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Recurrent Orbital Rhabdomyosarcoma in a Two Years Child: A Case Study

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ABSTRACT

Introduction: *The most common childhood orbital malignancy is the rhabdomyosarcoma contributing almost 10% of all RMS cases. The mean age of orbital RMS is 6-8 years and rare case of RMS from birth to the 8th decade was reported. Male are more affected as compared to female. Total 60.4% male were affected with RMS as compared to female was 39.6%.* ***Objective:*** *The objective of the current study was to describe a case of recurrent orbital rhabdomyosarcoma and effectiveness of chemotherapy to RMS in a 2 year old child.* ***Methods:*** *Computer tomography, magnetic resonance images and excision biopsy were done for confirmation of tumor. After confirmation patient was treated with chemotherapy.* ***Results:*** *Excision biopsy showed tumor composed of round blue cells with high nodular cells ratio and hyper chromatic nuclei and finding also presented favors of Embreyonal rhabdomyosarcoma and child was treated with chemotherapy after completion of chemotherapy course this little child was came to his normal life but after 6 months sudden symptoms of recurrence has been appeared and this time tumor was most aggressive and cannot be controlled with only chemotherapy treatment.* ***Conclusion:*** *To achieve a good survival rate of RMS in children and elderly age there is a need to complete surgical tumor resection in anywhere of the body. Only chemotherapy should not be helpful and cannot be preventable from recurrence. Early diagnosis and stared treatment is the best approach and any delay in diagnosis can have a negative effect on health status.*

Keywords: Orbital malignancy, Rhabdomyosarcoma, Ophthalmology, Recurrent RMS, Childhood, Oncology

INTRODUCTION

Rhabdomyosarcoma (RMS) is a malignant tumor of soft tissue which originates from mesenchymal cells of primitive pluripotent. This tumor contributes 5% of all cancers of childhood. Every year in the United State 250-350 new cases of RMS has been reported; orbital involvement ratio was reported to be 10% [1]. RMS has been classified as small round blue cell tumor and it can be categoried histologically into 4 categories such as embryonal (ERMS) 55%, alveolar (ARMS) 20%, spindle, and botryoid (ERMS) 5% and undifferentiated pleomorphic sarcoma (USD) 20% [2]. Head and neck RMS affects 40% in children [3]. Orbit RMS is also the most common sarcoma of soft tissue which affects 4% of orbital malignances in all age of childhood, the mean age of orbital RMS is 6-8 years [4]. Approximately, 50% diagnosed solid malignancies has been reported in patients age under 10 years, on the other hand, RMS rarely affects adult age group [5]. Orbital tumor tissues usually originate from eyeball tissue (primary orbital tissue) and also from surrounded structures like eyelids, nasopharynx, paranasal tissue, and cranial cavity. RMS when became metastatic then this malignant tumor can be originating elsewhere in the body [6].

Objective

The objective of the current study was to describe a case of recurrent orbital rhabdomyosarcoma and effectiveness of chemotherapy to RMS in a 2 year old child.

Data Sources

Data sources to collect the relevant literature were used including Google scholar, PubMed, Cochrane, and Science direct.

Case Presentation

A 2 year old male child was presented in ophthalmological out patients Department of Shaikh Zayed hospital Lahore Pakistan, with compliant of left eye swelling and eye discharged as noted by patient mother. On physical examination, there was a hard immobile mass under the lower eye lid in the orbital cavity. MRI finding showing approximate 2.8 cm × 2.3 cm enhancing signal abnormality was seen involving left anterior ethmoid air cells disrupting lamina papyracea extending into medical aspect and floor of orbit with the involvement of preseptal space. The finding of MRI showed the infective process of probably fungal etiology and the patient was treated with antifungal but the swelling was not settled down. Excision biopsy was done and finding of excision biopsy showed tumor composed of round blue cells with high N/C ratio and hyper chromatic nuclei and finding also presented favors of embryonal rhabdomyosarcoma. After final biopsy approval chemotherapy has been stared. After 2 sanctions of chemotherapy MRI was repeated and finding of MRI showed there was significant interval decrease in size of soft tissue lesion in left orbit which was measured (1.0 × 0.5) after completion of 8 sanctions of chemotherapy, MRI was again repeated and finding of MRI this time showed no evidence of mass lesion in the orbital cavity.

Finding of CT scan abdomen, CT chest, CT brain, bone marrow biopsy, and bone scan showed no evidence of metastatic disease. So, 2 year old child came to his normal life after completion of 8 sanctions of chemotherapy. After 5-6 month's sudden symptoms of recurrence appeared and this time tumor showed its aggressiveness. Tumor size was increased measuring as 40 mm × 20 mm × 30 mm (AP × TRV × CC) along medial wall of left orbit causing moderate proptosis. The mass was extending antero-posteriorly along the medial orbital wall. No intracranial extension was observed. Right orbital was normal. Again chemotherapy was started and MRI was repeated after 4 sanctions of chemotherapy, the finding of repeated MRI showed the size of the tumor was decrease but not significantly reduced when compared with the previous finding of MRI. Size of tumor in repeated MRI was 20.0 mm × 10.0 mm. The finding of the current observation showed recurrent rhabdomyosarcoma respond to chemotherapy was not satisfactory or not quick as to non-recurrent rhabdomyosarcoma.

Cases Reported

A case of RMS was reported in 2010, 11 year old healthy male child was presented to a clinic with a chief complaint of a headache, difficulty in moving eye and blurry vision for the last 2-weeks. According to his mother, he had a history of bumped in the head while swimming by his friend. His mother took him to a local ophthalmologist clinic for clinical evaluation. The finding of MRI showed a heterogeneous mass that was originating in the ethmoid to sphenoid sinus. Histopathology finding showed small, round, blue cells which showed feature of embryonal rhabdomyosarcoma and bone marrow biopsy, bone marrow scan and CSF cytology showed no evidence of metastatic disease. PET scan finding showed sphenoid and ethmoid sinus lesion of malignancy with lymph node involvement. So the patient was classified as group III stage 3 T2bNIMo disease and IRS (intergroup rhabdomyosarcoma) [7].

Another case was reported in 2014 in Iran with RMS in a male patient of 15 years age presented with a complaint of swelling of right upper lid since 2-months ago. On external eye examination a firm non tender, the non-mobile subcutaneous mass was palpated. No history of recent trauma and no family history of malignancy were observed. Ecchymosis was developed on lesion about 1-month before visiting the hospital. This lesion showed suspicious of insect bite and allergic reaction. Different medications were used but swelling of an eyelid did not reduce and became normal. Fundoscopy, biomicroscopic, papillary reflex and visual acuity were normal. CT scan of orbital cavity showed soft tissue mass without bony erosion and intra orbital extension. On histopathological examination embryonal rhabdomyosarcoma after confirmation from patient has gone under treatment of chemotherapy for 6 months, after completion of 6 months treatment with chemotherapy patient came to his normal life with no recurrent evidence of tumor [8].

A rare case of rhabdomyosarcoma in old age was present in October 2015 in Mexico, a 96 years old man present with a complaint of pain, swelling, visual disturbance and deposition of an eye. On CT scan a lesion of 38.18 mm × 33.00 mm was noted that showed suspicion of a tumor, and surgical resection of the tumor was done and was sent for histopathological finding. On histopathological finding, rhabdomyosarcoma was diagnosed with the involvement of muscular and adipose tissue as well as with part of sclera [9].

Another rare case of aggressive RMS in adulthood has been reported in Syria in July 2018-Sep 2018; initially patient

went to a local ophthalmologist with a complaint of the watery eye (epiphora) and redness of eye conjunctiva, a reddish mass rising from the left lower eyelid. According to these symptoms patient was diagnosed with perceptual cellulites and was treated with oral antibiotics, but symptoms were not settled. CT scan and excision biopsy was performed. The biopsy showed small round malignant cells that were consistent in orbital embryonal RMS. Two shot of chemotherapy and radiotherapy were given before surgery. Again CT scan of head and neck was repeated which showed soft tissue mass with involvement of left maxillary sinus and the ethmoid cells. CT of abdomen, chest, lungs, and pelvis was performed. A nodule was found in the right lung which indicates it is metastatic nodule. Lung biopsy was suggested to find out for metastatic but the patient refused. Surgical resection was done with left ethmoidectomy and left maxillectomy after surgery patient was followed up with chemotherapy and radiotherapy for 4-months. After 4-months of surgery, the patient became normal and was doing well [10].

Fourteen months old male baby was reported with RMS in India in 2017, the patient was presented with complaints of swelling on the right temporal region during past 20 days and fever, no history of trauma and chronic cough was observed. About 4 cm × 3 cm non tender, firm and immobile mass was palpated. USG of orbit cavity showed mixed echogenic lesion and the orbital wall was involved. But USG abdomen was normal and CT scan with contrast showed a heterogeneous lesion with involving lateral and inferior wall of right orbital, temporal and intra temporal fossa. Embryonal RMS was diagnosed on excision biopsy. The patient was under treatment with chemotherapy including vincristine, cyclophosphamide, and doxorubicin [4].

A case of RMS was reported in the USA in 2018, a 10 years old male was presented with swelling of the right lower eyelid and increase in size for over 2-weeks, no history of pain and trauma was observed. On clinical examination non tender, the immobile subcutaneous nodule was palpated. Visual acuity of left and right eye was 20/20. A solid mass measuring 35 mm × 35 mm × 30 mm has been identified by MRI in the inferotemporal orbit along with the orbital rim and no bone erosion has been noted. Embryonal group II rhabdomyosarcoma has been diagnosed through excision biopsy of the pseudo encapsulated mass. The further patient was managed by complete resection of the tumor and was followed by chemotherapy and radiotherapy [11].

DISCUSSION

The most common childhood orbital malignancy is the rhabdomyosarcoma contributing almost 10% of all RMS cases. The mean age of orbital RMS is 6-8 years and rare case of RMS from birth to the eight decades was also reported. Male was more affected as compared to female [4]. Total 60.4% male were affected with RMS as compared to female which was 39.6%, incidence for children under 5 years was 0.5%. Head and neck RMS were 28.1%, gastrointestinal tract was 17.9%, orbital was 7.9%, retroperitoneal was 3.4% and other sites in the body were 7.9% [12]. Aggressive surgical resection is the best treatment followed by chemotherapy and radiotherapy in RMS because RMS is mostly sensitive to chemotherapy and radiotherapy over last 30 years consequences and there prognosis improvements were significant with 5 years survival rates, 80%-85% achievement in most of the cases in children was achieved but outcome of chemotherapy and radiotherapy in adults was not satisfactory [12-14]. On the basis of the current study result, recurrent rhabdomyosarcoma doesn't have a good response to chemotherapy as compared to the non-recurrence tumor. Surgical resection is the best treatment and was followed by chemotherapy and radiotherapy, these approaches can be helpful to prevent recurrence of RMS.

CONCLUSION

Recurrent orbital rhabdomyosarcoma is rarely reported. To achieve a good survival rate of RMS in children and elderly age there is a need to complete surgical tumor resection anywhere in the body. Chemotherapy and radiotherapy approaches are helpful with complete tumor resection. Only chemotherapy should not be helpful and cannot be preventable from recurrence. Early diagnosis and stared treatment is the best approach and any delay in diagnosis can have a negative effect on health status.

DECLARATIONS

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Ethical Disclosures

The authors declare no human and animals experiment were performed for this study.

Confidentially of Data

The authors declare that no personal data of patient appear in this study.

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Conflict of Interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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