Frontal intraparenchymal schwannoma

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ABSTRACT

تعرضت مريضة تبلغ من 39 عام لنوبات صرع متقطعة وصداع لمدة 9 سنوات وانخفاض في الذاكرة لمدة عام واحد، بدون إشارات عصبية مسبقة. أظهر الكشف بأشعة الرنين المغناطيسي MRI عن وجود آفة معززة بمقدار 2x2سم في الفص الجبهي. مع وجود كيس و وذمة التي لم تكون متصلة للأم الجافية أو المنجل. قبل العملية الجراحية تم تشخيص ذلك بورم دبقي. أدت جميع المناطق الجراحية التي تمتت إزالتها من الآفة إلى نتائج مقبولة. أظهر الفحص المرضي والنسيجي بعد العملية الجراحية وجود خصائص انتوني (A و B) في المناطق المصابة بورم الغمد الليفي العصبي ويعد ذلك عبارة عن آفة غير شائعة والتي يمكن رؤيتها في الغالب لدى البالغين الشباب والأطفال. تمت مناقشة الاضطرابات الرئيسية والمصادر التطويرية المحتملة والسمات النسيجية والتصويرية وبروتوكول المعالجة بالكامل.

A 39-year-old female had been subject to headache, and intermittent seizures for 9 years and decreasing memory for one year, without obvious neurological signs. An MRI revealed a 2x2 cm contrast-enhanced lesion in the frontal lobe, with a cyst and peritumoral edema, which was not attached to the dura or falx. 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.

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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**). The preoperative diagnosis was a glioma. The tumor was totally removed by a frontal craniotomy, and it was observed that the lesion was soft, fleshy, and highly vascular, with a well-defined border. The excised mass had no attachment to the dura or falx. Subsequent histological examination showed an encapsulated mass composed of benign spindle cells arranged in Antoni A and B patterns; also, the formation of Rosenthal fibers and a cystic lesion appeared in some regions (**Figure 2a**). Immuno-histochemical results revealed: S100 protein (+), Vimentin (+), epithelial membrane antigen (EmA) (-), glial fibrillary acidic protein (GFAP) (+/-), oligodeoxynucleotide 2 (oligo 2) (-), and MIB-1 labeling index (MIB) <1% (**Figure 2b**). The final pathological diagnosis was a typical intraparenchymal schwannoma. She was discharged 2 weeks after operation, the follow-up MRI 3 months after operation showed no recurrence of the tumor (**Figure 3**), and she was seizure-free and had no headache during the following year.

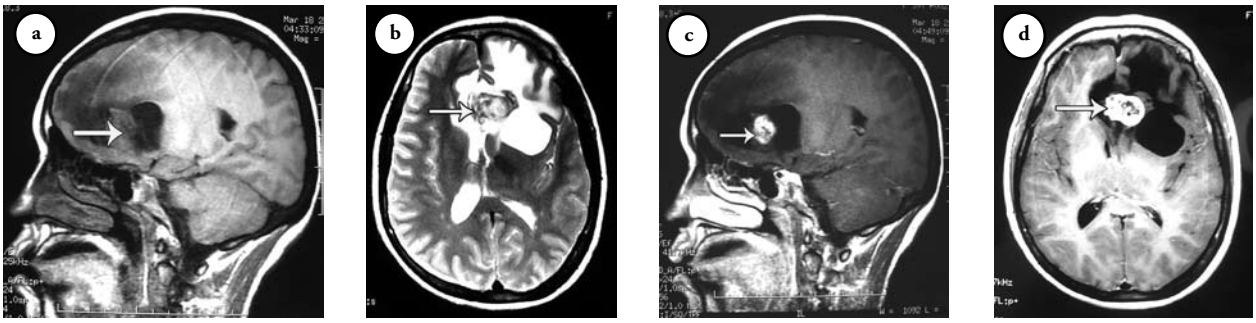

Figure 1 - The MRI showed an intracerebral lesion accompanied by a cystic-solid lesion and remarkable peritumoral edema. **a)** Sagittal T1-weighted image showed isointense-signal intensity mass (arrow). **b)** Hyperintense-signal intensity on Axial T2 weighted image (arrow). **c & d)** The nodule with a significant enhancement by GD-DTPA (arrows).

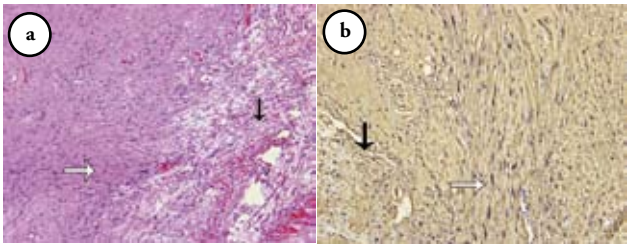

Figure 2 - Photomicrograph showing **a)** showing interlacing bundles of spindle cells in classic Antoni A (white arrows) and classic Antoni B (black arrows) patterns (stain: hematoxylin and eosin x 100). **b)** Photomicrograph of S-100 staining diffusely positive.

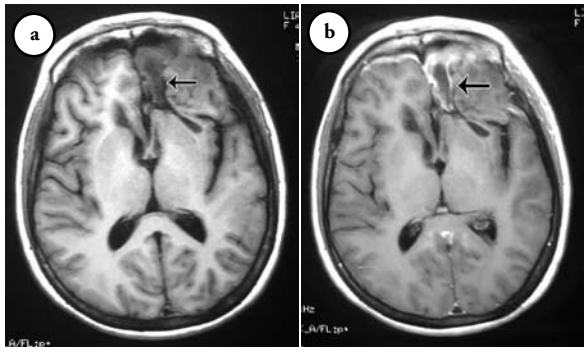

Figure 3 - Patient MRI 3 months after operation showing **a)** axial T1-weighted image. **b)** no significant enhancement by gadolinium-diethylenetriamine penta acetic acid (arrows).

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,[5] but Feigin and Ogata[8] 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[6] to 84-years-old.[7] 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.[9]

Characteristic pathologic and imaging features include calcification, cyst formation, peri-tumoral edema, and/or gliosis, and a superficial or periventricular location. Zagardo et al[9] 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.[9] 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.[9] Also, 20% of them occur around ventricles.[9] Most occur in the fourth ventricle, and the brain stem and suprasellar cistern are other possible locations.[6] Interestingly, Sharma et al[6] 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.[10] Furthermore, MR imaging findings of intracerebral schwannomas are varied.[9,10] 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.[6] 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 MRI will be necessary due to paucity of the prognostic data.

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