CT and multimodal imaging findings of primary orbital Ewing's sarcoma involving the middle cranial fossa: a case report

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Abstract

This report details the CT and MR imaging findings of a primary orbital Ewing's sarcoma case involving the middle cranial fossa in a 13-year old boy. CT showed an ill-circumscribed homogeneous soft-tissue density mass with needle-like bone reaction. On MRI, the mass showed homogeneous iso-intensity on T1WI, heterogeneous hyper-intensity on T2WI, and marked homogeneous enhancement. Meanwhile, the mass was hyper-intense on DWI, and ADC value was $575 \times 10^{-6}$ mm$^2$/s. The mass showed as high rCBV and rCBF, prolonged MTT based on DCS-PWI, and wash-out pattern of TIC derived from the DCE-MRI. Our case suggests that functional MRI modalities, including DWI, DSC-PWI or DCE-MR, could provide additional information for differential diagnoses. Both CT and MRI should be performed and comprehensively analyzed for limiting differential diagnoses, determining lesion extension and facilitating operative approach.

Keywords: Ewing's sarcoma, orbit, child, computed tomography, magnetic resonance imaging

Introduction

Ewing's sarcoma (ES) is a malignant neoplasm that mostly arises in the skeleton and was first described by James Ewing in 1921[1]. Skeletal Ewing's sarcoma typically involves the long bones and rarely occurs in the orbit. To date, fewer than 20 cases of orbital ES have been reported in the English literature and most reports focused on pathological or clinical finding[1-3]. To our knowledge, the CT or MR imaging (MRI) findings of orbital ESs have not been reported. Therefore, we report the CT and MRI findings of a primary orbit ES involving the middle cranial fossa in a 13-year old boy. This case report was approved by the Institutional Review Board in our hospital according to the CARE guidelines.

Case report

A 13-year-old boy, who presented with one month history of swelling of the right eye, was admitted to our hospital in 2014. He denied any past medical problems, especially neurological or ophthalmological complaints. Physical examination revealed proptosis of the right eyeball. The ophthalmologic evaluation was otherwise normal.
**Fig. 1 Conventional MR images and ADC map of the mass.** A–C: MR images reveal an intra-orbital mass manifesting as homogeneous iso-intensity on T1WI and heterogeneous hyper-intensity on T2WI, and homogeneous markedly enhancement after contrast administration (white arrow). D: The mass shows as low-value on ADC map (white arrow).

**Fig. 2 PWI and TIC of the mass.** A–C: PWI indicates that the mass shows as high CBV, high CBF, and prolonged MTT map (white arrow). D: The TIC shows a wash-out pattern.
The MRI examination performed 4 days before hospitalization showed a 30×30×30 mm irregular mass located in the superior lateral quadrant of the right orbit. The mass extended to the middle cranial fossa; however, the brain seemed not to be involved based on the T1 weighted images (T1WI) after contrast administration. The boundary between mass and brain tissue seemed clear and no obvious enhancement was found on the adjacent brain tissue. Additionally, the mass did not involve the lateral rectus and the posterior wall of the eye ball, although these two structures were being heavily pushed. Compared with the adjacent muscles, the mass showed homogeneous iso-intensity on T1WI (Fig. 1A), heterogeneous hyper-intensity on T2 weighted images (T2WI) (Fig. 1B), and marked homogeneous enhancement after administration of gadolinium-diethylene triamine pentaacetic acid (Gd-DTPA) (Fig. 1C). In addition, diffusion weighted imaging (DWI), dynamic susceptibility contrast perfusion weighted imaging (DSC-PWI) and dynamic contrast-enhanced MR (DCE-MR) were performed. The mass showed as hyper-intensity on DWI, low apparent diffusion coefficient (ADC) value on ADC map (Fig. 1D), and ADC value of the mass was 575×10 mm/s. It showed as hyper-perfusion on relative cerebral blood flow (CBF) map (Fig. 2A), relative cerebral blood volume (CBV) map (Fig. 2B), and prolonged mean transit time (MTT) map derived from DSC-PWI (Fig. 2C). The time intensity curve (TIC) derived from the DCE-MR images showed as a wash-out curve (Fig. 2D).

Computed tomography (CT) scan 5 days after hospitalization showed an irregular intra-orbital mass that involved the superior lateral quadrant of the right orbit and the greater wing of the sphenoid bone. The mass showed as iso-density and the CT value of the mass was about 57 Hounsfield units (HU). Obvious radial and needle-like bone reaction was noted (Fig. 3).

Six days after hospitalization, the mass was removed. Gross examination revealed a gelatinous and gray soft mass that measured about 30 mm in diameter; no obvious hemorrhage was seen. Microscopic examination showed that the tumor was composed of uniform, small and round cells (Fig. 4A). Immuno-histochemical stains disclosed CD99 (++) (Fig. 4B) and desmin (-) and GFAP (-).

The morphological and immunohistochemical results were consistent with the diagnosis of orbital ES. On the neck, chest, abdominal CT and brain MR images, no other lesions related to the Ewing’s sarcoma were observed. For this reason, the patient was diagnosed with primary orbital ES.

Discussion

Previous studies indicate that ESs occur in a peak age range of 5-13 years and 90% of cases occur by the age of 30 years[1]. The t (11; 22) (q24; q12) chromosomal translocation was found to be close with occurrence of ES, although the detailed pathogenesis of ES is not fully understood[4]. The transcriptional product of this translocation up-regulates insulin-like growth factor 1 (IGF1), which is a key factor in cell proliferation. The ESs were aggressive tumors with a high propensity for local recurrence and distant metastases. Metastases most frequently occur to the lungs, followed by the peritoneum, muscles and lymph nodes[4]. Till now, fewer than 20 primary orbital ES cases have been reported, and only a few reports mention imaging features of orbital ES, let alone the functional MRI features. Therefore, we report the CT and multimodal MR imaging features of a pediatric orbit ES case and focus on the diagnosis and differential diagnosis from the aspect of medical imaging.

As to the imaging features of ES, Somarouthu, et al. reported the imaging features of twenty-six extra-skeletal ESs[4]. They found that the extra-skeletal ESs
showed as an iso-density on CT, as a hypo- or iso-intensity on T1WI, and hyper-intensity on T2WI. Necrosis, hemorrhage and adjacent organ invasion could be partially detected, while calcification was never seen. As to the imaging features of orbital ES, only Kano, et al. and Naqvi, et al. mentioned it in their case reports that focused on the clinicopathologic features of orbital ES\cite{2-3}. The two irregular orbit ES masses in their case reports were located in the left lateral wall of the orbit, and the greater wing of the sphenoid bone was involved. The masses showed homogeneous enhancement on both CT and T1WI after contrast administration. The conventional imaging features of orbital ES in our case report were similar to their cases.

Besides the conventional MRI modalities, DWI, DSC-PWI and DCE-MR were performed in our case. Based on our case, we think that the orbital ES mass would show as hyper-intensity on DWI, hypo-intensity on ADC map, and hyper-perfusion on CBV, CBF and the MTT map which was derived from DSC-PWI. Meanwhile, the TIC based on the DCE-MR image would show as wash-out curve pattern. In our opinion, the hyper-perfusion of the tumor would be due to abundant vascularity within the tumor, and diffusion restriction of the tumor might be due to high cellularity within the tumor. To our knowledge, our report is the first case report that showed the multimodal functional MRI features of orbital ESs.

In view of the location and imaging features of orbital ESs, in our opinion, spheno-orbital meningioma is the most difficult and requires differential diagnosis. Using only conventional MR images, it is difficult to differentiate these two kinds of tumors. However, with spheno-orbital meningioma, associated expansion and sclerosis of the bone structure is commonly seen because of the benign nature of the tumor\cite{5}, while the radial and needle-like bone reaction shown on CT images may be a feature of orbital ES. DWI and TIC could aid in distinguishing these two tumors. The low ADC value and the washout curve are both indicative of malignant orbital tumors, which may assist in making the diagnosis of orbital ES\cite{6-7}. Besides meningioma, some extremely rare tumors, such as rhabdomyosarcoma, alveolar soft part sarcoma, or malignant fibrous histiotoma should also be considered.

Even though CT and multimodal MRI provide imaging information about orbital ES, definite diagnosis still could not be simply made based on CT or MRI. The definitive diagnosis of orbital ESs mainly depends on the pathology, especially immunohistochemical findings. In our opinion, the most important role of imaging modalities should be to determine lesion extension and facilitate the operative approach.

In conclusion, we present a rare orbital ES case from the aspect of medical imaging. Our orbital ES case showed as iso-density on CT, iso-intensity on T1WI, hyper-intensity on T2WI. The radial and needle-like bone reaction shown on CT images might be a distinguishing feature of orbital ESs. Functional MRI modalities, including DWI, DSC-PWI or DCE-MR,

\textbf{Fig. 4} \textit{Pathological results of the mass.} A–C: The tumor is composed of uniform, small and round cells (black arrow) (a, HE×100; c, HE×400). B–D Diffusely positive staining for CD99 (black arrow) (b, CD99×100; d, CD99×400).
could provide additional information for differential diagnoses. Both CT and MRI should be performed and comprehensively analyzed for limiting differential diagnoses, determining lesion extension and facilitating operative approach.

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