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Neuro-Ectodermic Tumor Primitive: About A Rare Case with A Parotid Location

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Abstract

Peripheral Primary neuroectodermaltumors (PNETs) are extremely aggressive small round cell tumors derived from neural crest cells. They belong to the family of Ewing's sarcoma [1]. Parotidial primitive neuroectodermaltumors are extremely rare. We propose, through the case of a patient treated in the hematology and oncology department of Rabat Children's Hospital, to study the clinical, radiological, anatomopathological and therapeutic aspects of tumors neuro-ectodermal peripheral Primary.

Keywords : neuro-ectodermictumor, child, parotide.

Introduction

Primary neuroectodermaltumors (TNEPs) are rare and very aggressive malignant tumors. They are made up of a proliferation of small round cells and developing mainly in the central nervous system [2]. Extra cranial and particular lyparotidlocalizationi sex ceptional. They belong to the family of Ewing's sarcoma. This family includes bone and extra osseous Ewing's sarcoma, A skin's tumor and peripheral neuroepithelioma or pPNET [3, 4]. This reconciliation between these different pathological entities was only possible thanks to the cytogenetic data and the demonstration of the translocation t (11,22) (q24, 12) [5, 6].

Case Report

15-year-old patient with no particular antecedents. Step resented 6 months before he hospitalization, a parotid right swelling rapid lyincreasing volume with second reappearance of ipsilateral facial and coulometer paralysis, all evolving in a context of deterioration of the general state. The clinical examination found a conscious patient, in poor general condition with a parotid right swelling, hard and painful on palpation, without inflammatory signs opposite, fixed in both planes, irregular contours 12 / 10.5 cm and deviating the flag right ear (Figure 1). The endo buccal examination showed an or pharyngeal swelling of firm consistency pushing back the right amygdale. In addition, the cervical examination did not findly mphadenopathy. The Peripheral right facial palsy with ipsilateraloculomotorpalsy was noted and ophthalmologic examination revealed ophthalmoplegia in the right eye. The rest of the somatic examination was peculiar. Cervical ultrasound showed a large angulomaxillary tumor, heterogeneous tissue with boneless, complemented by a cervical scan that showed a roughly rounded right parotid lobe formation measuring 10 * 6 * 8 cm. evoking evolutionary lesion with contact mastoid it is (Figure 2). The biopsy with histological study showed a small round cell proliferation, little differentiated, arranged in rosettes. The immunohistochemicalanaly sis revealed a positive labeling for the anti-enolaseanti bodies specific for the NSE neuron, protein S-100 and also for the CD99 which corresponds to the product of the MIC2 gene thus establishing the diagnosis of PNET. An extension

assessment was performed: the chest X-ray, the abdominal ultrasound, medullogram and osteomedullary biopsy, returned normal outside the bones cintigraphy which showed a zygomatic and right occipital hyper fixation related to bone involvement (Figure3). The surgical approach was difficult because of the extension of the lesions and as a result the patient received firstly multi drug therapy IVA (ifosfamide + vincristine + action mycin). The evolution was marked by the establishment of a syndrome of intracranial hypertension secondary to a thrombosis of the right jugular gulf extended to the ipsilateral sigmoid sinus on the cerebralangio scan (Figure 4), and by the increase of the tumoral volume on control CT (Figure 5). The patient later presented with a disorder of consciousness and died on day 7 of the 2nd cure.

Discussion

Peripheral neuroectodermaltumors are highly aggressive and rare tumors, their frequency is estimated at 1.1% of soft-tissue tumors in the general population and between 6.3 and 17% in the pediatric population, in itially described by STOUT in 1918 in the ulnar nerve under the term peripheral neuroepithelioma [7], are known in the literature under different names: primary neuroectodermaltumors, malignant neuroepithelioma or peripheral neuroblastoma, primary soft tissue neuroectodermaltumors or soft tissue peripheral neuroectodermal sarcoma [8] . Beside bone sites (Ewing's sarcoma), about half of the PNETs are developed at the expense of soft tissues, with a clear predilection (40%) for the thoraco-pulmonary region (A skin tumor), extremities (25 %), cervico facial regions and pelvis. [9]. According to the data of the literature, only 4 cases of PNET with parotidlocalization were reported with extreme age between 5 and 60 years, again the clinical course was rapid with an average of 6 months, 2 cases presented a peripheral facial paralysis as in our patient. All patients benefited from imaging (computed tomography) which is useful for delineating the limits of respectability, in the distant detection of metastases, and in the evaluation of the response to treatment, but the definitive diagnosis of PNET was made on histopathological and immune histochemical analysis: malignant proliferation of small, round, mono morphic and poorly differentiated cells. The fickle presence of neural differentiation elements such as rosettes or pseudo-rosettes with a fibrillar center (from Homer Wright) suggests their neuroectodermal origin, which is associated with immune histochemical analysis which is marked by at least two markers of differentiation. Neuronal (S100 protein, NSE, chromogranin A CGA, synaptophysin). Over expression of the MIC2 gneisses in nearly 84 to 100% of PNETs and over 95% of Ewing's sarcomas [1,6, 9]. The cytogenetic study is very useful in case of doubt diagnosis, since more than 90% of the PNETs have translocation t (11,22) (q24, q12), It could not be realized for our patient for lack of means . There is no clear therapeutic consensus for the management of PNETs. The treatment is super imposable to that of Ewing's sarcoma. It associates a radical surgical resection, the facial nerve is most often sacrificed, Chemotherapy makes it possible to optimize the local treatment and to control the metastatic disease. It is continued after the local treatment to reach a total treatment time of about one year. Several chemotherapy regimens based on cyclophosphamide, doxorubicin, vincristine, etoposide, busulfan, mephalan and carboplatin are currently used. Radiation therapy may be used alone in case of tumors that are surgically inextensible, but also and more often in addition to excision to help loco regional control, especially in cases of positive surgical margins [10,11]. Given the tumor extension in our patient, the surgical procedure was not retained. The evolution is ofte nun favorable in relation to loco regional extension or a metastatic stage from the outset. The mortality rate is about

50%. The most pejorative prognostic factor seems to be the presence of metastases, especially pulmonary, clinically detectable at the time of diagnosis. The average survival time is 12 months [12].

Conclusion

PNETs with parotid localization are rare and very aggressive malignant tumors. Their positive diagnosis is mainly based on immune histochemistry, which can often be characterized and distinguished from other small round-cell tumors. Cytogenetics may be useful in case of doubt diagnosis. Their treatment is multidisciplinary, calling on the surgeon, oncologist and radiotherapist.

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Legends Figures

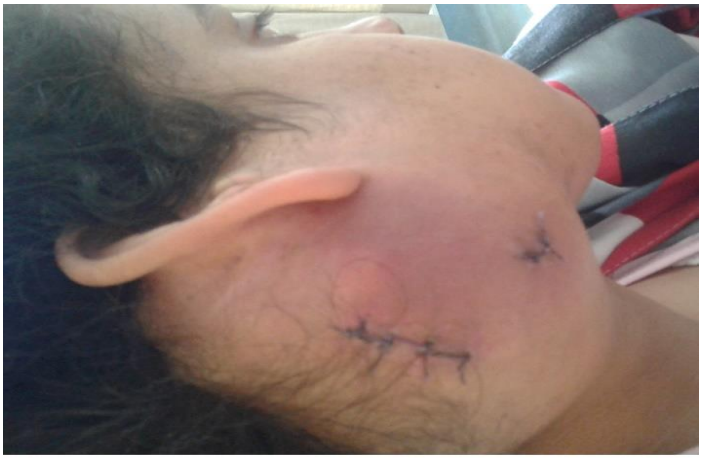

Figure 1: right parotid swelling, hard, without inflammatory signs opposite, fixed, irregular contours making 12 / 10.5 cm and deviating the flag of the right ear.

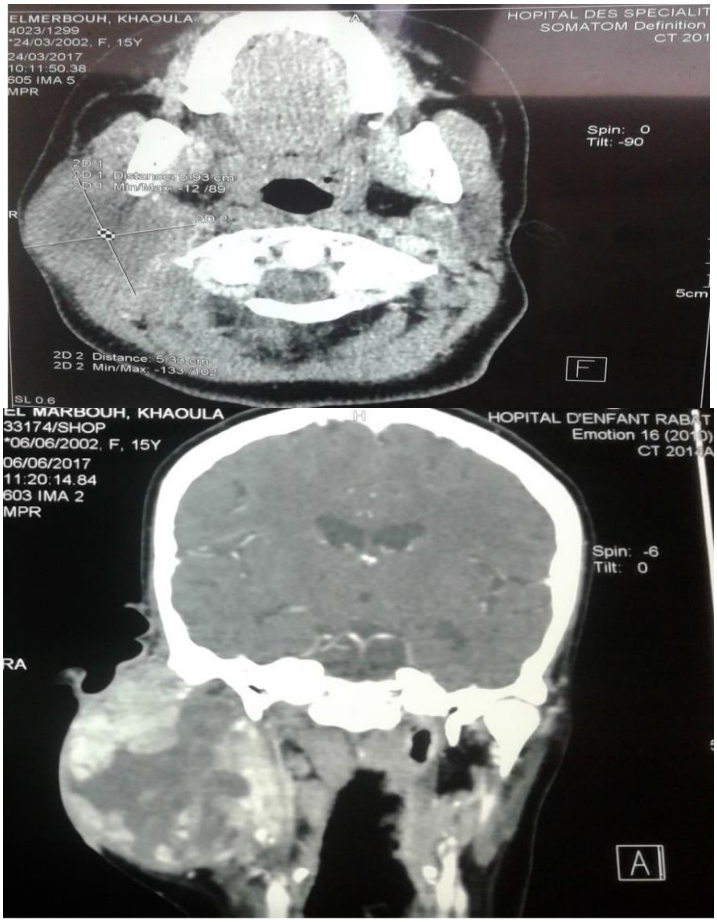

Figure 2: Cervical CT showing a roughly rounded right parotid lobe formation measuring 10 * 6 * 8cm evoking an evaluative lesion with mastoid it is on contact

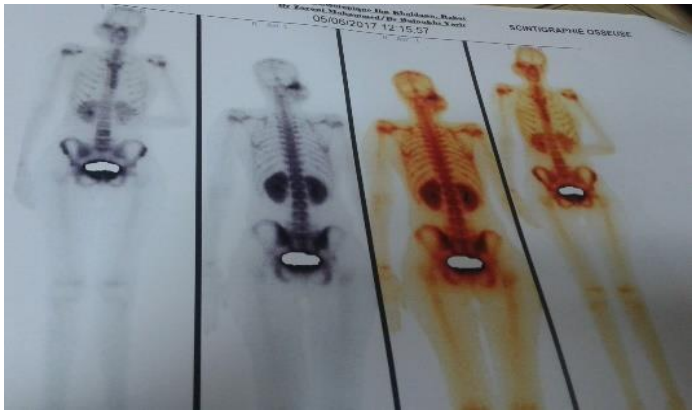

Figure 3: bones cintigraphy showing zygomatic and right occipital hyper fixation in relation to bone involvement

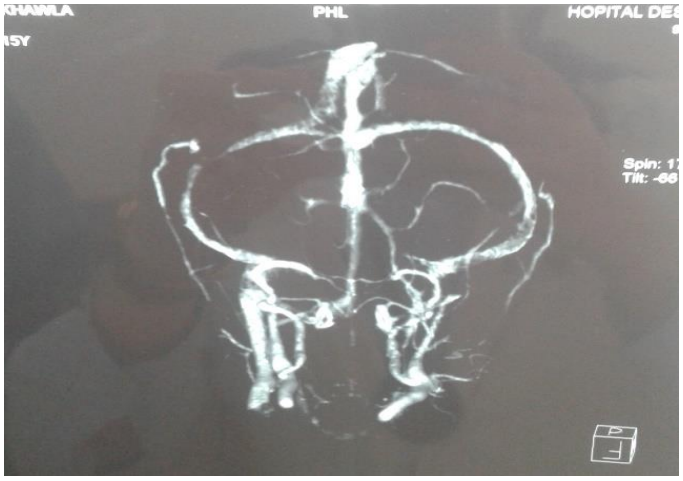

Figure 4: Cerebral CT angiography showing thrombosis of the right jugular gulf extended to the IPSI lateral sigmoid sinus

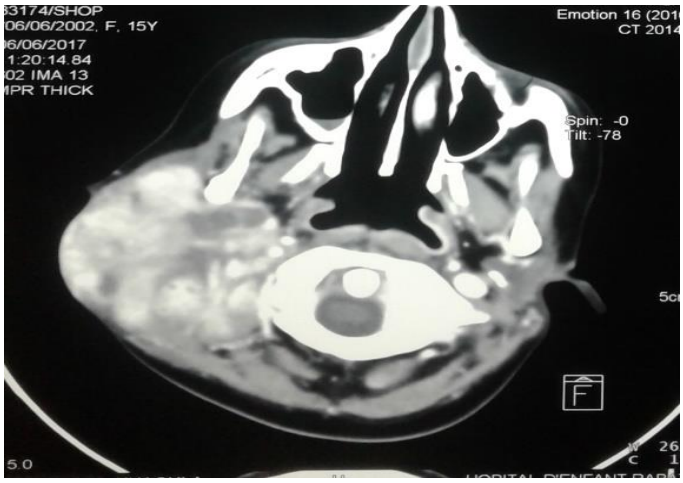

Figure 5: Control CT after the first course showing an increase in tumor volume