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Unusual presentation of a tectal plate tumor in a young adult

Nietypowa prezentacja guza blaszki czworaczej u młodej osoby dorosłej

INTRODUCTION

Tectal plate tumors are a subgroup of brain stem tumors that rarely occur among adults. Brain stem tumors account for approximately 10-30% of brain tumors in children and for only 2.5% in adults. The peak incidence prevailed in the first decade in children and in the 3rd and 4th decade in adults. The most common type of tectal plate tumor is the glioma. Histopathologically brain stem gliomas can range from WHO Grade 1 to 4. Grade 1 is the juvenile pilocytic astrocytoma, grade 2 – the diffuse astrocytoma, the most common type in children and adults, grade 3 – the anaplastic astrocytoma, and grade 4 – the glioblastoma multiforme (6,16). Apart from gliomas, other types of tumors localized in the quadrigeminal plate or cistern include oligodendroglioma (3), ependymoma (10), primitive neuroectodermal tumors (4), metastasis (7), lymphoma (5), lipoma (15), melanoma (20), dysembryoplastic neuroepithelial tumor – DNT (9), cavernomas (8), arachnoid cycts (19), hamartoma (12) and periaqueductal gliosis (1). Intrinsic non-neoplastic tectal lesions include vascular (occult cerebrovascular malformation, mesencephalic arteriovenous malformation with associated vein of Galen aneurysm and ischemic disease), traumatic (diffuse axonal brainstem injury and hemorrhage), inflammatory (multiple sclerosis, cysticercosis), degenerative (progressive supranuclear palsy) and congenital (Chiari II malformation) (5). Lesions with slow growth pattern and low grade histological features (e.g. hamartomas) are called "benign intrinsic tectal tumor(s)" (18).

Here we present a case of benign tectal plate tumor in a young adult presenting with epilepsy.

Case report

In 2002 a 14-year-old, previously healthy girl was admitted to a Children's Outpatient Department because of a severe headache with nausea and vomiting for about a week. In neurological examination papilledema and a positive Romberg test were found. Computed tomography (CT) scan of the brain

revealed non-communicating internal three-ventricular hydrocephalus with signs of increased intracranial pressure, without focal lesions of the brain (pictures are not available presently), but MRI was not done. Mannitol therapy first, and then a ventricular-peritoneal shunt alleviated the symptoms. Hydrocephalus of unknown etiology was diagnosed. Since 2005 there were recurrent episodes of fainting, without pathological changes in EEG and cardiological examination, and no new lesions in CT of the brain. At the age of 19, involuntary movements of the right upper limb with disturbed consciousness repeated especially in connection with the sleep. There were episodes of cluster seizures. She was hospitalized and EEG showed paroxysmal bilateral discharges in fronto-temporal regions. Complex partial seizures were diagnosed and valproic acid therapy was introduced. Epilepsy was well controlled however excessive sleepiness, memory problems and psycho-motor slowness were rapidly progressing. She was admitted in December 2007 to the Neurology Department of our hospital because of increasing cognitive impairment and upward gaze paralysis. She complained of severe memory problems and declining school performance. She required help with every day activities and bodily needs. Apart from cognitive impairment (MMSE = 25 pts.), bradyphrenia and bradykinesia, in neurologic examination there was also the Parinaud syndrome and unstable Romberg test. There were no changes in her laboratory (blood and cerebro-spinal fluid) results. EEG showed paroxysmal discharges in fronto-temporal-occipital regions bilaterally with slight slowing of the background activity. Control CT scan of the brain showed a small unidentified structure with focal calcification in the region of tectal plate. MRI of the brain was performed corroborating the diagnosis showing an enlarged tectal plate (10 x 15 mm), hyperintensive in T2 and hypointensive in T1, with focal isointensive, non-enhancing lesion on its left side. The picture was suggestive of the tumor of the tectal plate. There was no active hydrocephalus. Neurosurgeons recommended observation of the patient. Steroid-tapering therapy was applied. In April 2008 the patient's state improved, cognitive problems and bradykinesia withdrew, the palsy of the upward gaze persisted. Control MRI showed the known lesion that remained stable. In November 2008, there were no new abnormalities in neurologic examination and in MR spectroscopy in the area of brain tumor there was an elevated choline peak, increased choline/creatinine ratio, with no changes in N-acetyl aspartate peak; the morphologic picture was suggestive of tectal glioma of low-grade malignancy, e.g. pilocytic astrocytoma. Presently, after 3 years of observation, the patient's state is stable, with no symptoms of intracranial hypertension and no abnormalities in neurological examination. Last follow-up MR spectroscopy (March 2010) did not reveal any progression of the tumor.

DISCUSSION

Presenting symptoms of tectal plate tumor include symptoms of raised intracranial pressure (headache, drowsiness, nausea, and vomiting) due to hydrocephalus caused by compression and occlusion of the aqueduct (11). Tectal lesions may present with diplopia reflecting an internuclear ophthalmoplegia, indicating involvement of the medial longitudinal fasciculus. Less frequently seen is the Parinaud syndrome, with palsy of the upward gaze and accommodation, dissociation of light and accommodation (loss of pupillary reflex to light with preservation of pupilloconstriction in response to convergence), eyelid retraction, and convergence-retraction

nystagmus. Rarely, behavioral changes or seizures may be seen in children. Older children may have deterioration of handwriting and speech (6,16,18). In our patient the history of seizures (since the age of 17) and increasing cognitive impairment were atypical, occurring mostly in younger patients. The development of dementia and epilepsy, that were the reasons that revealed the presence of the tumor, was probably caused by its slight progression with possible increase of intracranial pressure, and the following improvement was the consequence of steroids administration. We cannot exclude also that the incidence of epilepsy might have been a complication of the ventricular-peritoneal shunt placement.

The pathogenesis of seizures in brain neoplasms can be interpreted in several ways: the peritumoral microenvironment could be associated with altered level of enzymes such as lactate dehydrogenase, adenosine 3',5'-cyclic monophosphate dehydrogenase, enolase, and thymidine kinase, which may cause metabolic imbalance. Altered expression of connexins may be the cause of hyperexcitability of the perilesional environment, and it may spread to the cortex. Another cause of epilepsy can be altered expression of neurotransmitter receptors (glutamate, kainite) that contribute to seizure generation (2,14,17). Such pathogenesis of seizures can be taken into consideration also in our case.

Benign tectal tumors remain often radiologically unrecognizable in CT; the majority of cases appear as a noncommunicating hydrocephalus, although sometimes calcification on the tectal plate may be observed, as in our case. MRI of the head is the diagnostic test of choice. The tumor was probably not recognized earlier in our patient because of not applying MRI. The tumor has low-to-normal signal intensity on T1-weighted images and high signal intensity on T2-weighted images. There is usually no contrast enhancement, it is usually minimal or absent in small lesions; contrast enhancement should raise suspicion of a metastatic lesion. MR spectroscopy has been used to help distinguish between tumor and nontumor lesions in the brain. An elevated choline peak suggests neoplasm (1, 5, 11, 16, 20).

The importance of MRI of the brain in the diagnosis of hydrocephalus should be stressed. Hydrocephalus can be successfully treated with an endoscopic third ventriculostomy; a simultaneous biopsy of the tumor is possible (13,18). However, in our patient, no MRI of the brain was done as the signs of increased intracranial pressure appeared and instead of endoscopic third ventriculostomy, ventriculoperitoneal shunting was done. The lack of appropriate diagnostic tools at the beginning of symptoms led to the delayed diagnosis and the lack of the right follow-up (i.e. annual MRI).

Adult tectal plate tumors have frequently indolent course, are more likely to be low grade and remain localized, so survival may be significantly longer in adults comparing to younger patients. Some adult patients with a tectal lesion, with mild symptoms of long duration, may be candidates for observation alone; radiotherapy can be reserved for patients with clear evidence of a tumor progression. Typically, biopsy and/or surgery are not required for diagnosis or treatment of tectal gliomas and cannot be recommended routinely. Although diagnosis can be made by MRI alone, MR spectroscopy is helpful in establishing the type of a lesion. The tumor should be monitored by regular clinical examination and annual MRI (6,16,18).

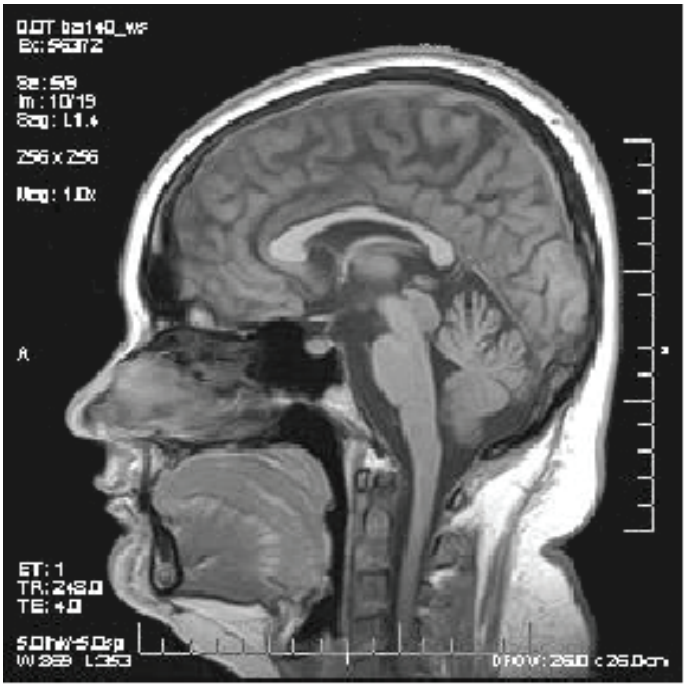

Fig. 1.T1-weighted image, sagittal view, with contrast, showing non-enhanced tectal mass

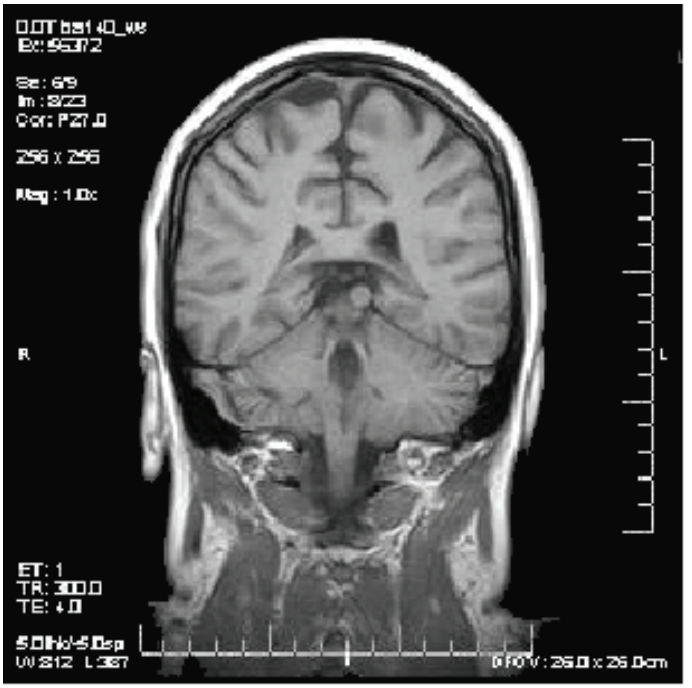

Fig. 2. T1-weighted image, frontal view, showing pathological tectal mass

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ABSTRACT

Tectal plate tumors are a subgroup of brain stem tumors uncommon among adults. We describe a case of tectal plate tumor in a young adult of atypical appearance. As 14-year-old girl the patient had a ventricular-peritoneal shunt placed because of symptoms of increased intracranial pressure and hydrocephalus of unknown etiology. At the age of 19 recurrent episodes of complex partial seizures (including cluster seizures) appeared and therapy with valproic acid was introduced. The seizures ceased, however rapidly progressing cognitive impairment and psycho-motor slowness were observed. In neurological examination bradyphrenia and bradykinesia and the Parinaud syndrome were present. MRI of the brain showed a tumor of the tectal plate without active hydrocephalus. In MR spectroscopy the morphologic picture was suggestive of tectal glioma of low-grade malignancy. Steroid-tapering therapy was applied and the patient's state improved. After 3 years of observation, the patient's state is stable, with no abnormalities in neurological examination.

Keywords: tectal plate tumors; brain stem tumors; the Parinaud syndrome; seizures

STRESZCZENIE

Guzy blaszki czworaczej to grupa guzów pnia mózgu, które są rzadkie wśród dorosłych. Opisujemy przypadek guza blaszki czworaczej u młodej osoby dorosłej o nietypowej prezentacji. W wieku 14 lat pacjentce założono dren komorowo-otrzewnowy z powodu objawów wzmożonego ciśnienia śródczaszkowego i wodogłowia o nieustalonej etiologii. W wieku 19 lat wystąpiły nawracające napady drgawkowe częściowe złożone (włączając napady gromadne), w związku z czym włączono leczenie kwasem walproinowym. Drgawki ustąpiły, ale zaobserwowano szybko postępujące upośledzenie funkcji poznawczych i spowolnienie psychoruchowe. W badaniu neurologicznym obecne były bradyfrenia, bradykinezja i zespół Parinaud. Badanie MR głowy uwidoczniło guza blaszki czworaczej bez aktywnego wodogłowia. W spektroskopii rezonansu magnetycznego obraz morfologiczny sugerował glejaka o niskim stopniu złośliwości. Zastosowano sterydoterapię w zmniejszających się dawkach i stan neurologiczny pacjentki uległ poprawie. Po 3 latach obserwacji stan pacjentki pozostaje stabilny, nie ma żadnych odchyleń w badaniu neurologicznym.

Słowa kluczowe: guzy blaszki czworaczej; guzy pnia mózgu; zespół Parinaud; drgawki