Int J Clin Exp Pathol 2017;10(4):4545-4551
www.ijcep.com /ISSN:1936-2625/IJCEP0051775

Case Report

Post-traumatic embryonal rhabdomyosarcoma of the scalp: report of two cases and review of the literature

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Received February 27, 2017; Accepted March 24, 2017; Epub April 1, 2017; Published April 15, 2017

Abstract: Embryonal rhabdomyosarcoma (ERMS) development after trauma is a rare occurrence. We first report two cases of head ERMS that developed in the scar of head trauma. The first is that of a 12-year-old boy who was incorrectly considered as hematoma calcification at the first stage because of his shorter head traumatic history (56 days) and the relatively stable feature of mass. Preliminary CT scan and MRI results did not fulfill the diagnosis of malignant tumor. But further pathological examination in the resected tissue demonstrated the feature of lesion and referred to ERMS, leading to the second radical excision of the tumor and further interstitial iodine 125 intervene in the region and adjuvant chemotherapy, with a good prognosis at last. The second case concerns a 10-year-old girl who was same with a head injury history (about 2 months) before presenting a mass in her forehead. However, the volume of mass significantly increased in the few months followed by severe headache and instable walking. MRI and PET-CT both showed local malignant lesion with systemic metastasis. Pathology further confirmed the diagnosis of ERMS. The girl died soon after abandoning treatment. The presented cases suggest that traumatic cerebral lesions may also be a predisposing factor for the development of ERMS and reminder us not to forget the possibility of tumor formation after head injury, especially for these posttraumatic adolescents. As stated by other authors, an association between head trauma and tumor risk cannot be ruled out and should be studied further.

Keywords: Head injury, posttraumatic tumor, embryonal rhabdomyosarcoma

Introduction

The relationship between head injury and the development of tumors has been controversy so far [1, 2]. Previously, several cases have been reported the relationship between traumatic head injury and the development of intracranial tumors, such as glioma [3-8]. However, there have been seldom cases about the incidence between traumatic head injury and embryonal rhabdomyosarcoma (ERMS) of scalp. Here we first report two cases of scalp ERMS that developed in adolescents with previous head trauma, which adds further support to the relationship between trauma and tumor.

Case 1

A 12-year-old boy without previous family history of physical health presented with a traumatic mass in his left temporal for 56 days (**Figure 1A**). Local hot compress was intermittently treated but the mass was not reduced. Computed tomography (CT) showed some clump-shaped mixed density in the left temporal muscle (**Figure 2A**) and magnetic resonance imaging (MRI) documented a large heterogeneous annular contrast-enhancing lesion in the same region (**Figure 2B**). Admission diagnosis was traumatic hematoma calcification, and then subcutaneous mass in the left temporal was resected under local anesthesia. Intraoperative seeing, the lesion in the temporalis muscle was gray, solid, rich blood supplied, with the size of about 3.3 cm × 3.3 cm × 4 cm and significant adhesion to the surrounding tissue. When peeling the tumor, its capsule was damaged with significant hemorrhage and white granulation tissue exposing. Postoperative pathological results showed that pleomorphic fea-

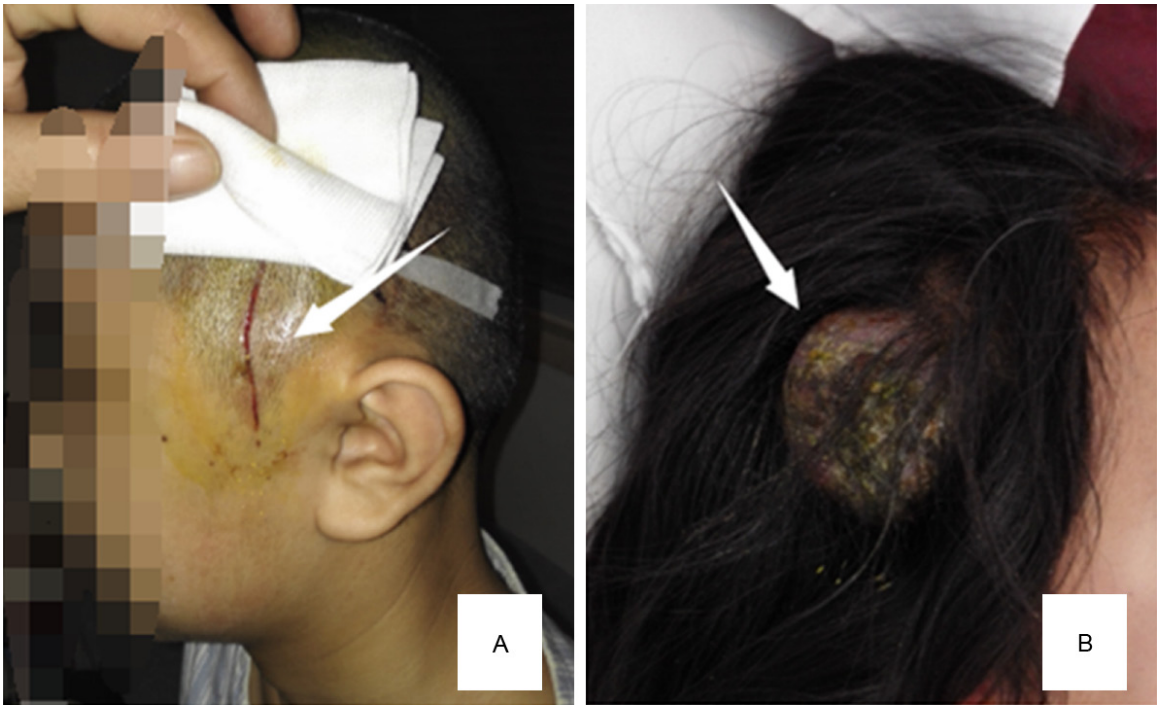

Figure 1. Large subcutaneous mass in the two cases. A. Showed the location of the mass and incision located in his left temporal after the first operation. B. Showed a huge mass in her right forehead and its diameter was up to 5 cm with apparent ulceration.

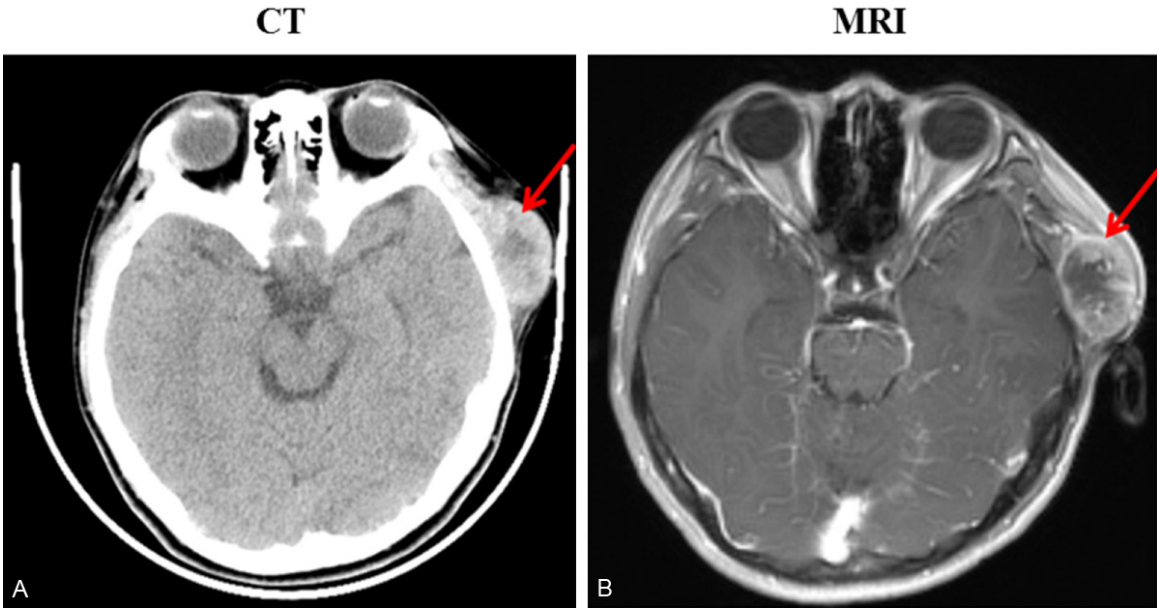

Figure 2. Radiological pictures of the lesion in case 1. A: Computed tomography (CT) scan showed some clump-shaped mixed density in the left temporal muscle (red arrow). B: Magnetic resonance imaging (MRI) revealed a large heterogeneous annular contrast-enhancing lesion in the same region. Hemorrhage and necrosis in lesions was also evident (red arrow).

tures with multiple atypical mitoses, sparse arrangement, rich mucus matrix and longitudinal muscle fibers or stripes were observed and the histopathological diagnosis was embryonal rhabdomyosarcoma (**Figure 4A**). Immunohistochemical result (**Figure 5**): CK(-), Vimentin(+),

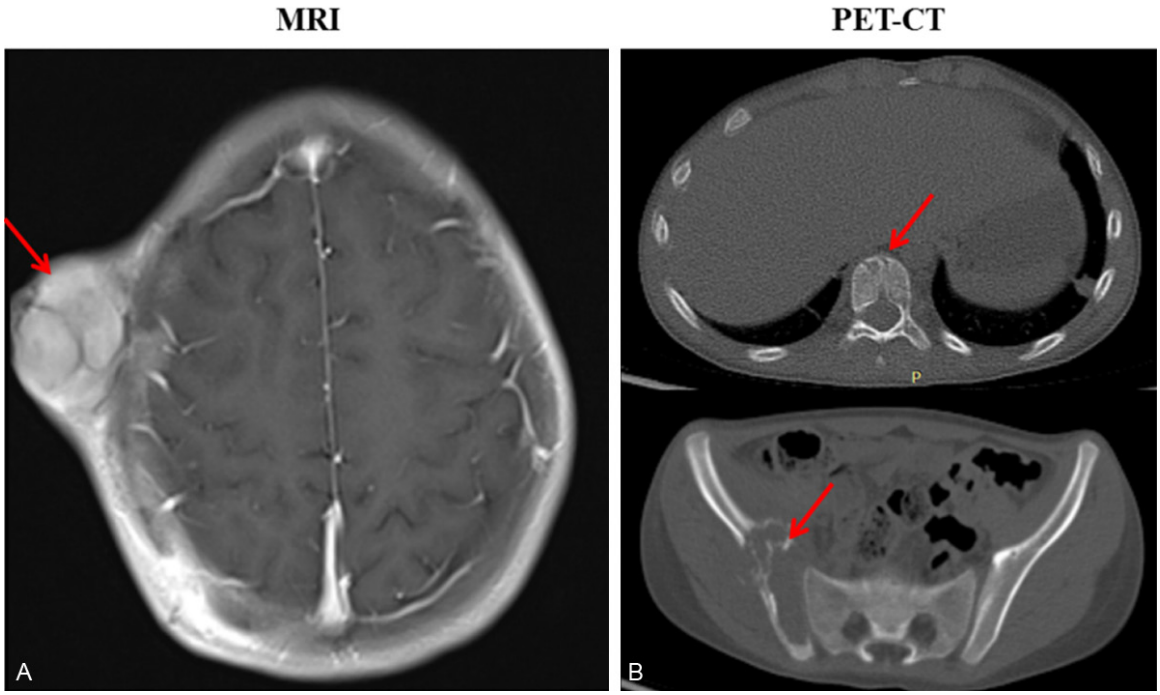

Figure 3. Radiological pictures of the lesion in case 2. A: Magnetic resonance imaging (MRI) revealed a large contrast-enhancing mass in the right frontal temporal region and multiple abnormal signals in the surrounding neck and ear soft tissues. Hemorrhage and necrosis in lesions were also evident (red arrow). B: PET/CT showed systemic multiple bone destruction with active metabolism, including vertebral body and pelvis (red arrow).

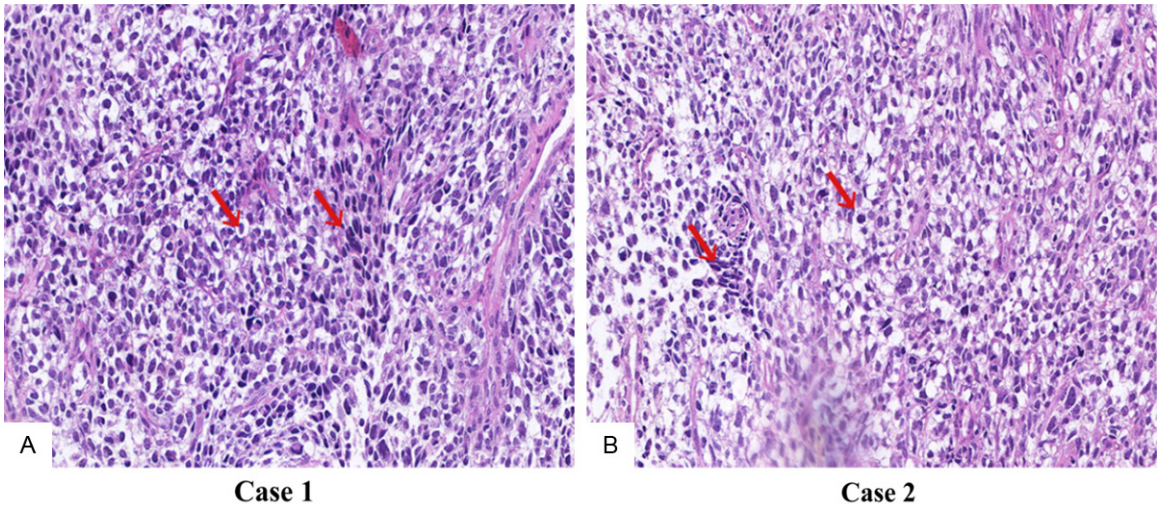

Figure 4. Histopathologic specimen demonstrating multiple atypical mitoses, sparse arrangement and rich mucus matrix with longitudinal muscle fibers or stripes (HE × 200, red arrow). (A) refer to case 1 and (B) refer to case 2.

Desmin(+), MyoD1(+), Myogenin(+), S-100(-), LCA(-). CD34(-), CD30(-), SMA(-), Calponin(-), CD99(-), CD56(+), Syn(-), Fli-1(+), Ki-67(+50%). Considering the malignant neoplasm, the second radical operation "maxillofacial neck mass resection and fascia flap plasty" was underwent in the Department of Oral and Maxillofacial Surgery after 10 days of the first operation under general anesthesia. The incision was about 6 cm from the hairline along the temporal front down the ear to the ear lobe, then cut the skin, subcutaneous tissue, separated the superficial temporal fascia, flipped the flap from the parotid gland muscle fascia and the tumor

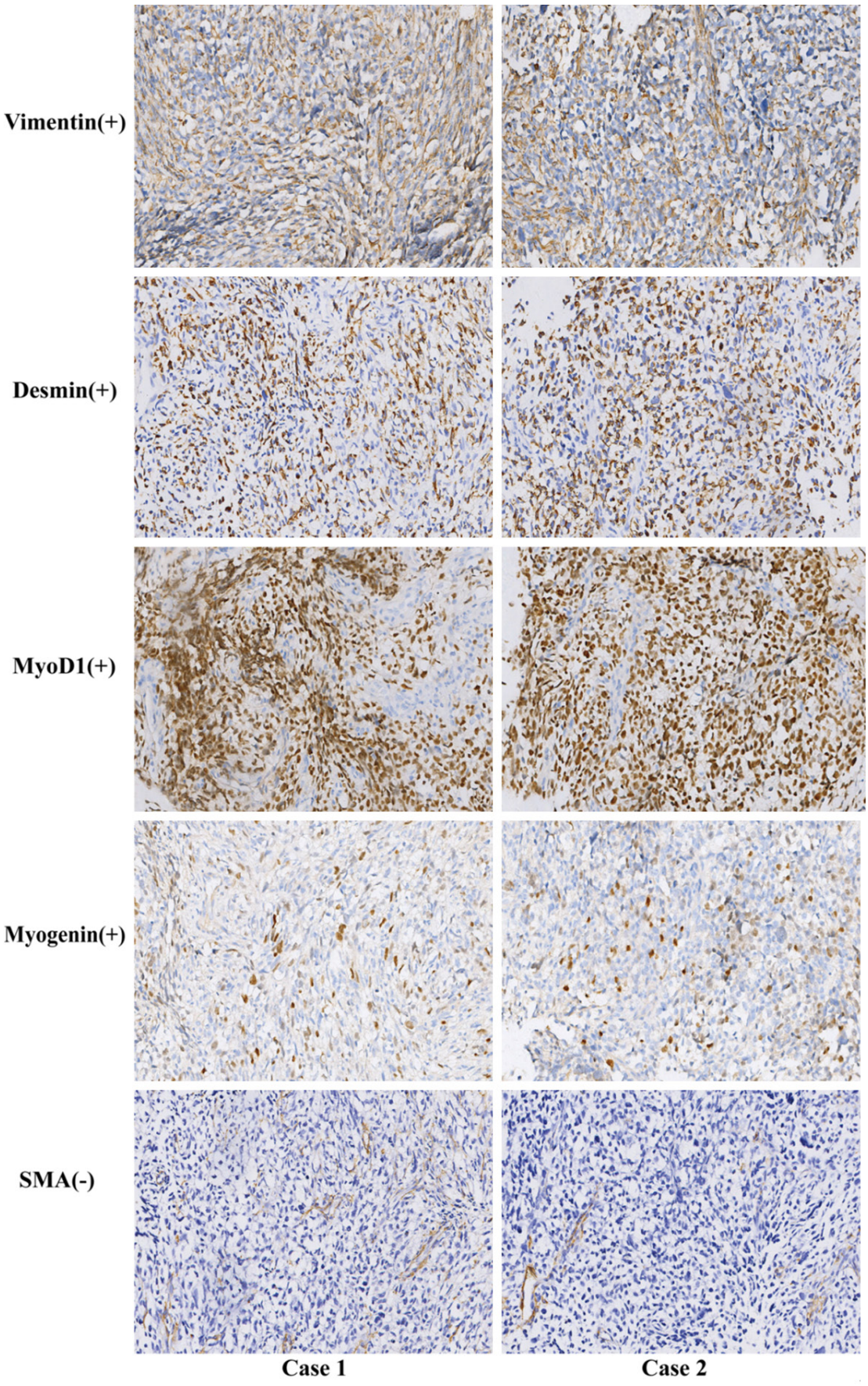
Vimentin(+)
Desmin(+)
MyoD1(+)
Myogenin(+)
SMA(-)
Case 1
Case 2

Figure 5. Immunohistochemistry of ERMS. Both cases show the strong positive staining with Vimentin(+), Desmin(+), Myogenin(+), MyoD1(+), and negative staining with SMA(-). Microscopic Image at 200 ×.

was seen to be solid, without capsule and clear surgical margins. Complete removal was conducted after dissociation, followed by the interstitial iodine 125 intervene to enhance the curative effect. After surgical excision, two cycles of dactinomycin-based chemotherapy were also performed. The patient healed without surgical complications and was discharged two weeks after the operation. At present, six months postoperatively, the patient is tumor-free.

Case 2

A 10-year-old girl was admitted to our hospital because of a significantly increasing mass in her right forehead obtained from a head injury about 2 months ago (**Figure 1B**). The mass had a brief reduction in short, but then its diameter sharply increased to 5cm with apparent ulceration. She complained about a persistent frontal headache, instable walking for 2 weeks and a novel mass in her right neck for 1 week. MRI after appearance of the current symptoms revealed a large contrast-enhancing mass in the right frontal temporal region and multiple abnormal signals in the surrounding neck and ear soft tissues (**Figure 3A**). PET/CT showed systemic multiple bone destruction with active metabolism, including vertebral body and pelvis, suggesting the malignant lesions of systemic metastasis (**Figure 3B**). Further fine needle puncture cytology examination showed that the histopathological diagnosis was also embryonal rhabdomyosarcoma (**Figures 4B** and **5**). Owing to the systemic metastasis, her parents gave up treatment and she died 3 months later.

Discussion

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma (STS) in children and adolescents. The incidence of RMS is 4.5 cases per million children per year, and the reported female: male ratio is 1.5:1 [9]. According to the present classification system at the IRS (The International Classification), RMS is divided into 5 subtypes, with 2 most predominant referred as to embryonal (ERMS) and alveolar (ARMS), accounting for about 60% and 25% of all RMS, respectively, while the remaining are classified as botryoid RMS, spindle cell RMS, and undifferentiated sarcomas [10]. Of them, ERMS commonly (about 35%) presents in the head and neck (including orbit and parameningeal sites like nasopharynx, nose, and paranasal sinuses, middle ear cleft, infratemporal fossa, pterygopalatine fossa, and the parapharyngeal space) and genitourinary areas of patients younger than 10 years of age. Besides, ERMS is highly heterogeneous and characterized by loss of heterozygosity on the short arm of chromosome 11 (11p15.5), suggesting inactivation of a tumor-suppressor gene [11-13]. Patients who have localized RMS have a 5-year survival greater than 70% following a multimodal approach that includes chemotherapy, radiation therapy, and surgery; however, overall survival of patients with metastasis still remains poor [14].

However, there are seldom reports about head injury and ERMS, especially the head injury at first stage and subcutaneous ERMS. Most of previous case reports are about the old intracranial injury and brain tumor formation, including glioma [3-8], Pott's puffy tumor [15], lipoma [16], meningiomas [17] and so on. Only Yeung [18] reported a case of anaplastic lymphoma kinase negative anaplastic large cell lymphoma manifesting as a scalp hematoma after an acute head injury. As for EMRS, only Nejat F [19] presented a case of subdural rhabdomyosarcoma that developed in a chronic refractory subdural hematoma of a 40-day-old boy with a head trauma. The patient repeatedly underwent different surgical interventions over 2 years. Finally, extensive bilateral front temporoparietal craniotomy was performed at the age of 30 months. Pathological examination confirmed the diagnosis of ERMS. Radiation therapy was performed, but the tumor recurred and the child died at the age of 3 years.

Here we first reported two cases that occurred an ERMS of scalp soon after a head trauma. The current medical community is still inconclusive about whether the trauma could act as a causative factor in tumorigenesis. It is generally believed that the occurrence of tumors and trauma has three relationships (1) tumor results from a natural factor and is irrelevant to trau-

ma; (2) tumor owes to an acquired factor induced by trauma because trauma could make local tissue degeneration, necrosis and the following immune clearance, new cells proliferation, repair and so on, which could make some locally metabolic changes and result in malignant transformation, such as endogenous carcinogen activation and tumor-suppressing gene dysfunction; (3) congenital factors and acquired factors both play roles in the tumor formation. Tumor cells may germinate in the embryonic period but are activated by trauma, leading their rapid proliferation and differentiation [20].

The two patients presented in our cases were diagnosed at 12 and 10 years old respectively, and both of the masses occurred in the scalp. However, the two patients were both in good health without genetic disease before the head injury and the tumors presented in the damaged regions after the head injury, which may suggest there are some relationship between head injury and ERMS. Furthermore, radiology and pathology both confirmed the diagnosis of ERMS. But the subcutaneous tumors did not meet the currently established criteria by Zulch et al [20] and Manuelidis et al [21] for accepting the traumatic origin of some intracranial tumors because of the relatively shorter time intervals, about average 58 days, which is less than the standard one more year. Maybe it could be explained by the highly malignant tumor cells in circulation accumulating at the injured site after trauma and forming a scalp ERMS at an alarming rate.

In conclusion, although posttraumatic embryonal rhabdomyosarcoma of scalp has seldom been described in the literature, its occurrence is possible, because trauma can trigger abnormal cell proliferation and differentiation. In our view, traumatic injuries may be a predisposing factor for ERMS, especially in the adolescents. The two cases may reminder us not to forget the possibility of tumor formation after head injury, especially for these posttraumatic adolescents, instead of incorrectly considered as the hematoma calcification.

Acknowledgements

This project was supported by grants from the National Natural Science Foundation of China (No.81572630, 81171177).

Disclosure of conflict of interest

None.

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