ejpmr, 2019,6(4), 98-101

SJIF Impact Factor 4.897

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

EWING'S SARCOMA OF THE MAXILLARY SINUS IN A CHILD: A CASE REPORT AND REVIEW OF LITTERATURE

Khaoula Hafidi*, Ilhame Toumir, Hasnae Taghzout, Zineb Alami, Touria Bouhafa and Khalid Hassouni

[1]Department of Radiotherapy, University Hospital Center Hassan II Fez, Morocco.

***Corresponding Author: Dr. Khaoula Hafidi**

Department of Radiotherapy, University Hospital Center Hassan II Fez, Morocco.

Article Received on 31/01/2019 **Article Revised on 21/02/2019** **Article Accepted on 14/03/2019**

ABSTRACT

Ewing's sarcoma is typically an aggressive, poorly differentiated tumor affecting children and young adults, it accounts for 4–6% of all primary bone tumors and facial primary localizations occur in only 1–4% of all cases, mostly in the mandible and calvaria. Paranasal sinus involvement is rare. Only 15 cases of extra-osseous Ewing sarcoma located in the nasal cavity and in the para-nasal sinus have been reported in the literature. The diagnosis of Ewing sarcoma is based on a complete imaging as well as on the anatomopathological analysis of the tumor tissues. It is characterized by dense clusters of small round cells in sheets, without tissue differentiation. It is a part of the family of neuroectodermal tumors, which have 11-22 translocation (or equivalent). Management must be multidisciplinary. The current treatment is based on the combination of chemotherapy and local treatment, at best surgical, sometimes radiotherapy. The two main prognostic factors are the existence of initial metastases and the response to chemotherapy. In the absence of treatment, the fatal course is usually rapid, with the appearance of pulmonary or bone metastasis. Advances in molecular biology can lead to hope for new treatments in the future. We report a new case of primitive Ewing sarcoma of the maxillary sinus in a 12-year-old child.

KEYWORDS: Ewing's sarcoma; Maxillary sinus; Chemotherapy; Radiotherapy.

INTRODUCTION

Ewing's sarcoma (E.S) is a rare mesenchymal tumor that typically develops in the bone.[1] Some extra bones cases have been described, particularly in the para-vertebral and retro-peritoneal regions. Less than 15 cases of extra bone ES located in the nasal cavity and in the paranasal sinuses have been reported in the literature.[2] Through this new case treated in our hospital university center, we will discuss the epidemiological, clinical, histological and therapeutic aspects of this rare tumor localization.

CASE REPORT

A 12-year-old patient with no notable pathological history, he complained of swelling of the rightcheek first noted 2 months ago. This swelling has been progressively increasing an size, which evolves in a context of slimming. The clinical examination found a child in good general condition, presenting a painful swelling, the palpation of the right zygomatic arch was painful without other associated signs. The rest of the somatic examination was normal, including the ophthalmological examination. Computed tomography and magnetic resonance imaging of the facial mass revealed the presence of a tissue process centered on the right maxillary sinus responsible for lysis of its various walls, measuring 61 * 51 * 52mm, with invasion of the soft parts of the eyes. (Figure 1). the diagnosis of ES has been evoked. An incisional biopsy was taken and the histological findings confirming an ES. The extension assessment revealed no argument in favor of secondary localization. The case was discussed with the onco pediatrician, and the decision was made to treat the patient by chemotherapy and complete by radiotherapy. So the patient received neoadjuvant chemotherapy protocol EMP (Vincristine-Ifosfamide-Doxorubicin-Etoposide), with good tolerance. Cranio-facial magnetic resonance imaging after 2nd, 4th and 6th cures showed a partial tumor response (Figure 2), then the patient was put under VAI (Vincristine-Actinomycin-Ifosfamide) consolidation chemotherapy in combination with conformational radiotherapy. by intensity modulation at a dose of 60 Gy, 2 Gy / fraction, 5/7 days, 50 days of spreading (Figure 3). The evolution was satisfactory without clinical and radiological recurrence with a follow-up of one year.

DISCUSSION

The Ewing's sarcoma family of tumors (EFT) includes ES of bone (ESB), extra osseous ES (EES), peripheral primitive neu- roectodermal tumor of bone (pPNET) and malignant small- cell tumor of the thoracopulmonary region (Askin's tumor), all of which are neoplasms of neuroectodermal origin.1 Ewing's Sarcoma of the facial bones is a rare pathological entity and involvement of the maxilla is even rarer. A rapidly enlarging, often painful mass is the most frequent clinical presentation.[3]

Signs and symptoms may include paresthesia, loss of teeth, and ulceration of the overlying mucosa. Tumor of the maxil- lary sinus might not be detected until the lesion protrudes into the nasal cavity and oral cavity, which can cause nasal obstruc- tion, epistaxis, and destruction of the palate.[4]

In the case described in tour study, the patient preented with a painful sweeling, with no oral or nasal protrusion. Imaging is essential, not only is it important for diagnosis and assessment, but it also allows evaluation of treatment efficacy and detection of recurrence. On computed tomography, a periosteal pluri-lamellar apposition known as onion bulb or spiculated in sun rays (lamellae perpendicular to the diaphysis) is characteristic of Ewing's sarcoma of long bones. However, Ewing's sarcoma of the maxillary sinus appears, most commonly as a permeative lesion (multiple small holes) with extensive soft-tissue invasion without calcifications indicative of its aggressive nature. Magnetic resonance imaging is systematic in the initial assessment, it typically shows a hypo intense to iso-intense lesion in T1 and hypo intense to hyper intense in T2.

The diagnosis of Ewing's sarcoma of the maxillary sinus is histological, like Ewing's sarcoma of other locations, preferably on a surgical bone biopsy performed in a reference center with laboratories capable of performing the necessary cytogenetics and biology examinations molecular on the tumor. It is preferable that the surgical bone biopsy be performed by an experienced surgeon who will take care of the patient afterwards because the tumor removal surgery includes the biopsy path and must remove the scar. In the current study the incisional biopsy was taken.[5]

Sarcoma tumor cells from Ewing, originating from the mesenchymal tissue are small, round, undifferentiated and positive to the CD99 membrane markers. However, this histological description is not specific to the Ewing tumor. The particularity of ES lies in its molecular signature and in the evidencing of a cytogenetic anomaly: the reciprocal chromosomal translocation t (11; 22) (q24; q12). This induces the fusion of specific genes carried by chromosomes 11 and 22, thus resulting in the formation of a mutated gene that codes for an abnormal protein called EWS / FLI-1 whose function is not perfectly known. At the present time, a translocation involving the EWS gene is considered pathognomonic for Ewing's sarcoma and it is this finding that makes it possible to establish the definitive diagnosis of Ewing's sarcoma.[6]

Ewing's sarcoma metastasises in approximately 18% of cases, most often at the pulmonary level (57%), bone (34%), and rarely at the cerebral and ganglionic level.[7] In the current study the investigations did not show evi- dence of metastatic disease.

Based on the results of numerous studies, several prognostic factors for ES have been defined. These make it possible to allocate patients with Ewing's sarcoma in different groups according to the prognosis and thus to establish a therapeutic protocol of intensity adapted to each.[8,9] The main prognostic factors are the presence of metastasis at the time of diagnosis and the response to neoadjuvant chemotherapy. Other factors were identified as: Age of the patient greater than 15 years, tumor size greater than 200ml and a high level of *a*ctate dehydrogenase LDH.

The management of Ewing's sarcoma of the maxillary sinus requires a multidisciplinary approach. Treatment includes chemotherapy and excision surgery. Radiotherapy will be discussed according to the post-operative pathological elements.

Although there are no prospective randomized studies comparing surgery and radiotherapy, retrospective studies suggest better local control of maxillary sinus Ewing sarcoma after surgery (with or without adjuvant radiotherapy) compared to radiotherapy alone. Tests CESS 81, CESS 86 and EICESS 92 even showed that the local control rate after surgery was significantly higher than after exclusive radiotherapy.[10] However, Grevener et al. Recently published the results of a data analysis of the German Society of Pediatric Hematology and Oncology, this analysis concerned Ewing's sarcomas of the head and neck, 7 cases of Ewing's sarcoma of the maxillary sinus were included. This analysis showed that there is no difference, or survival without recurrence or overall survival, between the different local treatment modalities: surgery, radiotherapy or surgery plus adjuvant radiotherapy

Data from the CESS and EICESS trials showed that patients with intra-lesional resection followed by radiotherapy had the same local control rates as patients who received radiotherapy alone.[10], thus clearly reversing the benefit of surgery.

Complete resection should be considered as the mainstay of the therapeutic management of Ewing's sarcoma. In cases where complete surgical excision is not feasible, adjuvant radiotherapy is indicated at a dose of 45 to 60 Gy, preferably in combination with systemic therapy.

In our case the patient did not have a the decision was to start chemotherapy followed by radiation therapy for local control.

The morbidity of local therapy is one of those key issues, which, in the face of Ewing sarcomas in the maxillary and maxillary sinuses, will have a huge impact on the choice of local treatment modality. Radical tumor excision is often limited by the proximity of adjacent critical structures, with the possibility of enormous complications: respiratory function, nutrition, phonation, vision, olfaction and aesthetic problems. The irradiation

of the head and neck may seem less deleterious but it can give serious side effects: mucositis, hypersialia, oesophagitis, regional pain, radiodermites, eye disorders, cognitive disorders... These side effects are certainly less and less frequent thanks to the new modern techniques of radiation intensity modulation to irradiate the tumor and spare as much risk organs as possible.

Chemotherapy can be given initially as a neoadjuvant, 3-6 cycles at 2-3 week intervals, to reduce tumor size and increase the likelihood of achieving complete surgical resection with negative margins. As an adjunct, 6 to 10 additional cycles of poly chemotherapy showed improved recurrence-free survival and overall survival.[11] The combinations of chemotherapy vary according to the teams, but the most traditional are: VACA (Vincristine, Doxorubicine, cyclophosphamide, actinomycin), VAIA and EVAIA (Ifosfamide replaces cyclophosphamide with or without the addition of Etoposide) and finally VIDE (Vincristine, Ifosfamide, Doxorubicin, Etoposide). The Euro-EWING 99 protocol is an international "intergroup" protocol, involving at least 5 European cooperative groups that have decided to build a common protocol for an infrequent illness, the only way to answer scientific questions in a scientific way. The French Society of Pediatric Oncology and the EORTC are two of these groups.[12,13]

The multidisciplinary approach of treating Ewing's sarcoma with chemotherapy, surgery and radiotherapy has increased long-term survival to more than 50%. In 1987, the intergroup Ewing Sarcoma Study (IESS) concluded that Ewing's sarcoma of the head and neck area had a better survival rate than other regions.

Recurrence of Ewing's sarcoma of the bone in general is most common within 2 years of initial diagnosis (approxi- mately 80%). The overall prognosis for patients with recurrent Ewing's sarcoma is poor[14,15]

In the case described in the current study we had a good local control with a follow up of one year without recurrence or metastasis.

CONCLUSION

Ewing's sarcoma of the maxillary sinus remains a relatively rare disease. His diagnosis is currently based on histology and molecular biology data. Its prognosis has been significantly improved thanks to modern therapies. His treatment is based on the combination of chemotherapy surrounding complete surgical resection of the tumor. Radiotherapy must remain reserved for some particular indications.

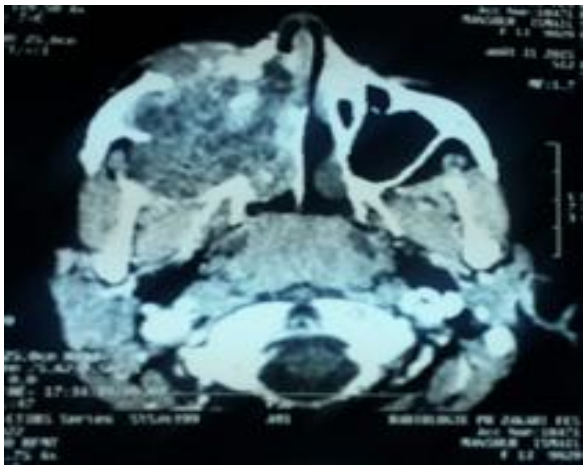

Fig 1: CT scan showing Ewing's sarcoma of the maxillary sinus

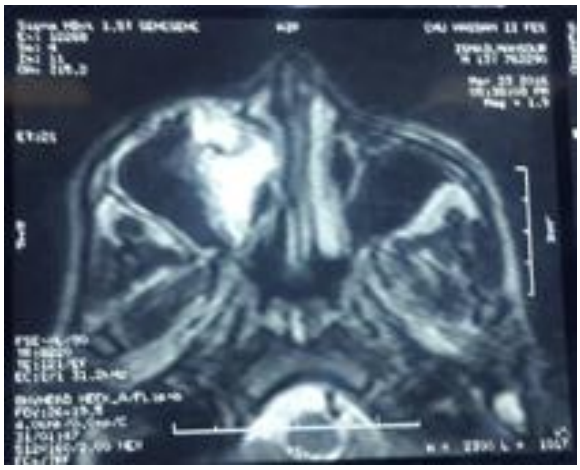

Fig 2: Sagittal magnetic resonance imaging (MRI) through the maxillary sinus

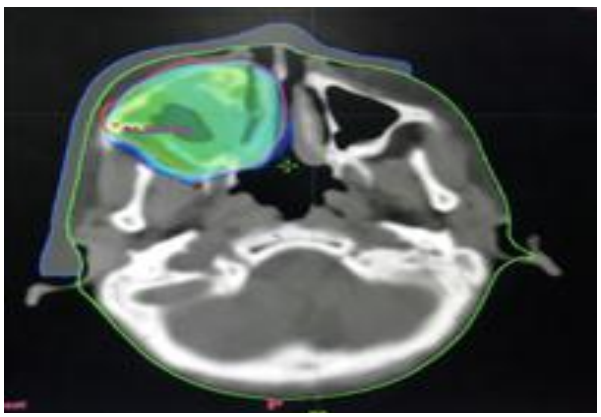

Fig 3: Dosimetric image of intensity modulated irradiation of Ewing's sarcoma of the maxillary sinus.

REFERENCES

1. Desai KI, Nadkarni TD, Goel A, Muzumdar DP, Naresh KN, Nair CN.PrimaryEwing'ssarcoma of the cranium. Neurosurgery, 2000; 46: 62-8.
2. Agrawal A, Dulani R, Mahadevan A, Vagaha SJ, Vagha J, Shankar SK.Primary Ewing′ssarcoma of the frontal bonewithintracranial extension. J Cancer ResTher., 2009; 5: 208-9.
3. Pedro Infante-Cossio, Jose Luis Gutierrez-Perez, Alberto Garcia- Perla, Manuel Noguer-Mediavilla, Francisco Gavilan-Carrasco. Primary Ewing's sarcoma of the maxilla and zygoma: report of a case. doi:http://dx.doi.org/10.1016/j.joms.2005.06.01
4. Jia Yu-Lin, Sedhain Bishwo P, Nie Xiu, Zhang Han-Dong. A maxillary sinus tumor in an adolescent girl, Huazhong University of Science and Technology, Wuhan, China. Oral Surg Oral Med Oral Pathol Oral Radiol, 2012; 114: 683–688.
5. Fiorillo A, Tranfa F, Canale G, et al. Primary Ewing's sarcoma of the maxilla, a rare and curable localization: report of two new cases, successfully treated by radiotherapy and systemic chemotherapy. Cancer Lett., 1996; 103(2): 177–182.
6. Carvajal R, Meyers P, et al. Ewing's sarcoma and primitive neuroectodermal family of tumors. doi:http://dx.doi.org/10.1016/j. hoc.2005.03.004.
7. N.G.Nikitakis,A.R.Salama,B.W.O'MalleyJr.,R.A.Ord,andJ. C. Papadimitriou, "Malignant peripheral primitive neuroectodermal tumor-peripheral neuroepithelioma of the head and neck: a clinic pathologic study of five cases and review of theliterature,"Head and Neck, 25(6): 488–498.
8. PAULUSSEN M., AHRENS S., DUNST J. et al. Localized Ewing tumor of bone: Final results of thecooperative Ewing sarcomastudy CESS 86. J. Clin. Oncol, 2001; 19: 1818-1829.
9. RODRIGUEZ-GALINDO C., NAVID F., LIU T. et alPronostic factors for local and distant control in Ewing sarcomafamily of tumors. Ann. Oncol, 2008; 19: 814-820.
10. A.Schuck,S.Ahrens,M.Paulussenetal.,"Localtherapyin localized Ewing tumors: results of 1058 patients treated in the CESS 81, CESS 86, and EICESS 92 trials,"International Journal of Radiation Oncology, Biology, Physics, 2003; 55(1): 168–177.
11. M. J. Krasin, A. M. Davidoff, C. Rodriguez-Galindo et al., Definitive surgery and multiagent systemic therapy forpatints with localized Ewing sarcoma family of tumors: local outcome and prognostic factors, Cancer, 2005; 104(2): 367-373.
12. APPELBAUM M.A. Clinicalfeatures and outcomes in patients with extra-skeletal Ewing sarcoma. Cancer, 2011; 117: 3027-3032.
13. Euro-E.W.I.N.G. 99 European Ewing tumourWorking Initiative of National Groups Ewing Tumour Studies, 1999.
14. Leavey PJ, Mascarenhas L, Marina N, et al. Prognostic factors for patients with Ewing sarcoma (EWS) at first recurrence following multi-modality therapy: a report from the Children's Oncology Group. Pediatr Blood Cancer, 2008; 51(3): 334–338.
15. Rodriguez-Galindo C, Billups CA, Kun LE, et al. Survival after recurrence of Ewing tumors: the St. Jude Children's Research Hospital Experience, 1979–1999. Cancer, 2002; 94: 561–569.