MRI Diagnosis of Primitive Neuroectodermal Tumors

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Abstract **Objective** To analyze the MRI features of intracranial primitive neuroectodermal tumors (PNET) and improve the diagnostic accuracy. **Methods** 10 cases of PNET were collected, which were all diagnosed by pathology. Of them there are 7 men and 3 women, their ages ranged from 6~58 years with mean age 26 years old. All of 10 cases were examined by conventional plain MR before operation, and postcontrast MR scans, spin echo (SE) sequences were adopted. **Results** The 10 cases were all supratentorial lesions included 5 cases of temporal lobe lesion, and one case in occipital, parietal, frantal, fronto-parietal,and tempo-parietal lobes respectively. All the tumours were big in size, and the largest one was 6.0cm×5.2cm×6.2cm, the smallest one was 4.5cm×3.8cm×3.8cm. All of the tumour were located in the shallow of the brain parenchyma and close to the dura mater , and the meniges around the lesions were not incrassation obviously. Seven of them were regular and three were irregular in shape, the borderline of the tumor was distinct. The parenchyma in 6 cases were uniformity, and small cystic degeneration could be seen in 4 cases. The peritumor area were with slight edema. The lesions demonstrated long T1 and long T2 signal intensity, mixed signal intensity on T1WI and T2WI. Seven cases demonstrated regular enhancement and 3 cases ring-like enhancement. **Conclusion** MR findings of PNET are short of characters , and the final diagnosis relies on pathology yet.

Key words Intracranial, Primitive neurotodermal tumour, Magnetic resonance image

Primitive neurotodermal tumour (PNET)was first reported by Hart in 1973, which is a scarce kind of nervous system tumour with high malignance. PNET is a kind of primitive tumor which comes from nervous crest, mostly produced by primitive neurepithelium and have a ability of multi-directional differentiation. PNET possesses the character of invasiving growth, general dissemination in cerebrospinal fluid, so the prognosis is bad and the final diagnosis in most cases must rely on the pathologist. Their histomorphology belongs to malignant small round cell tumor, and was divided into central and peripheral kinds. The peripheral tumor is common and much more reported, and the central is less, especially the supratentorial primitive neurotodermal tumour which only 0.1 percent in brain tumor, and often happen to children[3]. Now we retrospective analyses the PNET examples, and try to find PNET´s feature in MRI, in order to raise the accurate rate of imaging diagnosis.

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MATERIALS AND METHODS

Clinic materials

Ten cases of PNET were collected since Jan. 2000 to Jun. 2005, of them, 7 cases were male, and 3 were female, the age of the patients ranged from 6 to 58 years with mean age 26 years old. The courses of disease were from 7 days to 10 months.

In clinical manifestation: Six cases had intracranial hypertension symptome,such as headache and vomit, in them, 4 cases with limbs inability and spasm, and one with instability of gait, four cases had headache only. They all received head MRI plain scan and enhancement examination, and constrast medium is Gd-DTPA, dose is 0.2 ml/kg.

Examination method

1.5T Edge Eclipse size supraconduction magnetic resonance imaging machine produced by Maconi accompany in USA was used to scanning successively with SE sequence, layer thick is 10mm, layer distance is

5 mm, scanning parameter: axial view T2WI(TR/TE=4000/112ms), axial view and anteroposterior axes(TR/TE=350/12 ms)plain scan and enhancement scanning.

RESULTS

MRI appearance

The 10 cases were all supratentorial lesions included 5 cases of temporal lobe lesion, and one case in occipital, parietal, frantal, fronto－parietal,and tempo－parietal lobes respectively. All the tumors are big in size, and the largest one was 6.0cm×5.2cm×6.2cm, the smallest one was 4.5cm×3.8cm×3.8cm. The placeholder symptomes were evident. All of the tumour were located in the shallow of the brain parenchyma and close to the dura mater, but the surrounding dura mater are no obviously thickening. The appearance of tumor: 7 cases were round alike, 3 examples were irregular; six cases were solid tumor mass with clear and well－described edge and another 4 cases with cystic degeneration; there were light edema around tumor, but 2 cases without. Tumor signal were uniformity, T1WI shows slightly low or low signal, T2WI shows slightly high or high signal, two cases showed T1, T2 signal; there were 7 cases showed uniform enhancement, 3 samples showed circular enhancement. The dura mater are no obvious thickening and enhancement. Eight cases were diagnosed as neurogliocytoma, and the other two cases as germ cell tumor and lymphadenoma respectively before operation.

pathology result

Ten cases were all proved to be PNET by pathology after operation.

DISCUSSION

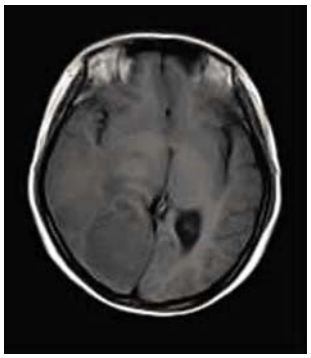

fig.1

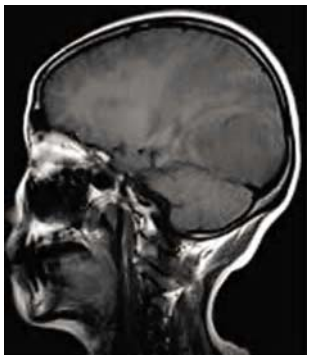

fig.2

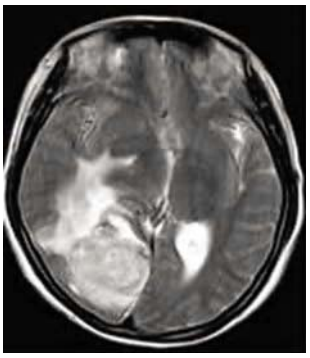

fig.3

Fig. 1–3: T1WI transverse position and anteroposterior axes position, T2WI transverse position: we can see round like solid tumor mass shadow in right occipital lobe, clear edge, peri－tumor edema, tumor shows T1 low signal,T2 high signal.

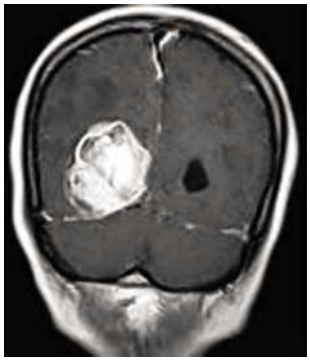

fig.4

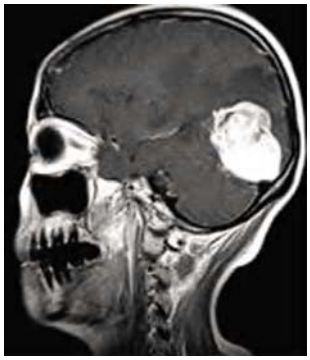

fig.5

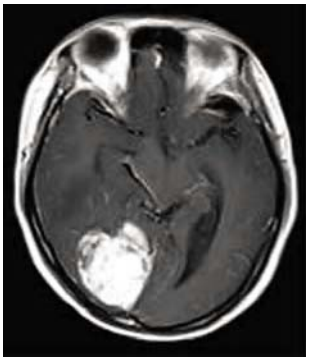

fig.6

Fig. 4–6: T1WI enhance coronal,anteroposterior axes and transverse position, tumor is obvious intensification, clear edge, the placeholder was evident, posterior horn of right lateral ventricle is compressioned.

Overview of PNET

Primitive neurotodermal tumor(PNET), which was first reported by Hart in 1973[1,2], is a kind of undifferentiation tuomor which originated from children´s cerebra, it is similar to medulloblastoma in cerebellum in histology and comes from undifferentiated precursor cell of neuroepithelial cell.

In 2000, neural embryonic tumors were divided into four kinds by WHO: medulloepithelioma, ependymocytoma, medulloblastoma as well as its variety and supratentorial primitive neurotodermal tumour as well as its variet(including neuroblastoma). Supratentorial primitive neurotodermal tumour once have other names [4~6], such as cerebral medulloblastoma, cerebral neuroblastoma, neuroblastoma of cerebral ganglionic cell and blue tumor.

At present, the definition of PNET is: a kind of highly malignant tumor located in cerebrum or above saddle constituted by undifferentiation or poorly differentiated neurepithelium. Intercranial PNET are all located above cerebellar tentorium, tumor cell can differentiate to the neurocyte, astrocyte, ependymal cell, muscle cell and melanocyte cell lineage[6].

The clinical and MRI features of PNET

The clinical manifestations of the disease are no specificities, but tumor is usually big in size, and can cause acute intracranial hypertension and occupied symptoms and physical sign. Patients usually show intracranial hypertension symptom firstly, such as headache,vomitting and so on. There are 6 cases went to hospital because of intracranial hypertension symptom and 4 cases had just headache only in this group, and 7 cases are adults, 3 cases are chindren.

The MRI appearance of PNET usually showed substantive tumor shadow, round like or irregular in shape with clear edge, tumor volume was comparatively large, placeholder effect was evident; there were cystic degeneration, calcification, and flow empty blood vessel shadow in the tumor; there was light edema or no obviously edema in the region around the tumor; the MRI signal was relatively uniformity, T1WI showed a little low or low signal, T2WI showed equal high or high signal; they showed uniform enhancement or circular ennoblement after enhancement. But we discover beside the appearances that we mentioned, the tumors were located superficially, close to dura mater, and the dura mater showed no obvious thickening or enhancement around tumor.

Main differential diagnosis

Elementary astrocytoma It was usually located in white matter, clear bounary, steady signal such as long T1, long T2 signal were not obviously strengthened, there were no edema around the tumor, and most tumors were located in deep part of the parenchyma of the brain. There were usually calcification for oligodendroglioma, which was difficult to distinguish from PNET, but PNET enhancement much more obviously and more uniformity than oligodendroglioma.

Metastatic tumors They usually happend to junctional zone of gray mater and white mater, often more than one. If the focus of infection is large, it is easy to happen cystic degeneration and bleeding, and obvious dropsy around tumor, this is valuable for distinguish, so we can according to patient´s history and display of MRI to distinguish metastatic tumor and PNET.

Lymphoma They usually happend to median line, single or more than one, shows focal glaebule shadow. It shows long T1, equal or little long T2 signal, less cystic degeneration, obviously strengthen, while PNET tumor are superficial, near to dura mater, but the dura mater had no obvious thickening or strengthen.

Germ cell tumor It need to distinguish with PNET located in third ventricle, its signal and strengthen was usually inequable and located in pineal body area, also occur above saddle or in basal ganglia area. It can be easily to distinguish from PNET according to tumors position and signal.

Analysis about misdiagnosis

This disease has a very high misdiagnosis rate, major reason is that we know not enough to PNET, its incidence is very low, especially in adult. There are much similarity between PNET and neurospongioma, but the focus of PNET is big, and the zone of necrosis is small, the boundary is clear, and shows "ring kind" obviously

after strengthening, which can help us to distinguish from neurospongioma. CT is a good supplementary examination way, it can discover calcification inside focus of infection, and we can use this to distinguish the poorly differentiated glioma (except oligodendroglioma) or metastatic tumor from PNET, but the final diagnosis still depends on the pathologic diagnosis.

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