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Case report

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Huge and Hemorrhagic Primary Undifferentiated Small Round Cell Tumor of the Orbit: A Case Report

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Introduction

Small round cell sarcomas are defined by malignant, round, small, relatively undifferentiated cells. When a tumor is poorly differentiated, identification of the diagnostic and morphological features is difficult and therefore, no definitive diagnosis may be possible [1]. Head and neck region are unusual sites for these tumors and obviously orbit is extremely rare [2]. Herein we present an infant with an unusual huge hemorrhagic mass in his face that invade inferior wall of right orbit.

Case presentation

During a routine fetal sonography of a pregnant woman at 37 weeks of gestational age radiologist reported a large echogenic subcutaneous mass in right premaxillary region which didn't exist in previous sonography a week before. After delivery an infant boy with pale appearance and a large hemorrhagic mass at right face hiding the globe, was observed (Figure 1).

A blood test was done and hemoglobin was 13.1 and platelet 233000 at day one. On 3rd day sonography from brain and abdomen was unremarkable but a heterogeneous and hyperechoic mass with a lot of vascularity and cystic spaces has revealed. As the infant was unstable, just a spiral CT scan with contrast could be done that showed a huge exophytic lobulated enhanced mass in right side of face and periorbita measuring 92 × 68 mm with hypervascularity and calcification. Bone destruction at right zygoma and maxilla was noted too. The lesion extended into right orbit and pushed globe superiorly (Figure 2).

Hemoglobin decreased to 8.3 due to necrosis and bleeding from top of mass therefore FFP and packed cell injected and condition stabilized. Because of bleeding property of mass and the size of lesion which caused Kasabach-Merritt phenomenon. The general condition got worse so tumor was resected wholly via inferior transconjunctival orbitotomy approach and re-constriction was performed (Figure 3).

The infant was treated with chemotherapy. Lumbar punctate was done for metastatic evaluation and result was negative. 7 days after first operation, a lateral tarsorrhaphy and debulking of tumor performed. 4 months later the tumor relapsed at the same site and biopsy revealed the same histopathology. Patient died after 2 months.

Histopathological study

Round cell tumors are a group of highly invasive and malignant tumors that can be divided into different types. This group is relatively small and uniform undifferentiated cells with increased nucleus to cytoplasm ratio [3]. Serial sections revealed diffuse sheets of tumor cells with round oval nuclei vesicular chromatin arranged in rosette like formation in some areas. Necrosis, apoptosis and nuclear debris are prominent (Figure 4).

In IHC staining, the mentioned cells showed positivity for CK, EMA, CD99, vimentin, CD57 (few) and negative for LCA, NKX2, chromogranin, synaptophysin, WT1, P40, PLAP and desmin. They are in keeping with undifferentiated malignant small round cell sarcoma.

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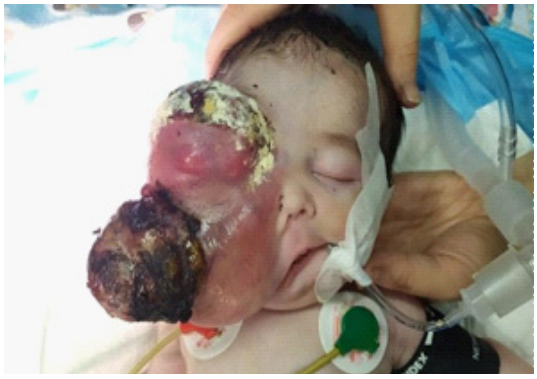

Figure 1: An infant boy with pale appearance and a large hemorrhagic mass at right face hiding the globe.

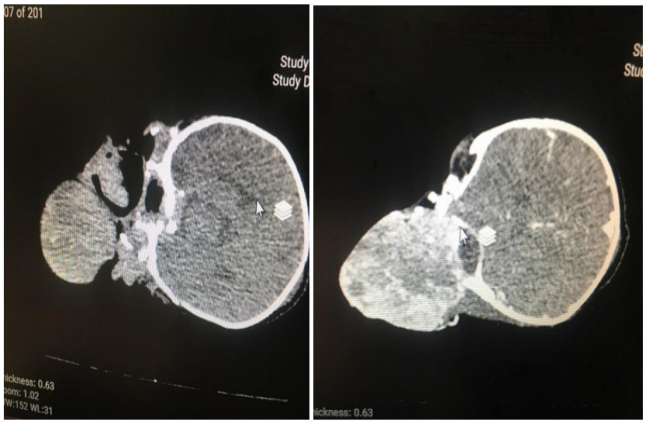

Figure 2: Spiral CT scan with contrast showed a huge exophytic lobulated enhanced mass in right side of face and periorbita measuring 92 × 68 mm with hypervascularity and calcification. Bone destruction at right zygoma and maxilla was noted too. The lesion extended into right orbit and pushed globe superiorly.

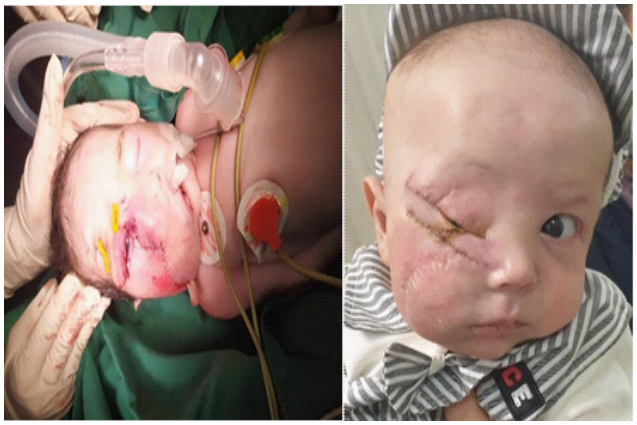

Figure 3: Tumor was resected wholly via inferior transconjunctival orbitotomy approach and re-constriction performed.

Discussion

In Udifferentiated Small Round Cell Sarcomas (USRCS) prognosis is mainly poor [4]. They have an aggressive clinical course with high rates of metastasis and lower sensitivity to chemotherapeutic protocols, with a 5-year survival of only 43% [5]. In 2020 the World Health Organization (WHO) classified soft tissue and bone tumors and USRCS has four subgroups which include pleomorphic, spindle cell, round cell, and epithelioid subtypes [6]. Nowadays most cases of USRCS can be precisely categorize using a combination of morphological, immunohistochemical, and molecular findings, however 10-20% cases cannot be classified into the existing classifications [7] and needs more research [8]. So USRC of orbit is extremely rare. There are seldom samples available to guide treatment. Only 3 case reports have explained similar characteristics to this type of malignancy priorly and all were infants.

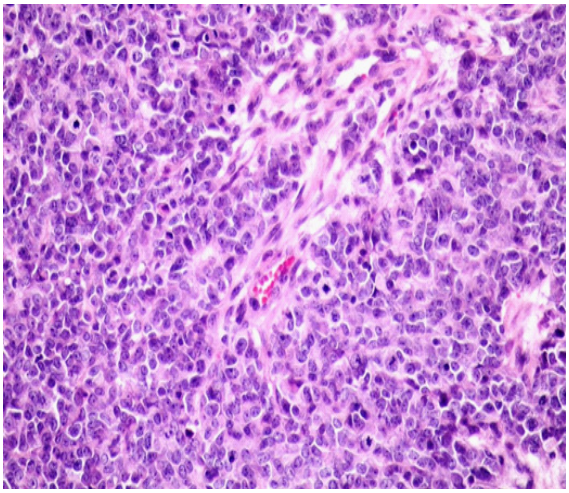

Figure 4: Histopathological study showed, tumor cells with round oval nuclei vesicular chromatin arranged in rosette like formation in some areas. Necrosis, apoptosis and nuclear debris are prominent.

Mendelblatt et al. [9] reported a case of undifferentiated orbital sarcoma present at birth. The initial therapy consisted of exenteration and recurrence was treated by excision and radiation and finally tumor proved fatal by cranial extension.

Gormley et al. [10] reported a congenital undifferentiated orbital sarcoma in a newborn with proptosis and a large right orbital mass. Despite treatment with chemotherapeutic regimen, the tumor continued to grow, and she died at 11 weeks of age. Nowadays more advance immunohistochemical methods have been developed and these methods determine tumor subtypes which not used in these 2 studies.

3rd case is reported by Adesina et al. [11] which is a 6 month old boy presented with recurrent, poorly differentiated orbital sarcoma diagnosed as non-rhabdomyosarcoma soft tissue sarcoma, despite a subtotal resection of his orbital tumor and adjuvant therapy with chemo-radio therapeutic regimen tumor recurrence almost 2 years after initial diagnosis.

In these reports a combination of chemotherapy and radiotherapy is applied. As mentioned before due to the rarity of this type of malignancy no specific treatment guidelines for USRCS are available. Main part of treatment is surgical resection with accurate margin then accompanied with adjuvant radiotherapy and/or chemotherapy [12]. Complete removal of tumor with 1 cm margin is the goal of surgery however in metastatic disease it can be presumed as palliative treatment [13,14]. Biological risk factors assigns the type of adjuvant therapy [15] but post or pre-operative radiotherapy are remarked as standard approaches [15] with considering incongruity and rarity of this tumor, demonstrating the efficacy of adjuvant chemotherapy is difficult so the advantage of this method is unsure and is not routinely recommended.

In our case due to the volume of mass and its bleeding property surgical debulking and primary repair at our center (Bahrami eye center, tehran university of medical science, iran) has done and for continuation of treatment a consult with pediatric oncology service applied and they recommended chemotherapy as post operation adjuvant therapy. Unfortunately, seven times of different types of chemotherapy failed to save the patient. Despite the advice of ophthalmologists to do radiotherapy, the parents and the oncologist refused to let it to be done. But it could help us to save the patient and we do not have enough information about the chances because of the lack of studies on it.

Conclusion

In conclusion, we report a rare case of USRCS of the orbit, an ultra-rare malignancy in this region, we made this diagnosis with help of these characteristics: this tumor has small round cells with hyperchromatic nucleus and hypo-chromatic cytoplasm, without distinct differentiation, also immune histochemical findings were positive for specific markers such as CD99. Aggressive multimodality therapy is needed however prognosis remains poor and adjuvant radiotherapy and/or chemotherapy after wide surgical excision is mandatory to improve prognosis.

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