computed tomographic (CT) appearance to that in the first case. The third involved the fourth ventricle, and histologic examination revealed a glial-ependymal cyst. Search of the literature has revealed only six reports describing similar cysts in the lateral ventricles. [3–6].

More recently, a large choroid plexus cyst (arising from the choroid plexus in the body of a lateral ventricle) was reported [7], and a case of arachnoid cyst occurring in a lateral ventricle was reported by Yeates and Enzmann [8]. The latter authors found no similar case in the literature. The remarkable resemblance of the CT appearance of Yeates and Enzmann's case to our cases and the recent surgical confirmation of our second lateral ventricular cyst have prompted this publication. Only five examples of symptomatic neuroepithelial cysts of the fourth ventricle have been found in the literature, to which we add another case.

#### **Case Reports**

Case 1: Lateral Ventricular Neuroepithelial Cyst

A 32-year-old man was admitted for investigation of possible temporal lobe seizures. He had been observed to suffer from spells of automatic behavior, often associated with violence, and total amnesia for the episodes. He complained of acute paroxysmal headaches, some of which were said to be followed by the spells of automatism. On neurologic examination, he was alert, oriented, and coherent, without abnormal neurologic findings. Two electroencephalographic examinations, including sleep studies, were normal. A plain film skull examination and a technetium pertechnetate radionuclide scan were normal.

A CT scan (EMI Mark I,  $80 \times 80$  matrix) revealed a localized expansion of the trigone and body of one lateral ventricle (fig. 1A). The occipital and temporal horns of this ventricle were moderately dilated. The medial wall of the posterior part of the body of the ventricle

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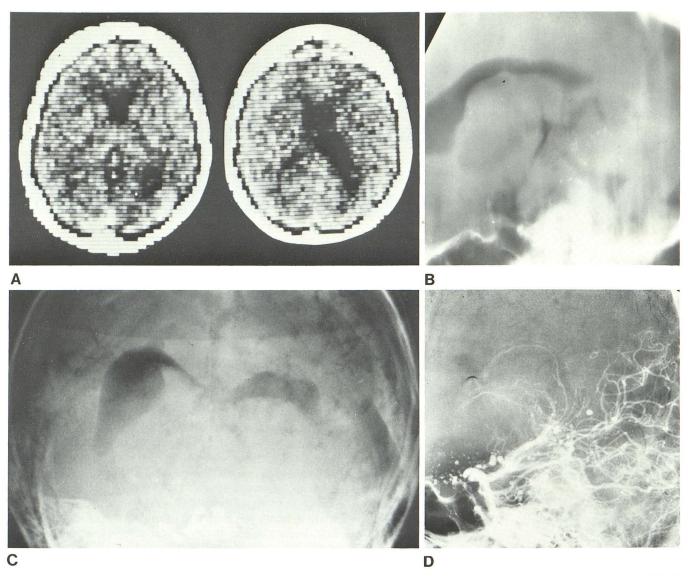


Fig. 1.—Case 1: Lateral ventricular neuroepithelial cyst. A, CT scan, 80  $\times$  80 matrix plain scan. Body and trigone of left lateral ventricle are expanded. Part of membrane separating ventricle from cyst is visible anteriorly and there is hint of another separating membrane in region of anterior part of occipital horn (arrows). Left occipital horn and, in lower sections, left temporal horn were enlarged, whereas frontal horn and entire right lateral ventricle were of

normal size. Attenuation within cyst was same as that of ventricular CSF. B, Pneumoencephalogram, lateral view in erect position. Smoothly lobulated mass projects upward into body and trigone of left lateral ventricle from floor. C, Frontal half axial view. Same findings as B. D, Vertebrobasilar angiogram, lateral view. Marked elevation of lateral posterior choroidal artery by avascular mass.

bulged across the midline.

Pneumoencephalography with tomography revealed a lobulated mass, projecting from the floor into the body and trigone of the left lateral ventricle (figs. 1B and 1C). Transfemoral carotid and left vertebral angiography revealed an avascular mass in the left thalamic area (fig. 1D).

A parietooccipital craniotomy with ventricular exploration revealed a large, very thin-walled, bluish cyst almost filling the ventricle in the region of the trigone and posterior body and attached to the ventricular floor. Clear serous fluid was aspirated. No abnormal vessels or solid abnormal tissue were identified and the cyst, which was firmly attached to the floor of the ventricle, was fenestrated. Histologic examination revealed a simple epithelial cyst. One year later the patient had no headaches or evidence of organic neurlogic disease.

#### Case 2: Lateral Ventricular Neuroepithelial Cyst

A 28-year-old woman awoke one morning with severe bifrontal throbbing headache and nausea. The headache was constant for 3 days, when she then developed episodic double vision, slurred speech, and right arm weakness. There was also gait imbalance. CT suggested a cyst in the left lateral ventricle. All symptoms except headaches resolved and it was elected to observe her progress. The episodic bifrontal headaches lasting several hours at a time continued.

Repeat CT 8 months later again showed a cyst containing fluid of cerebrospinal fluid (CSF) density in the posterior parts of the left lateral ventricle (fig. 2A). Neurologic examination revealed pale optic discs without papilledema, gross tremor of the right hand, and slight weakness and drift of the right upper extremity. The deep

C

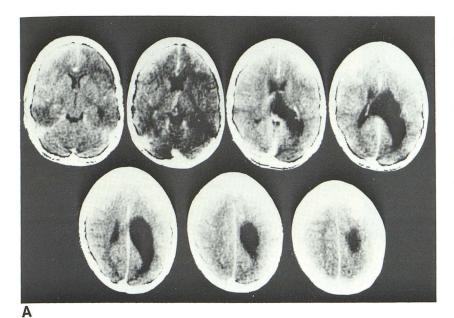
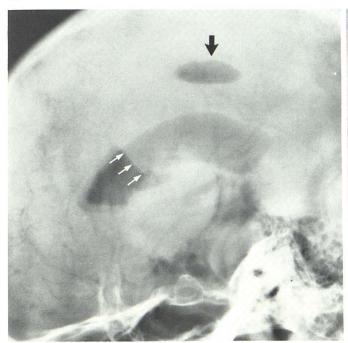
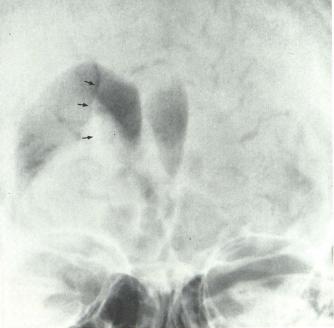


Fig. 2.—Case 2: Lateral ventricular neuroepithelial cyst. A. Contrast-enhanced CT scans. Expansion of left lateral ventricle extending from immediately posterior to frontal horn to occipital horn, in comma-shaped pattern. Left frontal horn not dilated, but occipital and temporal horns somewhat dilated. No definite membrane separating cyst from occipital horn is apparent. Suggestion of separating membrane anteriorly, though this could represent displaced choroid plexus. The septum pellucidum markedly dislocated to right, encroaching on body of right lateral ventricle, which is not enlarged. Attenuation of cyst contents was same as CSF. B, Pneumoencephalogram, lateral view, erect position. Anterior margin of cyst outlined by air in anterior part of body of left lateral ventricle (small arrows). At this stage in examination, small amount of gas had entered cyst, outlining its superior extent (large arrow). Body of right lateral ventricle is increased in height, consistent with compression exerted from left side. C, AP projection, right lateral decubitus position. Air has now entered expanded left trigone and posterior part of left temporal horn. Part of lateral wall of cyst outlined between this air and air that had entered cyst (arrows). Some air still trapped in left frontal horn anterior to cyst.





B

tendon reflexes were normal and symmetrical. The gait and the sensory examination were normal.

A plain film skull examination was normal. A lumbar puncture revealed no increase in CSF pressure and CSF analysis was normal. A fractional lumbar pneumoencephalogram (figs. 2B and 2C) revealed a large mass within the body of the left lateral ventricle. During the course of the study, a moderate amount of air entered the mass, confirming its thin-walled cystic nature. Transfemoral left internal carotid and right vertebral angiography showed a slight mass effect deep in the left cerebral hemisphere, resulting in minor depression of the posterior part of the internal cerebral vein. There was no neovascularity.

A ventriculovenous shunt was performed 10 months after onset of symptoms. The ventricular catheter was inserted through a left parietal burr hole into the posterior left lateral ventricle. The symptoms abated, but recurred in 2 weeks. A revision of the ventricular shunt was performed, including insertion of a low pressure valve. However, headaches returned a few days after the shunt revision and a second revision was performed 11 days later.

Over the next 21 months she continued to have headaches lasting several days at a time. An attempt to place a catheter within the cyst failed. A left occipital craniotomy, microsurgical drainage, and excision of the cyst was then performed. The posterior wall of the cyst was quite firm and a part was excised. The choroid plexus

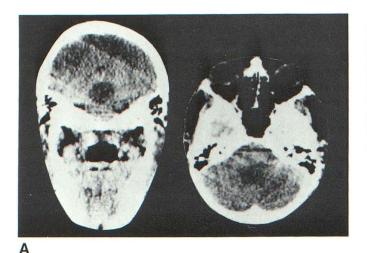


Fig. 3.—Case 3: Fourth ventricular neuroepithelial cyst. A, Coronal and axial CT scans, 320 × 320 matrix, without contrast enhancement. Coronal scan shows rounded contour of cystic lesion and fourth ventricle. No membrane separating low density cyst from remaining fourth ventricle identified in either plane. No abnormal enhancement evident after intravenous infusion of 42 g of iodine. B, Pneumoencephalogram, lateral view, erect position. Smooth convex cystic mass bulges into fourth ventricle from its floor. Part of cyst contour incompletely obstructs foramen of Magendie. Residual fourth ventricular cavity expanded, stretched longitudinally, and elevated. Aqueduct moderately enlarged in midline hypocycloidal tomographic section. C, Coronal hypocycloidal tomographic section. Air in displaced and expanded fourth ventricle. Small amount of air outlines lateral surfaces of cyst and caudal surface of most posterior part of cyst.





В

and subependymal veins could then be seen through the cyst wall. The free part of the wall was removed, leaving the part that adhered to the choroid plexus and ventricular wall. The wall was avascular. As the cyst was now collapsed, tubing was placed within the area of the cyst and brought to a reservoir beneath the skin surface.

Histological examination revealed fibrous connective tissue with focal epithelial cells, without evidence of neoplasia, consistent with a neuroepithelial cyst. The tissue fragment was too small to permit subclassification of the epithelium.

Eight days after subtotal cyst excision, a CT scan revealed marked reduction in the displacement of the medial wall of the left lateral ventricle to the right, some reduction in the dilatation of the occipital horn of the ventricle, and less bulging of the roof and lateral wall of the body of the ventricle than before surgery. One week after surgery she had no headache. There was minimal residual right upper extremity weakness and slight difficulty with tandem gait. Five months later she remained greatly improved, with no headaches or seizures, although memory remained somewhat

impaired. There was also a very slight association defect and a slight right homonymous hemianopsia.

Case 3: Fourth Ventricular Neuroepithelial (Glial-Ependymal) Cyst

A 37-year-old man had a 1½ year history of episodic dysequilibrium and vertigo lasting a few seconds, with increasing frequency, and provoked by sudden head movements. Neurologic examination revealed nystagmus on upward, right, and left lateral gaze. A mild left central facial weakness and increased deep tendon reflexes of the extremities, more on the left than the right were also present. Romberg's sign was absent and tandem gait was normal.

CT demonstrated a sharply demarcated low density mass, with homogeneous attenuation equal to CSF, in the area of the fourth ventricle and pons (figs. 3A and 3B). Left vertebral angiography revealed an avascular mass in the region of the fourth ventricle. Fractional pneumoencephalography with tomography revealed a sharply demarcated mass bulging into the fourth ventricle from its

floor (figs. 3C and 3D). The rest of the fourth ventricle was displaced posteriorly, enlarged, and stretched longitudinally and transversely. These radiologic studies suggested a nonneoplastic cyst or epidermoid tumor.

A suboccipital craniectomy was performed, with division of the vermis. The cyst wall abutted the ventricular roof and during the exposure was entered with release of clear watery fluid. The cyst then collapsed to the floor of the fourth ventricle. With microsurgical technique, a biopsy of the posterior cyst wall was taken and the cyst was marsupialized, leaving intact the part of the wall that was firmly attached to the floor of the fourth ventricle. There was no apparent neoplastic nodule. Histologic examination revealed that the cyst wall had an ependymal lining with a basement membrane, external to which there were collagenous and glial layers.

Neurologic examination 2½ years after operation was almost normal. The fourth ventricle remained enlarged, with effacement of its superolateral recesses and anterior bulging of its floor, expressive of atrophy of the mid-vermis and pons, sequelae of the previous chronic mass effect of the cyst.

#### Discussion

#### Pathogenesis

In recent years several investigators have offered explanations for the origin of intraventricular, intracerebral, and subarachnoid cysts of developmental origin, previously described as colloid, ependymal, arachnoid, and choroid plexus cysts, that would accommodate their similarities of histologic appearance, their differences, and their diverse sites of occurrence through the central nervous system [9–15]. Shuangshoti and his colleagues [12–16] emphasized the common origin of all such cysts from the primitive neuroepithelium that lines the neural tube.

Developmental cysts lined by neuroepithelium have been found in extracerebral, intracerebral, and intraventricular locations. They may also occur along the spinal cord [17–21]. Those occurring in a extracerebral position usually have walls formed by arachnoid [22–25], but extracerebral ependymal cysts also have been reported [26–34]. Intracerebral developmental cysts, not communicating with the subarachnoid space or ventricular system, have been reported. Most of these have been described as ependymal cysts [35–39].

Developmental cysts occurring within the ventricular system are typically confined to the anterosuperior third ventricle, in the region of the foramina of Monro, and arise from the neuroepithelium of the diencephalic roof. In this location, the cysts are generally classified as "colloid" cysts and differ from most neuroepithelial cysts occurring in other locations, in that the cyst contents are usually viscid, with gelatinous or mucinous appearance. Light and electron microscopy have shown these cysts to be lined by neuroepithelium of various types [9, 11–14, 40, 41]. In this location the incidence is about 0.5% of intracranial tumors [42].

According to Shuangshoti and colleagues [10, 12, 13], neuroepithelial cysts can develop in any part of the ventricular system lined by the neuroepithelial layer, either by invagination or evagination, with a connective tissue layer inside or outside the neuroepithelial layer, respectively. This concept is in accordance with the behavior of developing

neuroepithelia—choroidal, paraphyseal, and ependymal. When the folded epithelium at a site of invagination or evagination is pinched off, tubules form as part of the developmental process. Obstruction of these tubules and accumulation of secretory products with slow cellular proliferation result in the formation of these cysts. Fragments of choroid plexus may be present on the surfaces of cysts projecting into the ventricle.

Netsky and Shuangshoti [13] also considered ectopic neuroglial tissue containing ependyma-lined canals to be the possible origin of neuroepithelial cysts occurring in extra- and intracerebral locations, including those related to the spinal axis. Although small asymptomatic neuroepithelial cysts, single or multiple, occurring in the choroid plexus of the lateral ventricle have been described [12–14], we found reference to only six examples of *symptomatic* lateral ventricular epithelial cysts, excluding the well known typical colloid cysts of the anterior third ventricle [3–8] and those described by Ciric and Zivin [43] between the fornices and leaves of the septum pellucidum.

Shuangshoti et al. [40] also assert that the presence or absence of various cellular organelles or cell types is merely an expression of normal variation of the regional neuroepithelium. This would explain the occurrence of choroid plexus elements in some ependymal cysts. In particular, cilia are explained to be present in all primitive neuroepithelium, but to disappear from many cells with time. The term neuroepithelial cyst, first used by Fulton and Bailey [44] to describe the various developmental forms of cyst, whatever their location, has therefore come into common usage.

#### Fourth Ventricle Cysts

The occurrence of neuroepithelial cysts within the fourth ventricle is evidently extremely rare. Five cases have been identified in the literature [41, 45–48]. In the case reported by Parkinson and Childe [49], thought by them to represent two "colloid" cysts of the fourth ventricle, no attachment to the ventricular walls or gelatinous material were found. Subsequent review of a stained section of the specimen indicated cysticercosis [50]. Love and White [45], described the case of a 22-year-old woman with two epithelial-lined cysts of the fourth ventricle, causing increase in intracranial pressure and rhinorrhea. The cysts were excised, but their attachments and contents were not described.

Sharpe and Deck [46] reported a neuroepithelial cyst attached to the ventricular surface of the superior cerebellar vermis and expanding into the fourth ventricle. The wall of the cyst consisted of a thin layer of astrocytic glial tissue and was lined by two types of cells in a single layer, one type with frequent cilia, resembling mature ependyma, and a second cell type similar to normal choroid plexus epithelium. This 15-year-old girl had increased intracranial pressure due to obstructive hydrocephalus. Respiratory arrest and death occurred a few hours after lumbar puncture, due to cerebellar herniation. The cyst contained serous fluid.

Palacios et al. [47] described a patient of unstated age in whom a large neuroepithelial cyst occupied and obstructed

the fourth ventricle. Ventriculography showed a large, smoothly contoured mass encroaching on the fourth ventricle from its floor. Angiography revealed no abnormal vascularity.

Di Rocco et al. [48] described a 7-year-old boy who had headaches, macrocephaly, and slowly progressive intellectual impairment. Pneumoencelphalography showed an appearance very similar to that in our case 3 and that on the ventriculogram in the case of Palacios et al. [47]. The mass was avascular at angiography. The histology was that of an intraarachnoid cyst, with surrounding connective tissue. CT revealed the contents to have a density equivalent to CSF and at operation the fluid resembled clear CSF.

#### Cyst Contents

In each of our three cases, the cyst contained clear serous fluid. Protein concentrations were not determined. In each case, the CT attenuation of the cyst fluid was the same as that of the adjacent ventricular CSF. In the two examples of noncommunicating intracerebral ependymal cysts reported by MacGregor et al. [38], the protein content of the cyst fluid was 45 and 11 mg/dl, respectively. Both showed CT attenuations equal to that of CSF.

Friede and Yasargil [39] reviewed 15 cases of intracere-bral or convexity ependymal and glioependymal cysts and added two examples. In these cases, the fluid ranged from clear to turbid and was xanthochromic in two cases. Protein content ranged from 1.1 to 85 g/dl in the seven cases in which measurements were given. In the lateral ventricular arachnoid cysts reported by Yeates and Enzmann [8] and the choroid plexus cyst reported by Andreussi et al. [7] the contents were of CSF density on CT. Prior studies have demonstrated that CSF protein concentrations must be greater than 2 g/dl before CT can reveal abnormal attenuation values [51, 52]. In our three cases and the cases reported by Palacios et al. [47], Yeates and Enzmann [8], and Di Rocco et al. [48], the CT density of cyst contents were indistinguishable from CSF.

The source of the cyst fluid is debated. The finding of pinocytotic vesicles in the lining cells in some of these cysts, and the presence of choroid plexus elements in others, has suggested active cellular transport of fluid. The great variability of the protein content is not understood, although evidence of previous hemorrhage has been identified in some cysts in extra- and intracerebral locations. As in the case of typical colloid cysts of the third ventricle, growth of neuroepithelial cysts in other locations is apparently very slow, and clinical presentation is typically in adult life.

#### Radiologic Diagnosis

On CT, recognition of these cysts as mass lesions within the ventricles, rather than dilatations of the ventricles, is achieved directly by identification of the thin cyst wall separating the contents (isodense with CSF at protein levels below about 2 g/dl) from ventricular CSF. Owing to the relatively coarse matrices used in such cases to date, and

partial volume effects related also the curving cyst wall, identification of the wall has been difficult in the lateral ventricular cases and impossible in the fourth ventricular cases examined to date.

In the lateral ventricle, displacement of the septum pellucidum and choroid plexus, dilatation of the occipital and temporal horns relative to the small frontal horn, and absence of signs of cerebral tissue loss, such as may be reflected in widened cisterns and sulci (or actually signs of compression of the latter), combine to indicate the presence of a ventricular cyst. In the fourth ventricle, entrapment of the ventricle must be differentiated from a cyst (see below). Metrizamide ventriculography by ventricular needling or injection into a ventricular shunt or drainage tube could be used for identification of cyst walls as an alternative to pneumography.

Angiography can reveal a nonspecific mass effect without pathologic vascularity. Its main value is in the exclusion of cystic neoplasms with small vascular mural nodules.

#### **Differential Diagnosis**

#### Isolated (Trapped) Fourth Ventricle

If the foramina of the fourth ventricle are obstructed and a lateral ventricular shunt is performed, the aqueduct may become occluded, with or without clinical evidence of infection. The isolated fourth ventricle may then expand considerably due to choroid plexus secretion, leading to symptoms of a posterior fossa mass [53, 54]. Since the thin membrane of a cyst (developmental or parasitic) may not be resolved by CT, these conditions may appear identical, and differentiation will depend on the clinical context of the former situation.

#### Colloid Cysts of the Anterior Third Ventricle

Neuroepithelial cysts in this more common location offer no difficulty in differentiation from the cysts under discussion. Owing to their strategic location adjacent to the foramina of Monro, they are relatively small at the time of diagnosis, usually not more than 2 cm in diameter [1, 54]. The large majority of colloid cysts of the anterior third ventricle are considerably denser than brain, and CT with intravenous injection of contrast material commonly shows contrast enhancement of the cyst [55]. Hyperdensity seems to be due to high electron density [56], but trace amounts of higher atomic number elements could possibly contribute to the CT density of these cysts [57].

#### Cysticerosis

The cysts of this parasite not infrequently involve the cerebral ventricles [58]. Within the lateral and fourth ventricles, they may occasionally reach a size similar to the neuroepithelial cysts described here. When the cyst wall does not enhance and no calcification is visible within the larva, a CT, pneumographic, and angiographic appearance

identical with that of a developmental cyst may be produced. However, there will commonly be additional cysts in other locations, with or without peripheral enhancement and calcification, indicating the correct diagnosis.

#### Hydatid Cyst

Intracranial location of echinococcal cysts occurs in only 1%–2% and intraventricular hydatid cysts are even less common [59]. These cysts grow to very large size, show a tense rounded configuration, and their fluid has a CT density equivalent to CSF [60]. Angiographic avascularity is characteristic, but they may be differentiated from neuroepithelial cysts by the finding of calcification in the wall and/or daughter cysts. As in cysticercosis, immunologic tests and the finding of cysts elsewhere, particularly in the liver and lungs, may indicate the correct diagnosis.

#### **Epidermoid Tumors**

Epidermoid tumors may occur entirely or largely within the fourth ventricle or, less often, in a lateral ventricle, with ventricular expansion and obstruction. Typically, attenuation values are low and the epithelial wall is avascular, showing no enhancement on CT or abnormal vascularity on angiography. Attenuation may range from a few EMI units below to a few units above that of CSF, and does not change with intravenous contrast enhancement. While the mean density may be the same as CSF, attenuation tends to be somewhat heterogeneous, with spotty areas of somewhat higher attenuation [61, 62]. When these tumors occur within the fourth ventricle, the margins of the lesion may be somewhat irregular, but the contour can be similar to that of fluid-filled developmental or parasitic cyst [61].

Pneumoencephalography commonly reveals the contained whorls of keratinized debris, owing to breakdown of the thin epithelial membrane. In the case of an arachoid cyst described by Yeates and Enzmann [8], a dual kilovoltage scan of the lesion revealed a shift in attenuation values identical to that expected for CSF. The authors suggest that such scanning may offer an alternative to pneumoencephalography in the differentiation of simple cyst and epidermoid tumor [63, 64].

#### Cerebellar Hemangioblastoma

A solitary hemangioblastoma with a very small mural nodule, occurring without stigmata of von Hippel-Lindau disease, could cause differential diagnostic difficulty on CT. In these rare circumstances, angiography by demonstrating the small mural nodule can serve to differentiate.

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