Scientific Research Publishing

Journal of Biosciences and Medicines, 2023, 11, 103-107
https://www.scirp.org/journal/jbm
ISSN Online: 2327-509X
ISSN Print: 2327-5081

Mandibular Embryonal Rhabdomyosarcoma Confused with a Vascular Malformation: About a Case

Massilia Bouhmidi, Hajar Boudarbala, Ayad Ghannam, Aziza Elouali, Abdeladim Babakhouya, Maria Rkain, Noufissa Benajiba

Mother-Child Hospital, CHU Mohamed VI, Oujda, Morroco
Email: hajar.bdr2016@gmail.com

How to cite this paper: Bouhmidi, M., Boudarbala, H., Ghannam, A., Elouali, A., Babakhouya, A., Rkain, M. and Benajiba, N. (2023) Mandibular Embryonal Rhabdomyosarcoma Confused with a Vascular Malformation: About a Case. *Journal of Biosciences and Medicines*, **11**, 103-107.
https://doi.org/10.4236/jbm.2023.116007

Received: March 31, 2023
Accepted: June 13, 2023
Published: June 16, 2023

Abstract

Malignant mesenchymal tumors (MTM) in children represent 5% to 10% of malignant tumors in children. They constitute a heterogeneous group of tumors of various differentiations depending on their supposed tissue of origin. They mainly include tumors of muscular origin, those derived from connective, vascular, nervous, or adipose tissue. Rhabdomyosarcoma (RMS) is the most common mesenchymal tumor in children and adolescents (60% to 70% of them). And it accounts for 5.8% of all malignant solid tumors in children. Almost half of rhabdomyosarcomas occur in the head and neck. The prognosis for this type of tumor is particularly poor. A case of rhabdomyosarcoma in the mandible with extension to the abdominal wall and unilateral testis in a 6-month-old infant is reported with evolution since birth. It is a purplish lesion at the level under the right chin which was initially taken for vascular malformation, evolving very quickly towards a mandibular mass deforming the painful face with inflammatory signs, followed by the appearance of a hard swelling under the skin on the left flank taking on the same aspect of the mandibular mass. This observation illustrates the need to know how to systematically think about tumor causes in the face of atypical aspects and to carry out an anatomopathological examination.

Keywords

Rhabdomyosarcoma, Mandible, Mesenchymal Tissue, Vascular Malformation, Abdominal Wall, Testis

1. Introduction

Rhabdomyosarcoma (RMS) is a malignant tumor with striated muscle differen-

tiation preferentially affecting children and adolescents. The main localizations are cephalic, genito-urinary and skeletal, especially the extremity of the limbs [1]. Histologically, three forms of rhabdomyosarcoma exist: the embryonic form (most common, 80% of cases) with intermediate prognosis, the Alveolar form and the pleomorphic form with poor prognosis [2]. A case of embryonic RMS with mandibular location considered to be a benign vascular malformation, with evolution since birth, is reported.

2. Case Report

We report the case of an only son of his parents aged 6 months without parental consanguinity, born vaginally at term without notable pathological neonatal history, with a history dating back to the birth of a small purplish swelling under the right chin without inflammatory signs initially considered as vascular malformation of the angioma type. The evolution was unpredictable with a very rapid increase in the volume of the tumefaction deforming the face and the appearance of inflammatory signs with pain causing incessant cries in the infant, followed by an appearance of a parietal tumefaction on the level of the left flank. The clinical examination objectified a voluminous mass of the right mandibular region measuring 7.5 cm*11 cm of long axis deforming the face, red, hot, and very painful on palpation, with an irregular surface, extended to the cervical region below, laterally at the nasolabial fold and the right cheek, and above at the right labial commissure and the right lower lip (**Figure 1**). On examination of the oral cavity, the palate is preserved, with infiltration of the floor of the mouth and the internal face of the left cheek. And on abdominal examination, presence of a palpable subcutaneous mass on the left flank, hard, purplish, with opposite inflammatory signs, similar in appearance to the mandibular mass, estimated at 1.5 cm (**Figure 2**). Associated with three fixed laterocervical lymph nodes about 3 cm. Radiologically, the cervical MRI objectified a locally advanced tumoral process suggesting a sarcomatous origin (rhabdomyosarcoma or angiosarcoma) (**Figure 3**), with the anatomopathological study returning in favor of a rhabdomyosarcoma. Imaging of extension objectified a tissue mass of the soft tissues of the left flank with ADP of the right iliac fossa, with right unilateral testicular infiltration. Given these clinical-radio-anatomopathological criteria, the RMS was classified as high risk in the face of lymph node involvement, size of more than 5 cm. The localization was non-para meningeal. The patient was treated with chemortherapy according to the MMT 2005 protocol associating vincristine, ifosfamide, and actinomycin (IVA) was initiated, and the first three chemotherapy sessions were delivered initially in 8 weeks. This chemotherapy has reduced the tumor volume, and the volumetric response after these first cures was greater than 33%. The decision was to continue the chemotherapy with 6 sessions of IVA associated with surgery which was carried out at week 13 after the fourth cure of chemotherapy with good evolution (**Figure 4**).

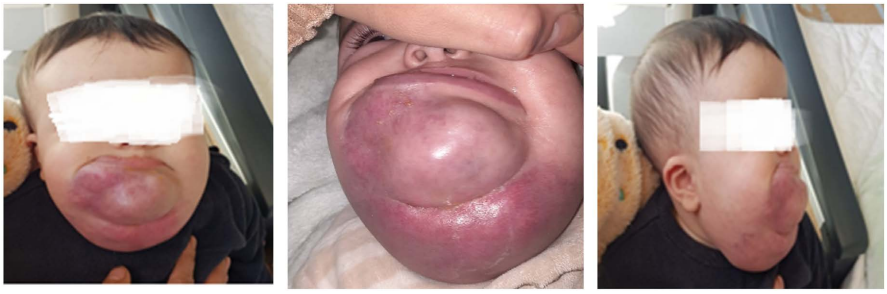

Figure 1. Purplish mandibular mass distorting the face with adjacent inflammatory signs.

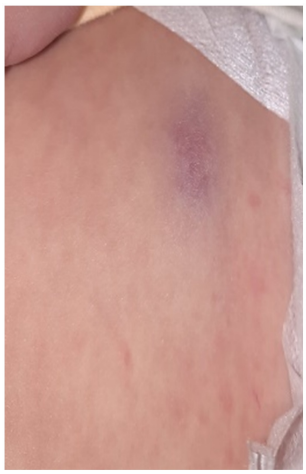

Figure 2. Parietal swelling of the left flank taking on the same appearance as the mandibular mass.

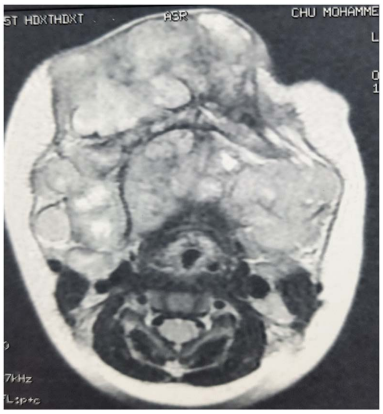

Figure 3. Cervico-occipital MRI for the current case.

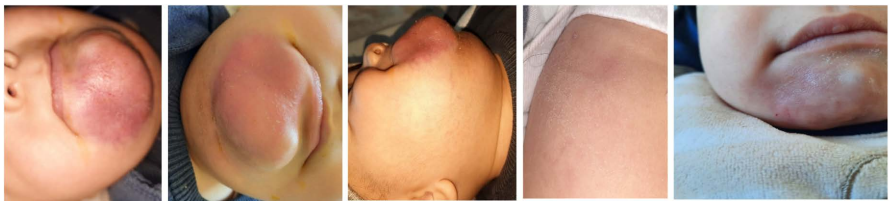

Figure 4. Evolution of swelling during chemotherapy sessions: Clear regression of the tumor process after chemotherapy.

3. Discussion

The RMS represents 7.5% of mesenchymal tumors of the oral cavity [1]. The distribution does not seem to be affected by sex [3]. The white child is three times more affected than the black child 6. The majority cases occur during the first two decades of life. The soft palate appears to be the preferred site for oral forms of RMS [3]. Histologically, four forms of RMS are distinguished, the embryonic type, the botryoid type, the alveolar type and the pleomorphic type; botryoidal RMS is considered a variant of the embryonic type [3]. There is a correlation between histological type and prognosis [2]. The embryonic type is characterized by very diverse aspects of muscle differentiation. The lesion strongly resembles fetal muscle tissue. Tumor cells are small, round or oval, poorly differentiated, contain little cytoplasm and a hyperchromatic nucleus of variable size and shape. In more differentiated forms, the cytoplasm of eosinophilic rhabdomyoblasts is more abundant and contains fibrillar material arranged concentrically around the nucleus. Striations are sometimes found on the periphery. This histological type is associated with a better prognosis, more particularly in its botryoidal variant. The 5-year survival rate for this last form of RMS, which mainly affects the genitourinary tract, is 95% [2]. Immunohistochemistry is of great contribution in the differential diagnosis which must consider peripheral neuroectodermal tumors (PNET) and malignant lymphomas. It uses monoclonal antibodies that are markers of striated muscle tissue (Ab antidesmin) or more specific for rhabdomyosarcoma (Ab antimyogenin). The karyotype of the tumor tissue and the study in molecular biology complete the anatomo-pathological diagnosis by eliminating the specific chromosomal translocations of the alveolar RMS and the PNETs. The etiopathogenesis of RMS remains poorly understood. No exogenous factor has been identified. The agents responsible for this mesenchymal proliferation seem to be linked to genetic factors, particularly in form [4]. The management of RMS is multidisciplinary, including polychemotherapy, surgery and external radiotherapy. The 5-year survival rate is 55% for children compared to 22% for adults.

Moreover, a very interesting message to remember from this observation is that we must always pay attention to the differential diagnoses of cutaneous angiomas in newborns and infants, the main one being the malignant tumor of the soft tissues of infants (rhabdomyosarcoma, infantile fibrosarcoma) and which must be evoked in a congenital context; a single mass; localized to the face or concerning a limb segment; indurated; fixed; deforming the superficial reliefs. These tumours, which sometimes have a purplish tint, can simulate a vascular tumour, which delays diagnostic and therapeutic management. This risk justifies a biopsy in the slightest doubt.

4. Conclusion

Rhabdomyosarcoma is the most common malignant mesenchymal tumor, occurring in children and very young adults. Localized forms have a good prognosis while metastatic tumors have very poor results. A well-defined treatment based

on surgery and chemotherapy gives good results.

Conflicts of Interest

The authors declare no conflict of interest.

References

[1] Peters, E., Cohen, M., Altini, M. and Murray, J. (1989) Rhabdomyosarcoma of the Oral and Paraoral Region. *Cancer*, **63**, 963-966. https://doi.org/10.1002/1097-0142(19890301)63:5%3C963::AID-CNCR2820630529%3E3.0.CO;2-0

[2] Nemon, W.A., Gehan, E.A., Wesser, B.L., Marsden, H.B., Unnik, A.J.M., Hammoudi, A.B., *et al.* (1995) Classification of Rhabdomyosarcoma and Related Sarcomas. *Cancer*, **76**, 1073-1084. https://doi.org/10.1002/1097-0142(19950915)76:6%3C1073::AID-CNCR2820760624%3E3.0.CO;2-L

[3] Chen, S., Thakur, A., Miller, A.S. and Harwick, R.D. (1995) Rhabdomyosarcoma of the Oral Cavity—Report of Four Cases. *Oral Surgery, Oral Medicine, Oral Pathology Oral Radiology*, **80**, 192-201. https://doi.org/10.1016/S1079-2104(05)80202-1

[4] Barr, F.G., Biegel, J.A., Sellinger, B., Womer, R.B. and Emanuel, B.S. (1991) Molecular and Cytogenetic Analysis of Chromosomal Arms 2q and 13q in Alveolar Rhabdomyosarcoma. *Gene Chromosom Cancer*, **3**, 153-161. https://doi.org/10.1002/gcc.2870030212