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Giant Orbital Rhabdomyosarcoma in a Newborn: Case Report

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Abstract: Rhabdomyosarcoma is the single most common type of soft tissue sarcoma in childhood and teenage, it has been reported from birth to the seventh decade, with the majority of cases presenting in early childhood but it is very rare in neonates. The orbit including the eyelids is the usual primary site in the head and neck, and it is a highly malignant tumor. There have been only a few cases of congenital orbital rhabdomyosarcoma previously reported in the literature. We report a case of a newborn girl that was admitted in our structure 2 days after her birth with giant right orbital mass inducing proptosis that was discovered at birth. MRI showed a poorly defined mass but biopsy of the tumor confirmed the diagnosis of Rhabdomyosarcoma. Chemotherapy was started accordingly with VAC regimen with a good evolution under treatment infortunatly the patient died at four month old from an infectious disease because she doen't receive any vaccination. Congenital RMS has a poor prognosis and must benefit from a multidisciplinary approach. Children with cancer need to be immunized against the common vaccine-preventable diseases sometimes during ongoing chemotherapy to increase their chance of survival. Prenatal diagnosis may also improve the prognosis of these patients.

Keywords: Rhabdomyosarcoma, Neonate, Vaccination

1. Introduction

Orbital rhabdomyosarcoma (RMS) arises from pluripotent mesenchymal cells and is the most common orbital malignancy in children. It accounts for approximately 4% of orbital mass in this age group. The mean age of diagnosis is 6-8 years; however, it is very rare at birth [1, 2].

Histologically, most orbital RMS in children are generally embryonal or alveolar [2].

Timely diagnosis and treatment can improve the prognosis, but the main problem is when we can vaccinate these patients that are treated by chemotherapy.

2. Case Report

The patient, a term new born girl, was born of a woman of 27 years old, primiparous.

Family history for drugs, malignancy, and hereditary disease was negative. Apgar score at birth was 9. Physical examination was normal except for right orbit necrotic huge mass with proptosis and downward displacement of the right eye. MRI has showed a poorly defined mass (hemangioma). Histological examination of incision biopsy showed a Rhabdomyosarcoma with fusiform cells.

Tumour node metastasis pre-treatment staging was T2N0M0.

The patient was discharged on 21th day of life, receiving systemic chemotherapy (VAC) with a good evolution (regression of the tumour), but she died at 4 months old from an infectious disease (pertussis) because she did not receive any vaccination.

3. Discussion

Annual incidence of RMS is: 4.3 cases per million persons age < 20 years.

It is the most common soft tissue sarcoma in children and

can occur at any age: with two peaks of incidence: 2_6 years and 14_18 years [3] May be present at birth but very rare: 5 to 10% of all cases occur in patients < 1 years [4].

Diagnosis may be difficult in new born. RMS should be considered in the differential diagnosis of any child with a unilateral proptosis. It consists of a detailed history, ocular examination, imaging studies including CT or MRI, and biopsy.

Differential diagnosis includes: orbital teratoma, haemangioma, meningocel, lymphangioma [5] Sometimes the diagnosis may be delayed because of that differential diagnosis which may worsen the prognosis.

That why in our case we have done biopsy immediately to clarify the diagnosis.

Some syndromes can be associated with RMS such Gorlin Syndrome, Noonan Syndrome, LI-Fraumeni Syndrome. [6] Our patient did not have any signs suggesting such syndromes.

The management of new born baby with RMS is challenging. It requires special therapy and careful monitoring to prevent life threatening complications.

The recommended chemotherapy protocol for treatment of RMS is VAC (Vincristine, Actinomycin D, and Cyclophosphamide). Meanwhile the immaturity of new-born organs exposes to a high risk of complications. That why protocols should be adapted according to the modified metabolism.

Ragab et al showed that the toxicity rate is very high in infant on comparison with older child: 5% of toxicity related death in infant versus 1% in older child [7]. The authors recommended reducing the dose of chemotherapy by 50% in infants. This finding have been confirmed by other studies: [8-11] Reduced dose of chemotherapy was well tolerated in many studies [8, 12]. No major renal, liver or neurologic toxicity was observed in new born. Meanwhile moderate myelosuppression have been reported as in our case. Doxorubicin and anthracycline are known to potentially cause cardiologic toxicity in new-born, and it's recommended to ovoid such molecules in infant under 3 months [13].

Another issue is how to calculate the dose. It's recommended to use weight instead of surface area in calculating the dose of chemotherapy.

Careful monitoring, reduced dose calculated on the basis of weight and not surface area, avoiding anthracycling are the mean important points to limit the side effect of chemotherapy in new-born with RMS.

Radiotherapy for local control is very important in the management of RMS. Meanwhile radiotherapy is not recommended at this age because of the high risk of complication especially in orbital localisation.

Prognosis of RMS is associated with the following factors: RMS location, histology, stage and age.

RMS location: parameningeal, orbit, head and neck, extremity, genitourinary, paratestis, retroperitoneal, studies showed the infant < 1 year present more often GU as primary site.

Histology: 5 histologic types of RMS known: embryonal, botryoid, spindell cell, alveolar and undifferentiated. Patient with undifferentiated and alveolar type are associated with poor prognosis. Studies showed the infant < 1 year are more often associated with undifferenttaited tumour than other types [14].

Stage [14].

Age: Many studies confirmed that age< 1 year is associated with poor prognosis. Age is an independent negative prognosis factor for RMS: failure free survival was 55% for infant < 1 year versus 83% for children 1-9 years. [15]. one of the reasons of such poor prognosis could be the histology of the tumour, the limited therapeutic resources.

There is little evidence-based literature to support immunization guidelines in immunocompromised children, some authors suggest that the vaccination should be given at least three months after the completion of chemotherapy, The cause of death in our case may cause physicians to reconsider the indication of appropriate moment for vaccination.

Figure 1. *Clinical photograph of the patient with the right orbit lesion.*

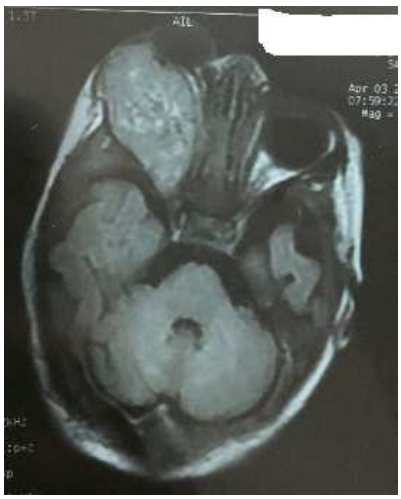

Figure 2. *MRI scan showing right orbit lesion.*

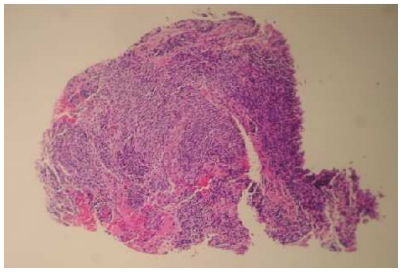

Figure 3. *Histopathologic findings small round cells.*

4. Conclusion

This case merits discussion for many reasons. it is reported because of extreme rarity of RMS to occur in neonates. Moreover no CNS metastasis was recorded in our case. And because it also highlights the poor prognosis associated with non vaccination of patients with RMS especially in neonates.

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