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Radiologic and Pathologic Findings of Intracerebral Schwannoma

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Summary: We report the radiologic and pathologic findings of an intracerebral schwannoma. MR imaging studies showed a superficially located cystic mass with an enhancing nodule and evidence of peritumoral edema or gliosis.

Intraparenchymal schwannomas of the brain and spinal cord are rare. Gibson et al (1) reported the first English-language account of an intraparenchymal intracerebral schwannoma in 1966. Since then, at least 33 additional cases of this neoplasm have been reported, but little has been written about its imaging appearance (2–24). We describe the MR imaging and pathologic findings in a patient with intracerebral schwannoma and, through a review of the literature, discuss the characteristic imaging features of this neoplasm.

Case Report

A 15-year-old boy was admitted to the hospital after accidental head trauma with loss of consciousness. His family history was noncontributory and there was no evidence of neurofibromatosis. Physical examination revealed a left-sided parietal scalp hematoma and laceration without focal neurologic deficit. Laboratory findings were normal. MR imaging studies showed a heterogeneous right-sided parietal mass (Figs 1A–D). The patient underwent a stereotactic biopsy, which yielded amber-colored cystic fluid. Microscopic analysis of solid tissue showed a benign spindle-cell neoplasm, consistent with a low-grade tumor, possibly a pilocytic astrocytoma. Ten days later, the patient underwent a right-sided parietooccipital craniotomy with resection of the tumor. Sonography was used to identify a solid nodule located immediately beneath the cortical surface with a prominent cystic component along the postero-lateral aspect of the nodule. The nodule was gently separated from the overlying brain and was removed in one piece. The cyst, which contained dark-greenish fluid, was decompressed. There was no visible evidence of residual tumor. Microscopic analysis of the tissue showed areas of nuclear palisading, typical of a schwannoma. Densely cellular tumor alternating with loosely textured myxoid tumor was present in equal portions, consistent with Antoni type A and Antoni type B tissue, respectively. A distinct interface between tumor and surrounding brain was present in some sections (Fig 1E). Immunohistochemical testing for S-100 protein was diffusely positive, whereas glial fibrillary acidic protein (GFAP) was negative in the tumor cells, confirming the diagnosis of schwannoma. An MR imaging study performed 1 day later showed no evidence of residual tumor. The patient remains asymptomatic 2½ years later.

Discussion

Intraparenchymal schwannomas of the brain are rare neoplasms, with only 35 cases reported over the last 30 years. Careful analysis of clinical and radiologic findings of these cases, including one of our own, has disclosed several characteristic features. Unlike vestibular schwannomas, those in the intraparenchymal brain exhibit no female predominance (female 17, male 18) (25). Furthermore, the majority of cases have occurred in children and young adults, with median and average ages at presentation of 21 and 29 years, respectively. Seventy percent of the reported tumors were clinically apparent before patients had reached the age of 30. In contrast, vestibular schwannomas rarely occur in children unless in association with neurofibromatosis type 2 (26). The most common signs and symptoms of intraparenchymal schwannomas are headaches, seizures, and focal neurologic deficits.

Characteristic pathologic and imaging features include calcification, cyst formation, peritumoral edema and/or gliosis, and superficial or periventricular location. Calcification, believed by some to be a characteristic finding in intracerebral schwannomas (21), has been seen radiologically or confirmed histopathologically six times. In contrast, calcification has rarely been seen radiologically or histopathologically in vestibular schwannomas (27). Cysts have been detected on MR images in 20% of vestibular schwannomas (28). Cysts were documented pathologically or by imaging studies in 19 of the cases we reviewed. The cystic component of the tumor may be central and secondary to necrosis. Alternatively, as seen in our case, the cyst may be large and located peripherally, with an associated mural nodule. These extramural/arachnoid cysts may be caused by mechanical trapping of CSF or by leakage of hemorrhagic material from the tumor, causing adhesions and secondary arachnoid cyst formation (28). Extramural cysts seen on MR imaging studies in 13% of acoustic schwannomas were confirmed pathologically.
or on imaging studies in 10 intracerebral schwannomas. The solid component of intracerebral schwannoma was found superficially or adjacent to the ventricular system in 12 (41%) of 29 cases (six cases were omitted because of data restrictions) and seven (20%) of 35 cases, respectively.

MR or CT studies were performed in 27 of the 35 reported cases. The presence of hypodensity or hypointensity, representing edema and/or gliosis, was specifically addressed in 22 patients and was found in 17. In our case, peripheral T2 hyperintensity was present with focal prominence of a sulcus (Fig 1D), suggesting atrophy, while the T2 signal abnormalities perhaps represented a combination of edema and gliosis. The cause of the edema surrounding this benign neoplasm is uncertain. Vascular endothelial growth factor (VGEF) messenger RNA expression has been implicated in edema formation around some meningiomas and may be an important etiologic factor explaining peritumoral edema in other benign neoplasms (29). The presence of VGEF in intracerebral schwannomas has not been confirmed however, and its role in edema formation requires further investigation. Chronic edema can result in degeneration of white matter and gliosis, possibly explaining the frequent association of these findings (30). Of the 27 patients who had MR or CT studies, 21 received contrast material, and the pattern of enhancement was specifically addressed in 20; in 10 of these, the pattern was described as homogeneous.

MR imaging findings of intracerebral schwannoma are varied. DiBiasi et al (23) reported a 1.5-cm well-
circumscribed mass located within the superficial frontal lobe showing hypointense T1 signal and hyperintense T2 signal that enhanced intensely and homogeneously. A 5-cm cystic frontal lobe mass with a solid, homogeneously enhancing component has been reported (21). The solid portion showed T1 hypointensity and mixed T2 hypointensity/hyperintensity. Calcification, seen on CT scans, may have contributed to the hypointense T2 signal. Similar T2 hypointensity was seen in our case and may be explained by the presence of hyalinized stroma and collagen deposition seen at microscopic examination. Cystic and solid brain stem and inferior vermian schwannomas have also been described (15, 20, 22, 24).

The origin of intracerebral schwannomas is uncertain. Schwann cells have been detected around arteries in the subarachnoid space and within the periphery of the brain along perivascular nerve plexus (31). Similar tiny myelinated nerves may reside in the tela choroidea, the embryologic anlage of the choroid plexus. The location of these nerves may help to explain the predilection of the reviewed cases for superficial or deep periventricular sites. The existence of Schwann cells deep within the substance of the human brain remains controversial. The only evidence of the presence of nonneoplastic Schwann cells in brain tissue is the rarely observed peripheral-type myelin in multiple sclerosis plaques or at the edge of old infarcts (32). These Schwann cells may undergo neoplastic conversion (33). Russell and Rubinstein (25) noted a close resemblance of mesodermal pial cells to neuroectodermal Schwann cells, hypothesizing that pial cells may sometimes undergo conversion to Schwann cells, thus serving as the origin of some intracerebral schwannomas.

The differential diagnosis of an intracerebral schwannoma includes several other neoplasms that may occur in children and young adults. These include pilocytic astrocytoma, pleomorphic xanthoastrocytoma, and ganglioglioma. Most pilocytic astrocytomas are discovered during the first two decades of life, and unlike in intracerebral schwannomas, surrounding edema is rarely present (34). Pleomorphic xanthoastrocytoma is a typically benign supratentorial astrocytoma occurring primarily in children and young adults. It affects males and females equally. MR imaging characteristics are variable, with T1 isointensity and hypointensity and mixed T2 isointensity and hyperintensity (35). Similar to intracerebral schwannomas, cysts and mild to moderate edema are common. This neoplasm is also located superficially, involving the cortex and leptomeninges, and may be firmly attached to the dura; however, calcification is rarely seen on CT scans or confirmed histopathologically, which may help differentiate pleomorphic xanthoastrocytoma from intracerebral schwannoma. Ganglioglioma is a rare neuronal tumor most commonly found within the temporal lobes. Cysts, contrast enhancement, and calcification may be present (36, 37), and its appearance may be nearly identical to the tumor seen in our case (38).

Conclusion

Intracerebral schwannoma is a rare, benign neoplasm. It is usually located superficially or adjacent to a ventricle. Characteristic imaging features include cyst formation, calcification, and evidence of peritumoral edema or gliosis. The recognition of this benign and potentially curable neoplasm and its differentiation from other neoplasms, some of which have less favorable outcomes, is of obvious importance.

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References