Case Report

Orbito-cranial mesenchymal chondrosarcoma in a young female: A rare case report

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ABSTRACT

Mesenchymal chondrosarcoma (MCS) of the head and neck is highly aggressive malignant small round cell neoplasm with cartilaginous differentiation, often with a pericytomatous vascular pattern. It represents approximately 0.1% of all head and neck neoplasms. Chondrosarcoma of orbit is extremely rare. We report a case of orbito-cranial MCS in a 24-year-old female who presented with exophthalmos. Imaging revealed a large heterogenous lesion arising from right orbit with intracranial extension. Patient underwent right fronto-temporal craniotomy with tumor decompression from intracranial part and right orbit. Histopathologic examination and immunohistochemistry was suggestive of MCS. Postoperative residual disease was treated with chemotherapy showing partial response.

Key words: Chondrosarcoma, intracranial chondrosarcoma, mesenchymal chondrosarcoma, orbital chondrosarcoma

INTRODUCTION

Mesenchymal chondrosarcoma (MCS) of the head and neck is an uncommon tumor with potential for exhibiting highly aggressive behavior. This lesion is a malignant small round cell neoplasm with focal cartilaginous differentiation, often with a pericytomatous vascular pattern. It represents approximately 1% of all chondrosarcomas and approximately 0.1% of all head and neck neoplasms.[1,2] It is usually seen in younger age group compared to conventional chondrosarcomas.[2] In the head and neck region, it presents with a predilection for the maxillofacial skeleton, particularly in the mandible and maxilla.[3] Chondrosarcoma of orbit is extremely rare; only 18 cases have been reported until 2004.[4] We report a case of orbito-cranial MCS in a young female who presented with exophthalmos.

CASE REPORT

A 24-year-old female presented with a history of progressive proptosis of right eye along with headache and the progressive loss of vision in the same eye for 10 months. Physical examination revealed forward displacement of right eyeball and decreased visual acuity in the right side. An incisional biopsy from right orbital mass was performed at a private hospital, and histopathological examination showed well-differentiated cartilage and proliferation of small round to polygonal cells with mild pleomorphism suggesting possibility of MCS. Magnetic resonance imaging of brain and orbit revealed a large expansile heterogenous lesion arising from lateral wall of right orbit and right greater wing of sphenoid, predominantly hypointense on T1-weighted images and mildly hyperintense on T2-weighted images and measured approximately 8.5 cm × 7 cm × 6.5 cm and it showed intense inhomogeneous enhancement [Figure 1]. The orbital part of the tumor caused marked proptosis of right eyeball and displacement of the optic nerve infero-medially. Intracranial portion caused marked compression of right temporal and frontal lobes. Left orbit was normal. Patient underwent right fronto-temporal craniotomy with tumor decompression from intracranial part and right orbit. Histopathologic examination showed a tumor composed of small undifferentiated cells with hyperchromatic nuclei arranged around blood vessels in a hemangiopericytoma-like pattern and islands of cartilage suggesting MCS [Figure 2a and b]. Immunohistochemical analysis was positive for S-100, CD99 and vimentin and negative for cytokeratin. Postoperative contrast-enhanced computed tomography (CT) scan of brain and orbit

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1½ months after surgery revealed large highly enhancing soft tissue density mass near the floor of middle cranial fossa on right side involving the sphenoid, lateral wall of right orbit and posterior wall of right frontal sinus [Figure 3]. Patient was advised for external beam radiotherapy, but she refused it. Later, she was given six cycles of chemotherapy comprising of injection ifosfamide 2 g day 1 to day 4 intravenous infusion and injection doxorubicin 40 mg in day 1 and day 2 as shot push. CT scan evaluation after 1-month of completion of chemotherapy revealed partial response.

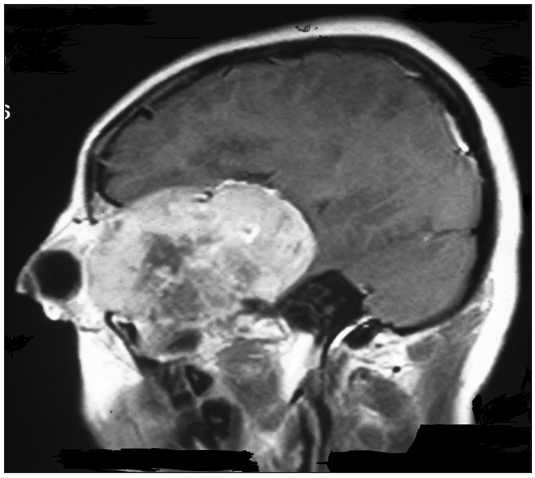

Figure 1: T1-weighted magnetic resonance image with contrast showing a large heterogeneous lesion arising from right orbit with intracranial extensiona - sagittal view

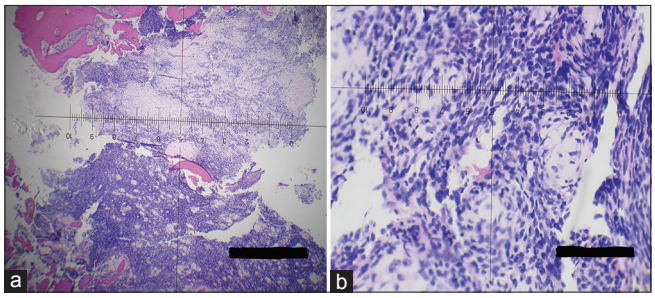

Figure 2: Histopathology showing small undifferentiated cells with hyperchromatic nuclei arranged around blood vessels in a hemangiopericytoma-like pattern and islands of cartilage (a) low power field, (b) high power field

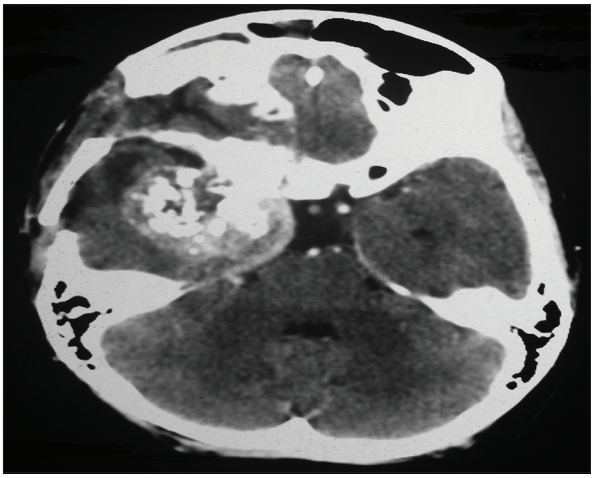

Figure 3: Postoperative computed tomography scan of brain and orbit revealed large soft tissue density mass near the floor of middle cranial fossa on the right side

DISCUSSION

The first case of MCS was described by Lightenstein and Bernstein in 1959.[5] This tumor tends to affect patients in their second or third decades of life, with a female preponderance.[1] Clinically, orbital MCS tend to present with progressive proptosis and/or abnormalities in vision, such as diplopia or reduction in visual acuity. CT scans usually reveal the presence of foci of calcification in the lesion. Histologically, this tumor exhibits a bimorphic appearance of undifferentiated mesenchymal cells with islands of mature hyaline cartilage.[6] Immunohistochemical analysis often reveals positivity for vimentin, S100 protein and CD99; while, actin, cytokeratin, and epithelial membrane antigen are typically negative.[7] The differential diagnosis of orbital MCS includes hemangiopericytoma, myxochondrosarcoma, osteosarcoma, and osteochondroma as primary tumors and lymphoma, neuroblastoma, synovial cell sarcoma, and chondrosarcoma as tumors affecting the orbit secondarily (direct invasion or metastasis).[8] This case presented no difficulty in the histopathological diagnosis since it showed a characteristic morphological pattern composed of round cells with an abrupt transition to well-differentiated hyaline cartilage and a hemangiopericytoma-like vascular pattern.

Surgical resection has been the main treatment modality for orbital MCS. Long-term survival with radical surgery seems to be favorable. Radiotherapy might play a role when accompanied with surgery.[2,9] Some authors used preoperative radiation therapy to reduce tumor bulk prior to radical resection, but it did not change the preoperative approach. There is evidence that postoperative radiotherapy demonstrates trend toward increased survival.[10] Chemotherapy has a limited role in the management of chondrosarcoma and should be used as an adjuvant therapy in cases with aggressive behavior with potential metastasis, rapid local recurrence, and high-grade lesions.[2,9]

The prognosis of patients with MCS is highly variable, ranging from a complete response and long-term survival to progressive disease with extensive metastasis. Among all types of chondrosarcoma, the mesenchymal histology carries a nearly 10-fold increase in 5-year mortality. The overall 5- and 10-year survival for patients with MCS, when considering all sites, is 55% and 27%, respectively.

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