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Intracranial Rhabdomyosarcoma Presented as Chronic Subdural Hematoma: A Case Report

Intracranial rhabdomyosarcoma is a very rare disorder. Subdural rhabdomyosarcoma has not been reported yet. It can be misdiagnosed with chronic subdural hematoma in CT images. Herein, we presented a 2.5-year-old boy with intractable chronic subdural hematoma who were treated with burr hole insertion, needle aspiration and shunting for about 2 years with partial resolution of his symptoms after each procedure. The final pathologic evaluation after extensive bilateral craniotomy revealed rhabdomyosarcoma. The role of CT and MRI in early diagnosis and management of this rare situation is discussed.

Keywords: chronic subdural hematoma, magnetic resonance imaging, rhabdomyosarcoma

Introduction

Chronic subdural hematomas (CSDH) are commonly seen in the infancy. The peak incidence is about six months of age. Chronic subdural fluid collections are rarely reported after one year of age. Certain traumatic events that occur in infancy usually remain unrecognized or unreported. The infants rarely have an underlying bleeding disorder or hematologic malignancy.¹ Computed tomography (CT) has been an established radiological modality for the diagnosis during recent years. However, there are certain pitfalls in CT diagnosis.² From the clinical point of view, the dilemma of differentiation between chronic subdural hematomas and subdural mass lesions such as primary or metastatic tumors has been of utmost importance but difficult to resolve with brain CT. Primary subdural sarcomas are extremely rare, and their association with subdural hematoma is even more unusual. Few cases have been reported in the literature.³ Here we reported on a case of subdural rhabdomyosarcoma that was initially managed as CSDH. The importance of magnetic resonance imaging (MRI) in the differential diagnosis of refractory CSDH is also emphasized.

Case Presentation

A 40-day-old boy was admitted to Children Hospital in Tehran, with coma and intractable seizure after severe head trauma which was first diagnosed by CT as acute subdural hematoma and frontal contusion (Fig. 1). The symptoms resolved with conservative treatments. The second CT five days later confirmed resorption process in the former hematoma and an obvious expansion of extra-axial fluid in both hemicranial spaces (Fig. 2). He was well for several weeks after he discharged from the hospital. Then, he developed progressive increasing of head circumference, vomiting and poor feeding. Another brain CT was performed that

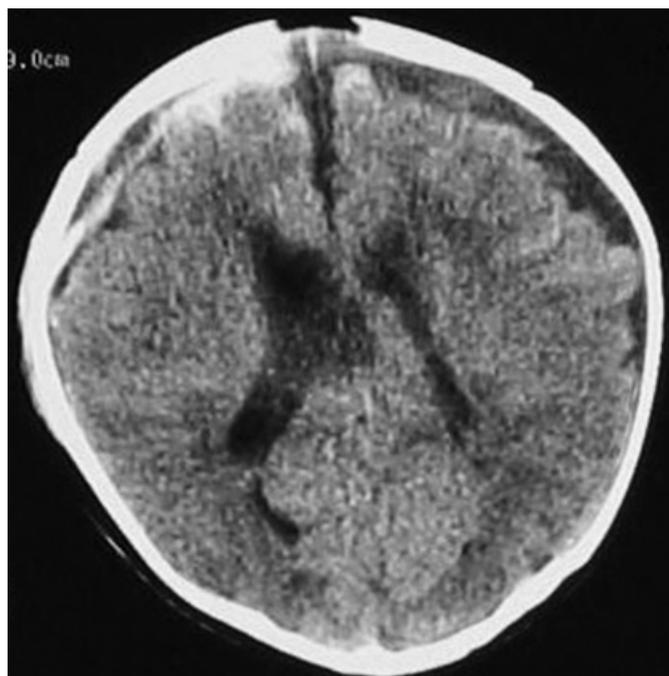


Fig. 1. Axial brain CT just after trauma reveals right frontal contusion and a small rim of acute subdural hematoma.

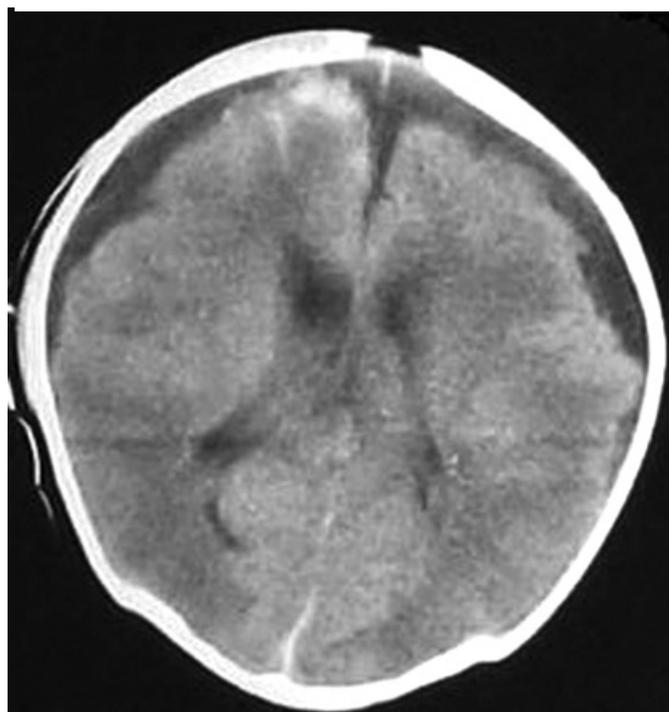


Fig. 2. The second CT performed five days later shows the process of resorption of the acute hematoma and the bilateral expansion of frontal subdural space.

typically causing sulcal effacement and ventricular compression (Fig. 3). The patient was managed with needle aspiration of the fluid, but his symptoms of intracranial hypertension returned back after several weeks. Different treatment modalities known for CSDH—needle aspiration, burr hole drainage, and

showed more expansion of the above-mentioned extra-axial fluid shunting—were all applied in the repeated attacks of the disease which were separated from each other only for several weeks. The fluid was watery, slightly blood-tinged at the first sampling. It was changed to a xanthochromic, proteinaceous fluid and later to a xanthochromic thick gelatinous collection that could not be drained using the previous methods. All specimens obtained from the subdural cavity were negative for bacteria, without abnormal cell count or glucose level, with rising level of protein in serial fluid samplings. The fluid was never sent for pathology. Peripheral blood examination, sedimentation rate and CRP were always normal.

Due to refractory CSDH, we had to perform craniotomy in order to approach the hematoma capsule. Brain CT before the operation confirmed the same lesion with severe compression of the underlying cortex and small nodules at the parietal capsule (Fig. 4). Extensive bilateral frontotemporoparietal craniotomy was done at the age of two, which surprisingly revealed a huge lobulated friable subdural mass, pinkish-gray, with low vascularity, thoroughly separated from dura matter and the underlying cortex. Histopathological findings and immunoperoxidase staining of the tumor, together, led to the diagnosis of rhabdomyosarcoma, embryonal type (Fig. 5). The child was referred to an oncologist for further treatment. There was a delay in starting the adjuvant therapy, consequently he developed previous symptoms. MRI without contrast done one month after gross total resection of the mass, revealed lobulated subdural mass, nonhomogeneously hypointense in T₁ and T₂ weighted images, which was not observed in previous CT images (Fig. 6). Standard radiotherapy was started. He was well only for two weeks after the last session of the radiotherapy, when he developed recurrence of all previous symptoms and recurrent seizures. Further treatment was refused by the parents. The patient passed away one month later.

Discussion

Development of an aggressive subdural mass in the setting of a CSDH is very rare. It is not our purpose to describe the etiology of this tumor and whether it existed initially or had developed subsequent to

chronic inflammation in CSDH. Herein, we would like to emphasize on the importance of MRI in the diagnosis of a subdural mass accompanying subdural hematoma that was not revealed by CT alone.

CT remains the primary imaging modality for diagnosing CSDH; this method is rapid and highly accurate. Over three quarters of hematomas are hypodense in appearance and many take on a lenticular shape. Such lesions that are isodense or hypodense on CT generally appear hyperintense on both T₁- and T₂-weighted images on MRI.¹ Uncertainties exist regarding strict diagnosis of subdural mass lesions with CT. CT shows subdural sarcoma to be hypodense, isodense, or slightly hyperdense. MRI is probably the method of choice for the evaluation of subdural sarcomas. The tumors are hypointense or isointense on T₁-weighted images and slightly hyperintense on T₂-weighted images—a finding that may reflect hypercellularity and correlate with a high grade of malignancy. All tumors show some enhancement following intravenous gadolinium administration which is most commonly solid and homogeneous.¹

There are several case reports that describe subdural mass lesions first diagnosed as subdural hematoma. Cinalli, *et al*, presented two infants with progressive macrocrania related to chronic pericerebral fluid collection. They were managed as subdural hematoma but the final pathologic findings led to the diagnosis of liposarcoma and fibrohistiocytic sarcoma.³

Brooks, *et al*, reported a patient with subdural sarcomatous granuloma diagnosed by MRI, where the value of this imaging modality in anatomical evaluation of the lesion and neurosurgical planning was advised.⁴ In a report of primary leptomeningeal lymphoma, it was recommended to perform proton-density MRI when progressive neurological abnormalities were found in patients with subdural hematoma.⁵ Other reports regarding the role of MRI are also available in patients with complicated arachnoid cyst, meningioma, dural liposarcoma, meningeal sarcoma, intracranial granulocytic sarcoma (chloroma) and malignant fibrous histiocytoma mimicking subdural hematoma in CT images.⁶⁻¹¹

Our results along with review of the literature demonstrated that subdural soft tissue mass may be confused with CSDH in CT images in many patients. Misdiagnosis of these patients results in delayed

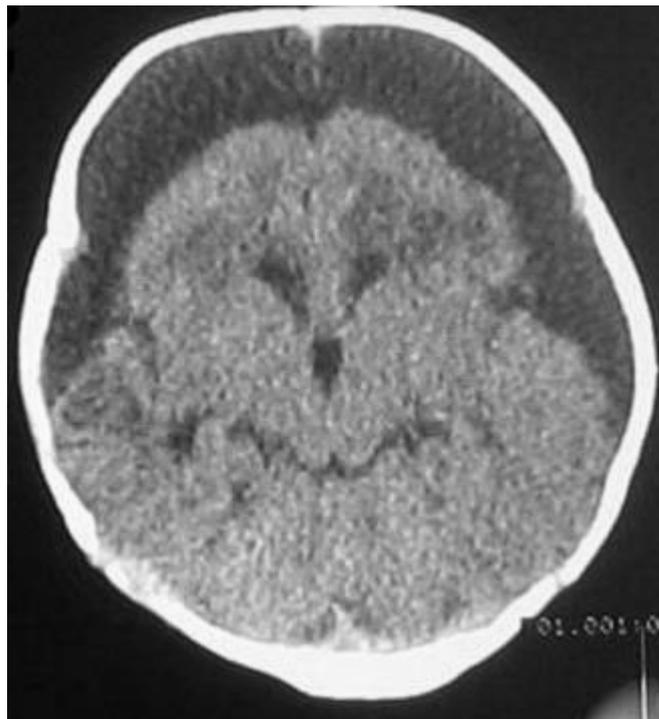


Fig. 3. Brain CT performed six months after trauma demonstrates obvious expansion of bilateral frontal subdural space. The fluid aspirated from this space was a blood tinged liquid.

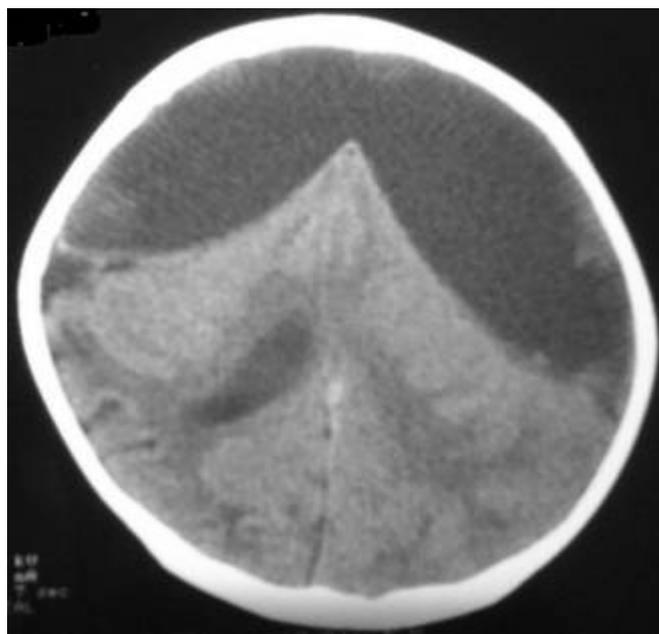


Fig. 4. Brain CT before the last operation confirms severe compression of the underlying cortex and small nodules at the parietal capsule. The fluid obtained during tumor mass removal was a thick yellow gelatinous material.

treatment and poor prognosis. Previous studies have emphasized the superiority of MRI to reveal the fine tumor mass details and the hematoma associated with the lesion compared to CT. It was concluded that MRI is a suitable method for differentiating soft tissue

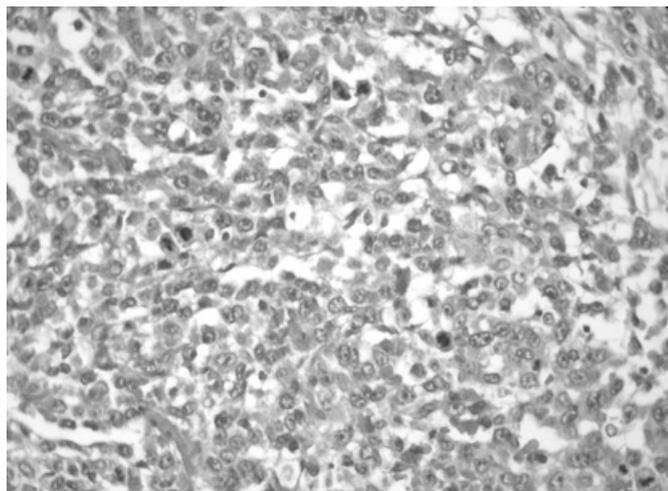


Fig. 5. Photomicrograph of the tumor. Microscopic features include pleomorphism with numerous typical and atypical mitotic figures, and fusiform cells with irregular, polylobulated nuclei and a high nuclear-to-cytoplasmic ratio (HE×100).



Fig. 6. T₂-weighted MRI reveals non-homogenous bilateral frontal lobulated hyper-intense mass originating mainly from parietal layer of subdural space accompanying with homogenous hyper-intense fluid collection. Sulcal effacement and cortical compression are also evident.

sarcomas from chronic traumatic hematoma.¹² Therefore, high resolution imaging modalities such as MRI

can possibly help clinicians facing this issue.

These findings suggest that in patients with refractory CSDH, MRI may help in diagnosis of the possible underlying subdural mass to offer appropriate treatment. Consequently, we recommend brain MRI as a routine protocol in patients with chronic subdural fluid collection that are resistant to conventional treatments to reveal sophisticated, rare and fatal conditions like primary and metastatic dural and subdural mass lesions.

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