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Case Report

Primary mesenchymal chondrosarcoma of the orbit: Histopathological report of 3 pediatric cases

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Abstract

Mesenchymal chondrosarcoma (MCS) is an unusual tumor mainly found in the skeleton. Around third of the cases occur in extra-skeletal sites with the orbit being the third most common site in these cases. In previous reviews of the orbital cases, it has been concluded that orbital MCS tends to occur in women in the second or third decades of life. However, 8 cases of orbital MCS have been reported so far in the pediatric age group (age less than 18 years-old) one of which has been considered congenital MCS in a 5-days old newborn girl. We describe 3 additional pediatric cases with primary orbital MCS and they were all males. Our cases presented with proptosis and calcific orbital masses on imaging studies. Histopathological examination of the excised masses shared the typical presence of undifferentiated mesenchymal cells and immature areas of cartilage. The diagnosis of MCS was further confirmed by immunohistochemical staining. Brief review of the literature in relation to this diagnosis in the orbit is also presented.

Keywords: Mesenchymal, Chondrosarcoma, Orbit, Proptosis, Cartilage

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Introduction

Mesenchymal chondrosarcoma (MCS), is an unusual tumor first described by Lichtenstein and Bernstein.[1] It is usually found in the skeleton, but around one third of the cases may also be found in extra-skeletal sites.[2,3] Orbital MCS was first described by Cardenas-Ramirez in 1959.[4] Subsequently, reports of orbital involvement by this neoplasm were published reaching 28 cases mentioned in the English-written literature mostly in adults.[5] As the orbit is a rare site of mesenchymal chondrosarcoma, here we report 3 pediatric cases with primary orbital MCS.

Case reports

Case I

A thirty-month old healthy boy presented with progressive right eye proptosis of 1 month duration. Magnetic resonance imaging was done and revealed right orbital mass. He was

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then referred to our institution with a provisional diagnosis of rhabdomyosarcoma for further management.

At presentation, he had right eye proptosis with hypoglobus and complete restriction of extraocular movements (frozen globe). Pupils demonstrated a 4+ afferent pupillary defect in the right eye. Anterior segment examination of both eyes was unremarkable. Fundus examination of the right eye showed a healthy optic disc, macula, tortuous retinal blood vessels and choroidal folds indicating a mass effect.

Physical examination showed two café-au-lait spots on the right arm and upper right chest.

Blood investigations included complete blood count, hemoglobin levels, urea and electrolytes and blood sugar which were all within normal limits.

Computed tomography (CT) scan revealed right orbital intraconal mass of homogeneous density filling the retrobulbar space of the right orbit and extending along the optic nerve. The mass caused expansion of the bony orbit with ill-defined medial wall.

Magnetic resonance imaging (MRI) was done and showed a right partially lobulated intraconal mass with an extension to the extraconal spaces, particularly on the medial aspect with remodeling of the lamina paprecea. On T1 and T2 weighted images, the lesion demonstrated isotonic signal intensity equal to the brain grey matter. Post-contrast images showed peripheral rim enhancement. The optic nerve has been evaluated during radiological imaging but it was not identified or visible in all obtained sequenced images. However, the canalicular and intracanalicular parts of the right optic nerve maintained normal morphology and signal intensity. These findings have led to a radiological suggested diagnosis of rhabdomyosarcoma or optic nerve glioma.

The patient was booked, for tumor excisional biopsy and debulking surgery. Intraoperatively, the tumor was found to be friable. On gross pathology, the specimen consisted of multiple, friable, pieces of tissue which aggregated to measure 55 × 30 mm in maximum dimensions.

Microscopic examinations showed predominantly undifferentiated mesenchymal round cells with hemangiopericytoma vascular pattern (Fig. 1A). Several areas of immature cartilage with focal calcifications were seen within a myxoid stroma (Fig. 1B). Immunohistochemical staining panel was performed. The tumor cells showed diffuse positivity for vimentin and showed focal expression of CD99, CD56 and NSE (Fig. 1C and D). S-100 stain was not performed in this case. A final diagnosis of undifferentiated malignant round cell tumor consistent with mesenchymal chondrosarcoma was made.

Case II

A 6-year-old healthy boy presented at Johns Hopkins healthcare center with his parents with a complaint that his right eye seemed bigger than the left. His vision was 20/20 in both eyes without correction. The extra-ocular motility was full. Both pupils were equal, round, and reactive to light. The right eye was displaced inferiorly by 4 mm. The anterior chamber examination was unremarkable.

Orbital ultrasound showed consistent findings with a dermoid cyst rather than a hemangioma or other vascular lesions (Fig. 2A). On CT scan, a large superior orbital mass with calcifications was identified.

The lesion was surgically excised. The gross pathology revealed a 30 × 15 × 11 mm, tan-colored mass with a smooth exterior surface. The lesion was bisected and the cut surface was firm with no evidence of cysts, hemorrhage or necrotic foci. The histopathological examination was identical in appearance to Case I with expression of CD99 by the

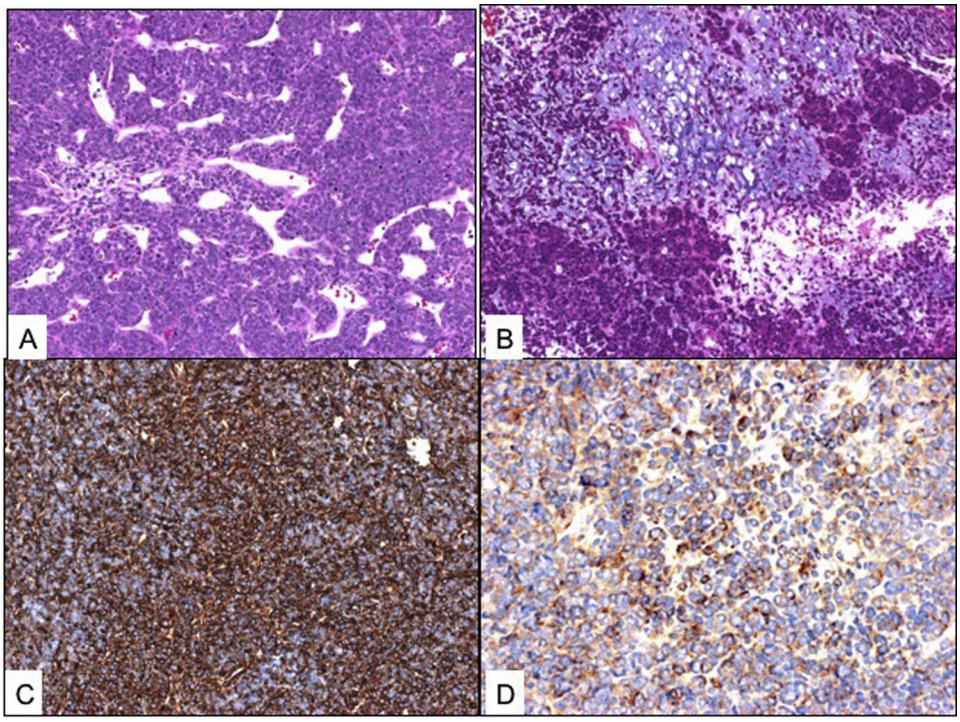

Fig. 1. (A) Tumor with hemangiopericytoma-like pattern in the first case (Original magnification x200 Hematoxylin & Eosin). (B) Ares of immature cartilage (Original magnification x200 Hematoxylin & Eosin). (C) Mesenchymal tumor cells expressing Vimentin (Original magnification x400). (D) Focal CD99 expression in noncartilaginous areas of the tumor (Original magnification x400).