

Ewing's Sarcoma of the Mandible: a Case Report

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ABSTRACT

Ewing's sarcoma of the mandible is a rare disease. The treatment is based on surgery and chemotherapy. We report a case of a seventeen-year-old girl diagnosed and treated for an Ewing's sarcoma of mandible.

Keywords: Cricotracheal stenosis, cricotracheal resection, single stage surgery.

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INTRODUCTION

Ewing's sarcoma (ES) was described by James Ewing in 1921 [1]. It's the second most common bone primary malignant tumors described in children, following osteosarcoma. ES is an aggressive tumor with poor differentiation, commonly arising in the diaphyses of long bones, ribs, pelvis and vertebrae at the ages of 10 to 15 years [2,3]. More than 50% of ES cases originate in the pelvis and long bones [4, 5, 6] with distant dissemination at diagnosis being the rule rather than the exception, reflecting its aggressive biological behavior [7]. Primary localization in the face is very rare and occurs in only 1% to 4% of cases [3]. Mandibular bone is affected more than maxillary bone, with only few cases reported.

CASE REPORT

A seventeen-year-old girl presented two months ago a painless in the right parotid region, with growths progressively, with limitation of mouth opening and difficulties in chewing. Panoramic radiograph noted an expansive osteocondansant process with resorption of the cortical. Cranial computed tomography and magnetic resonance imaging with contrast revealed a destructive lesion extensively involving the right mandible, measured 6,5*5,5 cm, with cortical's bone destruction and fracture. This mass reached the soft parts. Surgical bone biopsy confirmed mandibular infiltration by ES. Immunohistochemistry showed strong positivity for CD99. The pelvic thoraco abdominal CT was normal. Bone scintigraphy

showed hyperfixation at the level of the mandibular primary lesion. The patient was treated according to the Euro-EWING99 protocol with 6 cycles of neoadjuvant chemotherapy regimens combining vincristine, ifosfamide, doxorubicin and etoposide. At the end of this chemotherapy, the iconography revealed persistent swelling of the mandible with a tumor volume decrease of more than 70%. The surgical procedure consisted of complete excision of the mandible with reconstruction by osteosynthesis. Anatomopathological examination on surgical specimen showed a diffuse bone and gingival fibroinflammatory remodeling without viable tumor residue with no cervical lymph node metastasis. The post operator treatment consist on adjuvant chemotherapy associated vincristin, actinomycin and ifosfamide.

DISCUSSION

Ewing's sarcoma is the second most common primary malignant tumor of the bone in children and adolescents [2]. The site of predilection is mostly the posterior regions with a 4:1 ratio compared to anterior regions [3]. The review of the English language literature from 1950 to 2017 revealed 102 cases of ES family tumor in the jaws. Interestingly, the majority of these cases were reported within the past decade, possibly indicating an increased awareness of the diagnosis [8]. In the head and neck region, there are nonspecific clinical findings for ES; most of the patient's complaints at time of presentation are due to the mass effect of the tumor, its rapid growth, the swelling of the affected area and the pain sensation. Ewing's sarcoma is a poorly differentiated neurectodermal tumor with small, round and blue cells [9]. Histopathologically, ES is composed of small, poorly differentiated cells with medium size, round or oval nuclei exhibiting a fine chromatin pattern, small nucleoli and scanty cytoplasm [10,11]. The intracytoplasmic glycogen may be demonstrated by PAS stain in 75% of the cases, but it is not pathognomonic and conclusive because other small round cell may show the presence of glycogen as well [11,12]. Due to similarity

with many malignant tumors, the diagnosis of ES can be very difficult and the lesion must be differentiated from other small round cell tumors, such as small cell osteosarcoma, mesenchymal chondrosarcoma, embryonal rhabdomyosarcoma, neuroblastoma and lymphoma [13]. The use of immunohistochemistry has helped in the diagnosis of this tumor. More than 90% of cases show a characteristic translocation $t(11;22)(q24;q12)$ resulting in the fusion of the EWS and FLI-1 genes [14]. This gene rearrangement causes a fusion product which functions as an oncogenic aberrant transcription factor with structural variability and potentially prognostic impact. Immunoreactivity against FLI 1 and CD99 can confirm the diagnosis [9]. There are other chromosomal abnormalities such as multiple numerical and structural aberrations and the loss of chromosomes 5, 8, 9, 10, 12 and X [15].

Plain radiographs are the first tool for determining the presence of bone lesions and remain the most useful imaging technique for suggesting ES. Generally, ES presents as a lytic, permeative, poorly defined lesion. However, "onion skin" periosteal reaction, which is a common finding in long bones, occurs rarely in the mandible [10].

Combined therapy including surgery, radiotherapy and chemotherapy is the best approach for ES [13,16]. Multidisciplinary treatment protocols have dramatically improved the 5-year survival rate of patients from 16 to 75%. Radiotherapy can treat nonresectable primaries and chemotherapy can suppress micrometastasis and reduce tumor load before surgery. The chemotherapeutic agents commonly used are vincristine, doxorubicin, cyclophosphamide, ifosfamide and actinomycin-D [8]. ES has poor prognosis because of hematogenous spread and lung metastases occur rapidly. Systemic symptoms, high erythrocyte sedimentation rate, elevated serum lactate dehydrogenase levels and thrombocytosis are poor prognostic indicator. Major prognostic factors include the tumor site and its volume as well as the presence of metastases. However, tumors in jaws have a better prognosis than those in long bones.

REFERENCES

- [1] Vaccani JP, Forte V, De Jong AL, et al. Ewing's sarcoma of the head and neck in children. *Int J Pediatr Otorhinolaryngol* 1999;48:209-16.
- [2] 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;103:177-82.
- [3] Siegal GP, Oliver WR, Reinus WR, et al. Primary Ewing's sarcoma involving the bones of the head and neck. *Cancer* 1987;60:2829-40.
- [4] Vikas PB, Ahmed MBR, Bastian TS, et al. Ewing's sarcoma of the maxilla. *Indian J Dent Res* 2008 Jan-Mar;19(1): 66-69.
- [5] Da Fonseca MA, Abrams RB. Ewing's sarcoma of the mandible in a young patient: case report. *Pediatr Dent* 1992 Nov-Dec;14(6):402-404.
- [6] Infante-Cossio P, Gutierrez-Perez JL, Garcia-Perla A, et al. Primary Ewing's sarcoma of the maxilla and zygoma: report of a case. *J Oral Maxillofac Surg* 2005 Oct;63(10):1539-1542.
- [7] Heare T, Hensley MA, Dell'Orfano S. Bone tumors: osteosarcoma and Ewing's sarcoma. *Curr Opin Pediatr* 2009 Jun;21(3):365-372.
- [8] Casaroto AR, Sampieri MBDS, Soares CT, et al. Ewing's Sarcoma Family Tumors in the Jaws: Case Report, Immunohistochemical Analysis and Literature Review. *In Vivo*. 1 mai 2017;31(3):481-91.
- [9] Solomon L, Frustino J, Loree T, et al. Ewing's sarcoma of the mandibular condyle: Multidisciplinary management optimizes outcome. *Head Neck* 2008; 30:405– 410, Wiley InterScience VVC 2007 Wiley Periodicals, Inc.
- [10] Gorospe L, Fernández-Gil MA, García-Raya P, et al. Ewing's sarcoma of the mandible: radiologic features with emphasis on magnetic resonance appearance. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod*. 2001 Jun;91(6):728-34. PMID:11402290
- [11] Talesb KT, Motamedi MH, Jeihounian M. Ewing's sarcoma of the mandibular condyle: report of a case. *J Oral Maxillofac Surg* 2003;61:1216-1219.
- [12] Quesada JL, Alcalde JM, Espinosa JM, et al. Ewing's sarcoma of the mandible. *J Laryngol Otol* 2003;117:736-738.
- [13] Fonseca AS, Mezzalira R, Crespo AN, et al. Ewing's sarcoma of the head and neck. *São Paulo Med J* 2000;118:198-200.
- [14] Gosau M, Baumhoer D, Ihrler S, et al. Ewing's sarcoma of the mandible mimicking an odontogenic abscess – a case report. *Head & Face Medicine* 2008;4:24.
- [15] Manor E, Kapelushnik J, Brennan PA, et al. chromosomal aberrations in primary ewing sarcoma of the mandible. *J Oral Maxillofac Surg*. 2012 Sep;50(6):574-6. doi: 10.1016/j.bjoms.2011.09.016. Epub 2011 Oct 11. PMID:21996574
- [16] Brazão-Silva MT, Fernandes AV, Faria PR, et al. Ewing's Sarcoma of the Mandible in a Young Child. *Braz Dent J* (2010) 21(1) :74-79.

