Primitive neuroectodermal tumor in the jaw: report of a case

Toranzo Fernández José Martin * / Salgado-García Nickte-ha** / Santos-Díaz Miguel Ángel* * *

The neuroectodermal tumor is a malignant neoplasm of round cells originated from the neural crest. The occurrence of this tumor in head and neck is rare. A 5 month old Mexican patient with a primitive neuroectodermic tumor stage IV, in the jaw is reported.
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INTRODUCTION

The primitive neuroectodermic tumor (PNET) and the Ewing sarcoma are closely related, but they are not identical, they can happen in bony and soft tissues.[1-4] Stout described it for the first time in 1918.[5] It is the second most common bony tissue tumor in pediatric patients.

The annual incidence is 3.4 cases for each million of Caucasian American children, younger than 15 years old; compared to Afro-American children which is 0.6 for each million.[2] It is also known as peripheral neuroepithelioma, Askin's tumor and peripheral neuroblastoma.[6]

Most of the PNET are presented in the toracopulmonary region in 46% of the cases, continued in order of frequency by head and neck, superior and inferior members in 42%.

The metastasis is presented in 31% of the patients at the moment of the prognosis.[7] Only 3% of PNET´s have been reported in the jaw.[8]

The tumor has an aggressive local behavior and a high metastasis probability.[7, 9]

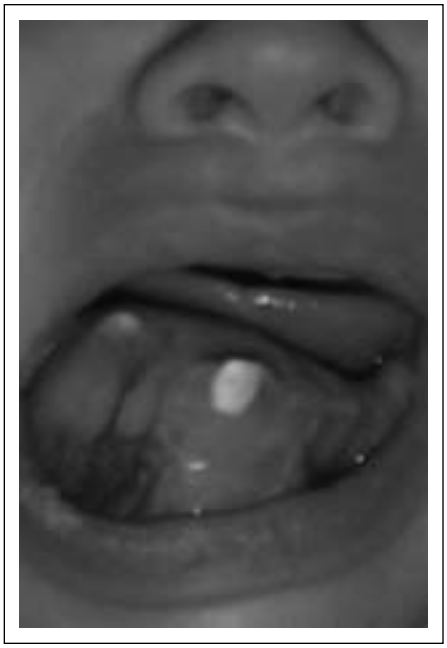


Figure 1. Image clinical intraoral. Female of 5 months with primitive neuroectodermal tumor in the jaw.

CLINICAL CASE

A 5 month old female, born in Esperanza, Guanajuato Mexico. She is the daughter of a young not consanguineous couple, without family records of importance. She was admitted to the ER of the Central Hospital "Dr. Ignacio Morones Prieto" of San Luis Potosí, Mexico, with a 3 week evolution of an increase of volume in the alveolar process of the lower jaw, with edema and pruritus previous to the eruption of the incisor teeth.

It was diagnosed as a possible cyst of the eruption and she received antibiotics and a non-steroidal anti-inflammatory for having presented fevers of 40° C (104° F) for a possible infection. She presented vital signs inside normal parameters, she weighed 7.5 Kg. (p50), and measured 65 cm. (p50).

An increase of volume of the skull was observed in occipital right region of approximately 5cm of diameter, without movement, nor pain. In the mouth, an increase of volume of 7 x 10 centimeters in alveolar process of the lower jaw with irregular borders and great vascularity, purplish coloration, with displacement of dental organs, fluctuating depressible bonded to adjacent tissues (Figure 1). Neck without adenopathies. The neurological exam and the rest of the exploration, anatomical and

* Toranzo Fernández José Martin, Chief of the Oral and Maxilofacial Surgery Department, Central Hospital "Dr. Ignacio Morones Prieto"

** Salgado-García Nickte-ha, 3d Year Resident of the Oral and Maxilofacial Surgery Department, Central Hospital "Dr. Ignacio Morones Prieto"

*** Santos-Díaz Miguel Ángel, Pediatrician and Geneticist, Central Hospital "Dr. Ignacio Morones Prieto"

Send all correspondence to Dr.Jose Martin Toranzo Fernandez, Vallejo #766, Barrio de San Miguelito, Zona Centro, San Luis Potosi, S.L.P., Mexico.

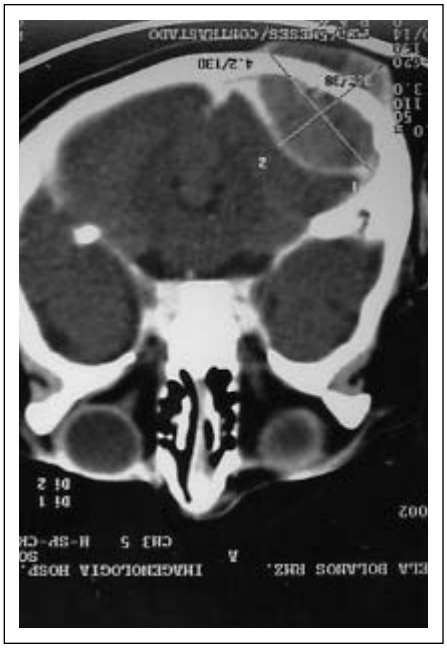


Figure 2. Tomography axial on-line contrasted skull where tumor is observed in later grave.

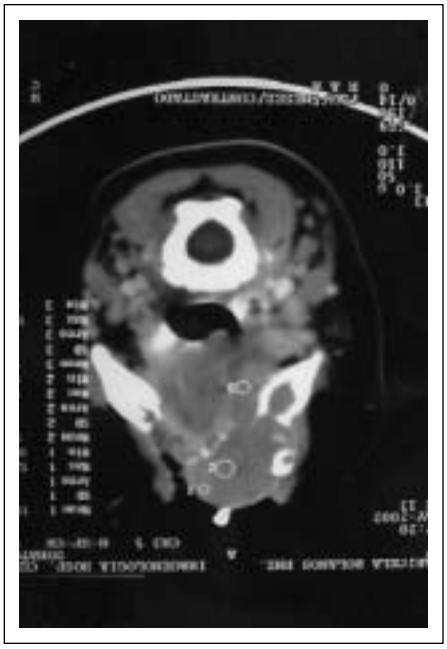


Figure 3. Facial Solid TAC contrasted in which tumoracion is observed in jaw of aggressive local behavior.

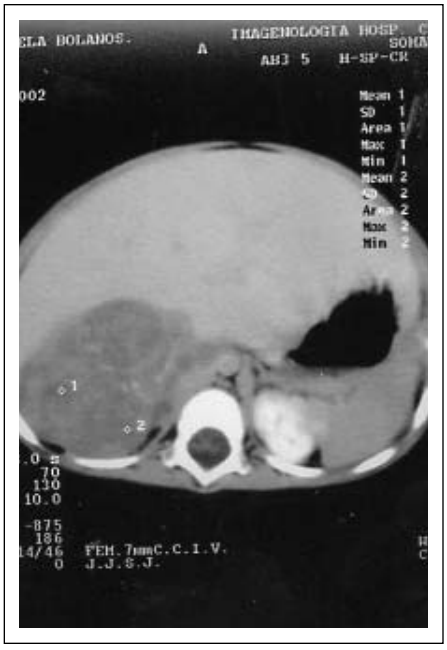


Figure 4. Abdominal TAC in which can observe for tumorous extravasacion in retroperitoneo, hepatic, renal.

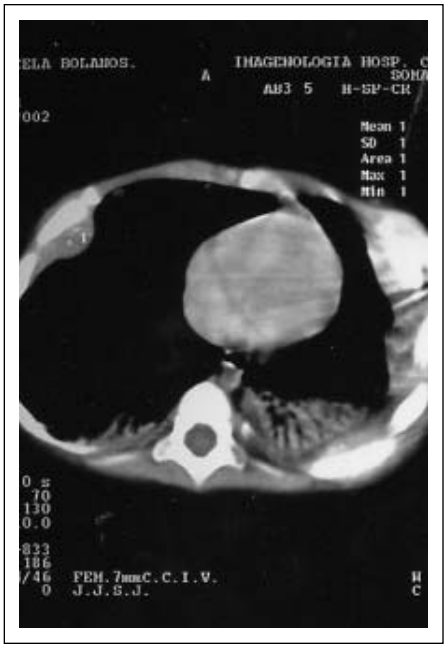
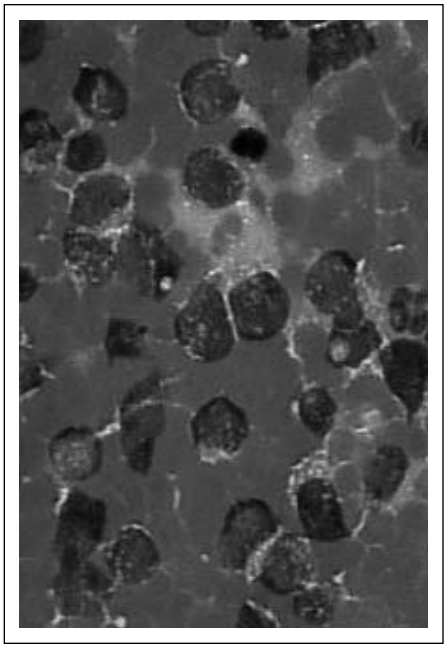


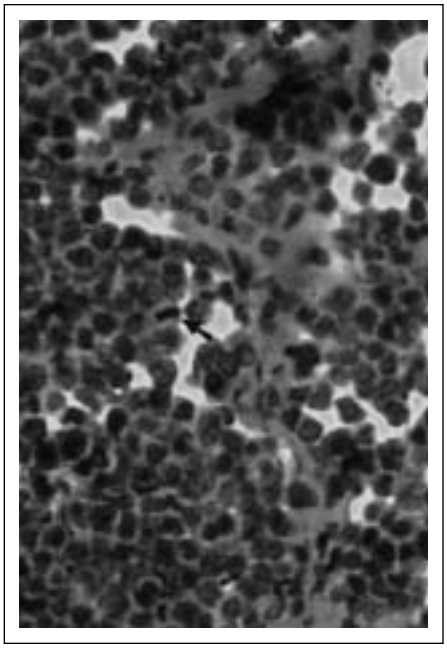
Figure 5. Thoracic TAC observes bony tumorous metastasis in right rib.

functionally normal. The laboratory exams showed: hypochromic microcytic anemia (Hb of 7 gr/dl), DHL 8784U/l.

The contrasted skull TAC (Figures 2) showed a hypodense lesion that was corroborated with the radio-opaque contrast medium that involved the bone in the posterior epidural pit and the soft tissue in the occipital region of 3.5 X 4.2 cm. In the facial TAC (Figures 3) a hypodense image was observed involving the mentoninan symphysis, body and right jaw angle, displacement of dental organs and bony erosion.

In extensive studies, the contrasted abdominal TAC (Figure 4) showed a heterogeneous hypodense lesion of 7.9 x 5.5 cm. in diameter in the superior right renal pole, as well as a conglomerate of ganglions retroperitoneal paracavales, intercavo-aortic. As well as lung metastasis and bony destruction of left costal arches (Figure 5). The bone





Figures 6 and 7. Microscopic image of tumoracion biopsy in jaw.

gamagram showed several bone metastases in the column, skull, and thorax.

The biopsy of the intraoral mass (Figure 6 and 7) showed the proliferation of small blue round cells with ovoid nuclei, mitosis, showing positiveness to the specific neuronal enolasa. The suction of bone marrow had enhanced cells, diminished megacariocites and infiltrated in a 70% with immature cells, scarce basofilo cytoplasm with abundant vacuole similar to the Burkitt type lymphoma and neoplasic cells. The hystopatological diagnosis was of a primitive neuroectodermic tumor stage IV.

Chemotherapy began with cisplatino, cyclophosphamide, and adriamicine during 5 sessions with poor results. The oral lesion presented changes in coloration and smaller vascularity but without perceptible decrease. Finally the patient was voluntarily released with tumorous activity.

DISCUSSION

Primitive neuroectodermal tumor or neuroepitelial are some of the names used to describe a tumor conformed by undifferentiated cells with germinal appearance or cellular matrix of the neural embryological tube. It is implicit in all the used names to refer to these cellular tumors, the capacity to differ along the glial lines. The term neuroectodermal has been used to describe the total substance of the central nervous system, the mesenquimal elements, blood vessels and microglia.

There are an extensive group of undifferentiated neoplasias that occur among children. These tumors are still an enigma because they are not common, for this reason, the primitive neuroectodermal tumors are generally called with a variety of names: neuroblastoma, cerebral meduloblastoma, undifferentiated cellular neoplasm.[10]

The PNET is presented in the thorax, femur, tibia, flue, pelvis, ribs, vertebrae, collar bone, jaw and skull, in the last, they can show certain characteristic, as thickness and epidural inclusion, adjacent commitment to the brain, as well as periostium and soft tissue.[4]

In the thorax it involves soft tissue and extra pulmonary tissue.

The abdominal occurrence is thought to be caused by the extravasation of the tumor, which crosses the diaphragm and the bone metastasis are relatively common. Any way the occurrence in the oral cavity is extremely rare.[11] In this case we can observe the different localizations and metastasis in not very requent places like the jaw.

The neuroectodermal tumors contain small blue round cells. Other tumors with the same histological pattern correspond to Ewing´s sarcoma, neuroblastoma, rhabdomyosarcoma and the non-Hodgkin lymphoma. It is characterized by the presence of big attachés of tumorous cells with an alveolar pattern separated by filaments of fibrous tissue. There are regular cells with oval nucleus.

The lightly granular cytoplasm, this framed by the cellular outline. The nucleus contains fine dispersed chromatin, giving it the appearance of grounded crystals. With occasional mitosis[4,12] some of the histopatoligcal characteristic can be observed under the microscope of our case. These tumors show Homer-Wright pseudorosetas under the microscopy and positive immunehistochemistry for sinaptofisine and specific neuronal enolasa.[1,13,14]

CD99 is observed in its membrane cells as a result of genes MIC215 and l6.95% contains translocation 11;22 or 21;22, the mechanism of the tumorogenesis remains uncertain.[7,17,18,19] The clinical manifestations depend on the localization of the tumor. The first symptoms are pain and swelling of the area close to adjacent tissues near the tumor. In the case of involving soft tissue, the tumor has a purplish color, fluctuating and with abundant vascularity. Fevers and increased heart frequency in 28% and metastasis can be present in 26% in the initial diagnose.[20]

The initial symptoms can mislead doctors to think it is an infectious problem, before there is a suspicion of a neoplasm.

PNET can clinically simulate in its first stages osteomyelitis for radiographic similarity and the presence of leucocytisis.[4] The most frequent place for metastasis is the lung and other bones.

Patients can look for the medical assistance due to the symptoms related to metastasis instead of the tumor.[12,21]

The local treatment should be achieved by means of surgical treatment, chemotherapy and radiotherapy. The size of the biggest tumor (more than 8 centimeter) or volume (more than 100ml) is correlated with the poor response to treatment.[22-24]

An initial phase of chemotherapy with 12-15 weeks of induction, to do the diagnosis biopsy avoids the complication of a future resection. The preoperative chemotherapy can reduce the size, the vascularity and the friability of the tumor, facilitating the resection and decreases the risk of rupture of the transoperatory tumor.[26]

In the traditional way, biopsies for these tumors have been done with open techniques and they can be difficult due to the extremely vascular nature of this tumor. The percutaneous biopsy is preferred in many cases since it provides the perfect material for histological and cytogenetic examinations.[25]

Right now, the normal chemotherapy for the non-metastasis illness consists of a régime from three to five drugs: leurocristine, doxorubicina, and cyclophosphamide, alternate with ifosfamida and etoposid for a total of 48 weeks.[2]

Historically, the presage for children and young adults is based on the size of the tumor, the situation and the magnitude.

The presage is poor, if it is associated with a) metastasis, b) the size of the tumor (bigger than 8 centimeters in diameter) and c) volume (bigger than 100 ml). Children with metastasis have a bad presage.

In several studies it has been described that high levels of lactic deshidrogenasa in serum have value presage. The necrosis and complete resection of the tumor plus the radiotherapy and complete chemotherapy have direct relationship with a favorable presage, with a survival rate of 5 years in 84% to 95%.[27,28]

The most frequent cause of failure in the therapeutic treatment is the systemic illness. Modern treatment outlines show a survival rate free of illness to 5 years in 73% of patients without metastasis evidence and of 39% in those that present pulmonary metastasis.[10]

The outlook for children that present metastasis, which involve the jaw are very poor and a short, but intensive course of radiotherapy consisting from 1 to 3 treatments for a total dose of 400 to 1200 e.g. besides palliatives for the pain is recommended.[29]

This reported case corresponds to a primitive neuroectodermal tumor with local aggressive behavior and with high metastasis probabilities and we consider it of interest due to the presentation in head and neck as primary tumor, which is a rare case. The early diagnosis as well as an appropriate surgical treatment can give a favorable result.

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