Case Report

Orbital Mesenchymal Chondrosarcoma: Report of a Rare Tumor in a Nigerian Girl

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Abstract

The orbit (eye socket) is one of the rare extraskeletal sites of mesenchymal chondrosarcoma (MC). This is a report of the clinical presentation, the results of radiological and histopathological investigations, as well as the treatment outcome of a case of orbital MC in an 11-year-old Nigerian girl who presented with right eye protrusion and worsening vision. Examination of the right eye revealed lagophthalmos with marked proptosis of the globe. Right eye modified exenteration was done due to progressive proptosis, pain, and no perception to light. Histology revealed a biphasic neoplasm that is composed of well-formed hyaline cartilage and sheets of undifferentiated mesenchymal cells with a diagnosis of MC. She had four courses of cytotoxic chemotherapy, and was initially clinically stable on follow-up, but had a recurrence due to noncompletion of chemotherapy and radiotherapy. Orbital MC should be considered among young females who present with progressive proptosis, with or without pain, and worsening visual impairment.

Keywords: Chondrosarcoma, mesenchymal, Nigerian girl, orbit

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Introduction

Mesenchymal chondrosarcoma (MC) is one of the very rare soft tissue neoplasms, with skeletal sites being three times more commonly affected than the extraskeletal sites.[1,2] The orbit is one of the common sites of extraskeletal MC, and it is common in young females aged between 20 and 30 years.[1] This tumor is a malignant small round blue cell tumor with cartilaginous differentiation.[3,4] To the best of our knowledge, there has not been a reported case of orbital MC in Nigeria. We report a case of orbital MC in an 11-year-old Nigerian girl with a review of the literature.

Case Report

An 11-year-old girl who presented at the ophthalmic clinic of University of Ilorin Teaching Hospital, Ilorin Nigeria, with a 6-week history of progressive right eye protrusion with minimal reduction in vision [Figure 1]. A review of her medical history showed 2 years of noticeable mild protrusion. There was no history of trauma, no redness of the eye, and no diplopia. Equally, there were no other ophthalmic or constitutional symptoms.

Ocular examination revealed normal visual acuity for the unaffected left eye (6/6), while the right eye had marked reduction of vision (6/18). There was lagophthalmos of the right eye with marked proptosis of the globe, about 28 mm in the axial direction (using Hertel exophthalmometer). There was marked restriction of the extraocular muscle movement across all directions of gaze.

Orbital magnetic resonance imaging (MRI) was done which revealed intraorbital mass extending superiorly to the medial wall, and an impression of possible orbital abscess collection was made. Other ancillary investigations were

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not remarkable. Fine needle aspiration under anesthesia was done, and the sample which yielded a minimal bloody aspirate was sent for microscopy and cytology. She was commenced on intravenous antibiotics; metronidazole, ceftriaxone, and amoxicillin-clavulanic acid. She also had oral and topical analgesics; timolol gutt, diamox, and diclofenac. The aspirate microscopy and culture yielded no microbial growth. There were no viable cells seen on cytology.

The vision on the right eye worsened in the immediate postoperative period, possibly due to central artery occlusion as a result of very high orbital pressure. There was also no perception to light (NPL). The proptosis was equally progressive, and the cornea was edematous with associated severe eye pain.

The patient and her parents were then counseled for right eye modified exenteration due to progressive proptosis, pain, and NPL. At surgery, a firm-to-hard orbital mass filling more than three-quarter of the orbit was found. There was associated erosion of the medial, superior, and inferior orbital walls. The right globe was removed with the optic nerve. The tumor was cleared and removed in piecemeal, and the periosteum was equally excised. Immediate postoperative period was uncomplicated. The patient was discharged home, and follow-up visit at the clinic was scheduled.

The specimen submitted for histopathology consists of the eye globe with the optic nerve and fragmented grayish-white solid masses. Microscopic features [Figures 2-4] showed a biphasic neoplasm that is composed of well-formed hyaline cartilage and sheets of undifferentiated round to spindled mesenchymal cells. There are few pleomorphic, hyperchromatic chondrocytes within the chondroid matrix. The undifferentiated cells appear predominantly spindle with hyperchromatic small nuclei and few stellate cells. There are areas of ossification and calcification seen. Diagnosis of orbital MC was made.

She was subsequently worked up for cytotoxic chemotherapy after counseling. Assessment of liver and renal functions, as well as full blood count before administration of the cytotoxic drugs were within physiological limits. The cytotoxic chemotherapy regimen includes intravenous vincristine (1.5 mg/m^2), cyclophosphamide (1.5 mg/kg), and doxorubicin (60 mg/m^2). This was preceded by premedication with adequate hydration and intravenous ondansetron and dexamethasone. The patient had 4 monthly courses of the cytotoxics, and throughout this period, there was no history of adverse drug reaction.

She presented for follow-up 2 months after completion of the fourth course of cytotoxic chemotherapy and was clinically stable with no signs of any orbital growth and no signs of metastasis clinically [Figure 5]. She, however, presented again 8 months after the follow-up visit with filled orbit, extrusion of the artificial eye, and evidence of metastases. She was sent for radiotherapy, but this could not be done due to financial constraint. She was subsequently lost to follow-up.

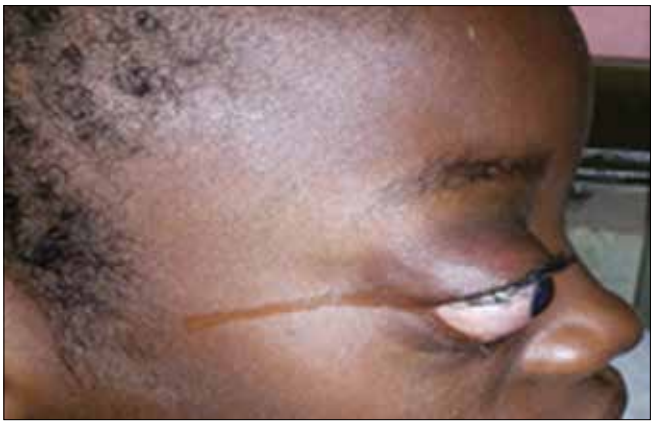

Figure 1: Right eye protrusion at admission

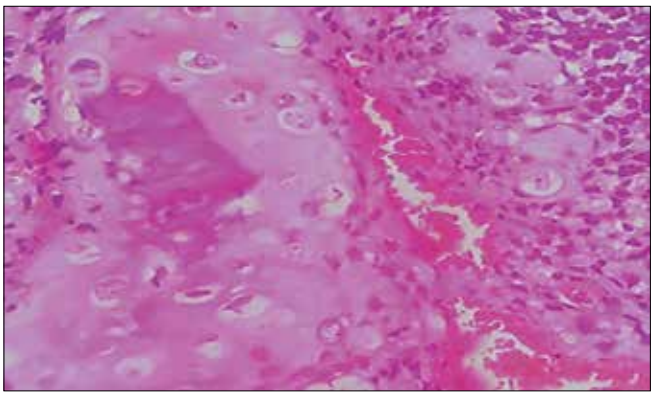

Figure 2: Histology micrograph section showing malignant undifferentiated mesenchymal cells with hyaline chondromyxoid stroma (H and E, ×400)

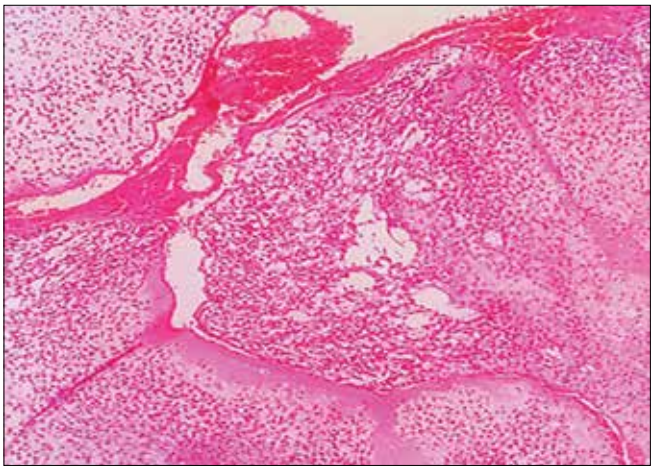

Figure 3: Histology micrograph section showing undifferentiated malignant mesenchymal cells admixed with chondromyxoid stroma (H and E, ×100)

Discussion

MC is a rare malignant neoplasm composed of primitive round to spindled mesenchymal cells with foci of chondroid differentiation.[2,3,5,6] Lichtenstein and Bernstein reported

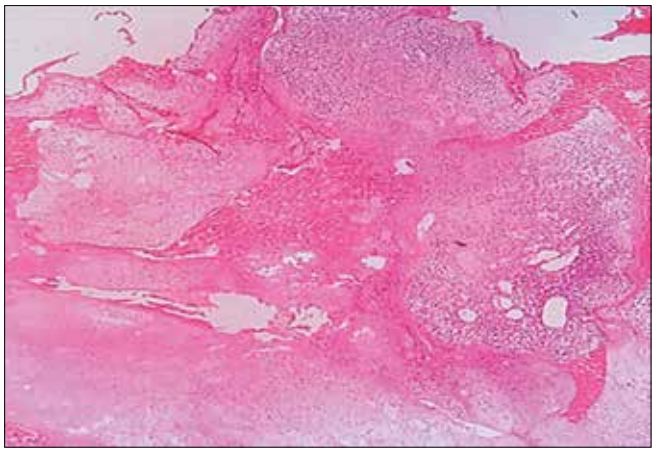

Figure 4: Histology micrograph section showing lower magnification view of the lesion (H and E, ×100)

the first case in 1959.[2,5,6] MC accounts for about 1%–8% of all chondrosarcomas,[2,5] and it is commonly seen in the mandible and maxilla among skeletal sites.[5,6] The meninges, mediastinum, orbit, and the visceral are the common reported extraskeletal sites.[5-7] Orbital MC is very rare, and up until 2018, only about thirty cases have been reported.[8] This is the first reported case in Nigeria, to the best of our knowledge.

Orbital MC tends to affect young female within the second and third decades of life.[3,9] Progressive proptosis and exophthalmos with or without pain are the common early symptoms, while progressive visual impairment is a late complication.[1,3,9-12]

Computerized tomography scan usually will demonstrate an isodense intraorbital mass with calcification.[3] Further, MRI will demonstrate isointense lesion with contrast enhancement.[1,13]

Histologically, MC is a high-grade sarcoma.[5-7] The tumor exhibits a biphasic pattern consisting of sheets of undifferentiated spindled/round small cells and areas of well-formed hyaline cartilage.[5,6] The undifferentiated mesenchymal area is usually hypercellular and typically has numerous vascular channels given hemangiopericytoma-like appearance.[5-7] Common differential diagnosis include dedifferentiated chondrosarcoma, Ewing's sarcoma (EWS) and its cohort, embryonal rhabdomyosarcoma, small cell osteosarcoma, and malignant lymphoma involving the bone. Usually, the biphasic pattern and hemangiopericytoma-like appearance with the typical age of occurrence are convincingly diagnostic of MC.[5,6]

By immunohistochemistry, MC is usually SOX-9 positive (a valuable marker of chondrogenesis), S-100 positive (variable and limited to the cartilalgenous areas), FLT-1 negative (positive in EWS), desmin, and osteocalcin negative.[4,5] In rare case of diagnostic difficulty, HEY1-NCOA2 and IRF2BP2-CDX1 gene fusion detection is highly confirmatory.[5]

Our patient is a girl child who had clinical symptoms strongly conforming to the clinical behavior of an intraorbital MC. Histologically, there was no difficulty in diagnosis; the tumor showed the typical biphasic pattern and the hemangiopericytoma-like appearance. The tumor responded well to cytotoxic chemotherapy initially with clean socket, and she was clinically stable with no signs of metastases 7 months after surgery and 2 months after completion of the fourth course of cytotoxics. However, the recurrence in this case about 12 months after the surgery was due to financial constraints because the parents did not have resources to pursue radiation therapy for the girl.

Figure 5: Picture of the right eye socket with artificial eye 2 months postcompletion of chemotherapy

Conclusion

Orbital MC is a very rare orbital tumor, which should be considered among young females that present with progressive proptosis, with or without pain, and worsening visual impairment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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