Frontal intraparenchymal schwannoma

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Schwannomas account for approximately 8% of primary intracranial tumors, and constitute almost 90% of tumors in the cerebellopontine angle, which is always associated with the eighth cranial nerve.1 Intraparenchymal schwannomas are extremely rare, and only 64 cases have been reported to date in the literature,2-7 among which the first case was described by Gibson et al in 1966.2 Although most of intraparenchymal schwannomas are benign, 7 cases of malignant intracerebral or intramedullary schwannomas have been reported previously.3-5 In order to better define the clinical and pathological features and therapeutic strategies of this entity, we report a case of frontal intraparenchymal schwannoma and review the literature.

Case Report. A 39-year-old female patient presented with a 9-year history of headache and intermittent seizures, accompanying decreasing memory for one year. Neurological examination revealed no abnormalities. Her MRI showed a 2-cm diameter circular mass located in the frontal lobe with a cyst, and the nodule showed isointense-signal intensity on T1-weighted images, hyperintense-signal intensity on T2-weighted images, with significant enhancement by gadolinium-diethylenetriamine penta acetic Acid (Figure 1). Preoperatively, it was diagnosed as a glioma. Total surgical removal of the lesion led to a favorable result. Post-operative histo-pathological examination showed characteristic Antoni A and B areas consistent with intraparenchymal schwannoma. Intraparenchymal schwannoma is an extremely uncommon lesion, which is seen mostly in young adults and children. The main clinical symptoms include rising-intracranial-pressure-related manifestations and associated seizure disorders. The possible developmental origins, histological, imaging features, and protocols of treatment for this entity are discussed.
Discussion. Intracerebral parenchymal schwannoma (neurinoma) is so rare that its oncogenesis is still enigmatic even now. It was hypothesized that smaller vessels in the cortical and periventricular regions had a propensity for developing into schwannomas, but Feigin and Ogata proposed that mesenchymal multipotential cells differentiate into Schwann cells. Without female propensity, approximately 70% of patients are teenagers and young adults, and the ages at diagnosis range from 6 months to 84-years-old. The clinical manifestations of intracerebral parenchymal schwannoma depend mainly on the locations and the sizes of the tumors. The most common symptoms and signs include headache, seizures, and focal deficits. Microscopically, analysis of the tissues has shown areas of nuclear palisading, characteristic of a schwannoma; and, dense, cellular tumor, alternating with loosely textured myxoid tumor is present in equal portions, consistent with Antoni type A and Antoni type B tissue. A distinct interface between the tumor and surrounding brain tissues was present. Immunohistochemical testing for S-100 protein is diffusely positive, whereas GFAP is negative in the tumor cells, confirming the diagnosis of schwannoma.

Characteristic pathologic and imaging features include calcification, cyst formation, peri-tumoral edema, and/or gliosis, and a superficial or periventricular location. Zagardo et al considered calcification as one of the specific features of intraparenchymal schwannoma. In contrast, calcification rarely has been seen radiologically or histopathologically in vestibular schwannomas. The cyst formation rate is 54.1%, much higher than that of acoustic schwannoma. Cysts could be formed after central secondary necrosis or hemorrhage, or it may occur around a solid nodule, as in this case. Approximately 41% of these tumors are located superficially; mostly in the cerebral and cerebellar hemisphere. Also, 20% of them occur around ventricles. Most occur in the fourth ventricle, and the brain stem and suprasellar cistern are other possible locations. Interestingly, Sharma et al reported that 6 lesions were in the left cerebral hemisphere, that could be observed more often than in right cerebral hemisphere. The CT findings of intracerebral schwannomas reveal that the tumor exhibits both high and low density. Furthermore, MR imaging findings of intracerebral schwannomas are varied. The lesion could be solid or a cystic-solid mass. The solid portion showed T1 hypointensity and
mixed T2 hypointensity/hyperintensity with significant enhancement. Calcification, seen on CT scans, may have contributed to the hypointense T2 signal. Similar T2 hypointensity may be explained by the presence of hyalinized stroma and collagen deposition seen under microscopic examination. On the imaging pictures, intracerebral schwannomas may mimic astrocytomas or malignant tumors, and it is important to distinguish them from other neoplasms. The neuroimaging features are non-specific, and a definitive diagnosis can only be made on the basis of histology. The protocol of treatment of intracerebral schwannomas is total excision; however, it depends on the locations of the tumors. Complete relief of clinical symptoms and signs is mostly achieved after total or radical surgical removal. In our case, the one-year follow-up evaluation, post excision, revealed no headache or epilepsy, and improvements in memory. Obviously, total surgical removal of the lesion led to a favorable result. As for the literature and our case, intraparenchymal schwannomas offer a rare differential diagnosis for intra-cerebral tumors. It is worthy of further studies on its origins and familiarization with its fundamental features and management for neurosurgeons. It should be noted that routine follow-up examination of skull MRT will be necessary due to paucity of the prognostic data.

References


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