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(CASE REPORT)

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Neonatal Rhabdomyosarcoma: About a new observation

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Abstract

Rhabdomyosarcoma is a cancer that develops from so-called "soft" tissue. In neonates, as well as in older children, rhabdomyosarcoma is the most common malignant soft tissue tumor. Malignant mesenchymal tumors are, after neuroblastoma, the second most common type of cancer in the newborn.

Our case is a female neonate, 2nd of 2 siblings, full term, eutrophic, admitted at H4 of life with respiratory distress rated 2/10 according to the silverman score with left choanal atresia. The oral examination revealed several solid pinkish masses and the cervico-facial CT scan showed an oval retro pharyngeal collection associated with infiltration of the superficial and deep left neck and upper mediastinum spaces.

The histopathological study found a morphological and immunohistochemical aspect of an embryonal rhabdomyosarcoma.

Rhabdomyosarcoma is rare in the neonatal period, it is located mainly in the limbs, genitourinary tract and head and neck region. As was the case with our newborn who presented with an oropharyngeal location.

Their diagnosis can be difficult and requires the advice of an expert anatomopathologist. Their management must be multidisciplinary.

Keywords: Rhabdomyosarcoma; In-newborn; Tumors; Embryonic; Neonatal

1. Introduction

Malignant soft tissue tumors are, after neuroblastoma, the 2nd leading cause of cancer in the neonatal period [1].

Rhabdomyosarcoma is a cancer that develops from the so-called "soft" tissues (mainly muscles and supporting tissues that are between organs). This tumor can occur in different parts of the body such as the head and neck, urinary tract, reproductive system, gastrointestinal tract and others.

Rhabdomyosarcoma is one of the so-called embryonic tumors because the cancer cells resemble muscle cells in the embryo.

2. Clinical case

Female neonate, 2nd of 2 siblings, at term, eutrophic with a birth weight of 3450 g and a birth height of 49 cm, and head circumference of 34 cm. He was born in a non-consanguineous pregnancy, well monitored, to a 24-year-old mother,

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with no particular pathological history. The delivery was by vaginal route; Apgar not specified; Negative infectious history and notion of stained liquid; Admitted at H4 of life.

The clinical examination showed respiratory distress rated 2/10 according to the silverman score. A left choanal atresia, a good hemodynamic and neurological status and well differentiated female genitalia.

In front of this state, we evoked in first an infectious cause considering that the newborn was symptomatic, a neurological cause is also evoked considering the absence of evaluation of the fetal well being and the non evaluation of the adaptation to the extra uterine life (Apgar not specified) and the notion of the tinted amniotic liquid and in last place a transient respiratory distress in front of a fast vaginal delivery.

Chest X-ray showed cardiomegaly with TIA at 0.56. Echocardiography showed pulmonary hypertension of respiratory origin.

Blood gas on admission was PH= 7.35; PCO2=43; HCO3-=23.2

The biological workup showed anemia at 8.3 g/dl on the blood count with normal WBC and platelet count, normal natraemia at 134 mEq/l, calcemia at 69 mg/l, negative CRP at 0.81 mg/l. The maternal workup showed CRP at 78mg/l and on ECBU leucocyturia at 450/field with a sterile culture. The biological stigmas of suffering came back to 650 for creatine kinase and 910 for lactate dehydrogenase.

The evolution was marked by worsening respiratory distress and respiratory acidosis on control blood gasometry, hence the need for artificial ventilation.

Intubation was difficult due to the presence of several pinkish solid masses without necrotic areas (Figure 1).

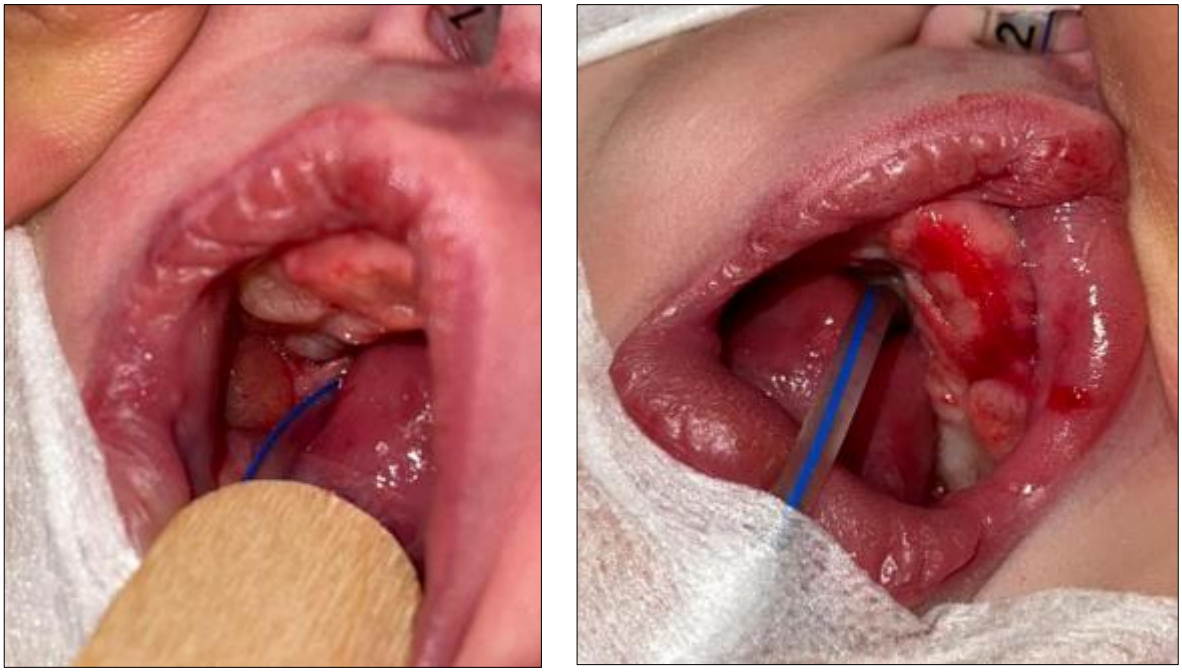

Figure 1 Appearance of the oral masses

A faciocervical CT scan shows an oval retro pharyngeal collection with infiltration of the left superficial and deep spaces of the neck and the upper mediastinum.

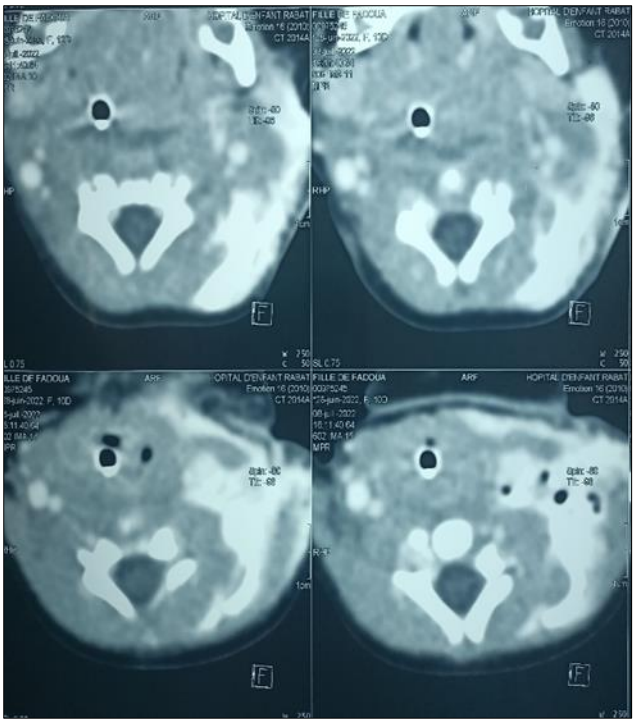

Figure 2 Retropharyngeal collection on transverse cervical sections

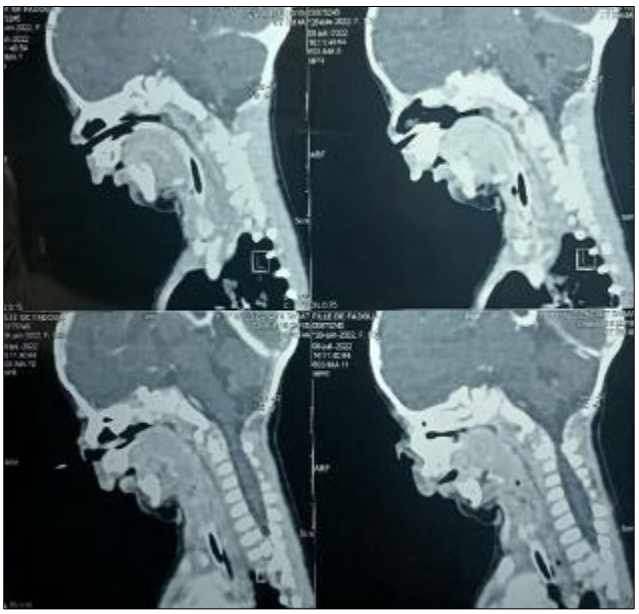

Figure 3 Retro-pharyngeal collection on sagittal sections

Laryngoscopy under general anaesthesia evoked a tumoral aspect of the oropharynx with multiple biopsies.

The histopathological study found a morphological and immunohistochemical aspect of an embryonal rhabdomyosarcoma.

The case was discussed with our oncologist colleagues who suggested to start with a mild chemotherapy in order to reduce the tumor compression and to try extubation until then impossible.

The evolution was marked by death in septic shock before the start of chemotherapy.

3. Discussion

Neonatal malignant tumors represent only 2% of childhood cancers. Among them, malignant mesenchymal tumors are, after neuroblastomas, the second most common type of cancer observed in newborns, representing, depending on the series, 10 to 20% of cases [2]. These malignant neonatal mesenchymal tumors do not seem to be associated with congenital malformations or specific chromosomal abnormalities. Similarly, no environmental risk factors (especially during pregnancy) have been identified. In newborns, as in older children, rhabdomyosarcoma is the most common soft tissue malignancy [3].

Rhabdomyosarcoma is rare in the neonatal period, accounting for 0.4 to 2% of all rhabdomyosarcomas depending on the series [4].

The clinical features of neonatal RMS are comparable to those of RMS in older children. The tumor presents as a more or less large, often painful, and rapidly growing mass. It is mainly located in the limbs, the genitourinary tract (risk of acute urine retention) and the head and neck region. As was the case with our newborn, who presented with an oropharyngeal location. At the time of diagnosis, the majority of tumors are localized, however 6-14% are already metastatic (mainly in the lungs).

A case of congenital embryonal rhabdomyosarcoma of the right shoulder is described. The patient was the first child born to a 24-year-old woman who had previously been treated for infertility. The diagnosis was made by ultrasound in the 36th week of gestation. This full-term boy was delivered by cesarean section because of the size of the tumor. Surgical treatment was supplemented by chemotherapy [5].

Histologically, RMS presents as a round cell tumor, the rhabdomyoblasts. There are 2 forms: the embryonic form and the alveolar form.

In our patient, the rhabdomyosarcoma is of embryonic type.

The management of this disease in newborns and, more generally, in small children is difficult because of the poor tolerance of chemotherapy due, among other things, to the immaturity of the tissues and enzyme systems at this age and the long-term risks of "unacceptable" sequelae, which contraindicate the use of external radiotherapy [1].

Chemotherapy is adapted to the age and weight of the child. Local treatment is essentially represented by conservative surgery, sometimes associated with brachytherapy (vesicoprostatic RMS). Surgery can sometimes be mutilating (amputation of 1 limb, total cystoprostatectomy), but it should be preferred to external radiotherapy [1].

The prognosis of localized RMS in children under 1 year of age is comparable to that observed in older children, with an overall 5-year survival of 72% [6]. The only study specifically evaluating the survival of neonates with RMS is an American study of 14 children, half of whom survived long term [5]

4. Conclusion

Neonatal soft tissue masses have a unique and mostly favorable course and prognosis. Their diagnosis can be difficult and requires the advice of an expert anatomopathologist. Their management must be multidisciplinary, in a specialized environment, and based on adapted chemotherapy associated with conservative surgery.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from guardian included in the study."

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