Case Report
Volume 5:1,2020
DOI: 10.37421/2577-0535.2020.5.47

Journal of Cancer Clinical Trials

ISSN: 2577-0535 **Open Access**

Primitive Neuroectodermal Tumor (PNET) of the Parotid Gland in a 13-year-old Female: A Rare Case Report

Ait Taleb Oum'hand Hajar*, Mahiou Noureddine , Belhaj Najoua, Nitassi Sophia, Bencheikh Razika, Benbouzid M A, Oujilal Abdelilah and Essakalli Leila

ENT Department, Hospital of Speciality of Rabat, University Hospital Avicenne, Rabat, Morocco

Abstract

Primitive neuroectodermal tumor (PNET) belongs to the Ewing family of tumors. It's a rare and highly malignant small round cell tumor that generally occurs in adolescents and young adults. Its location at the parotid gland is extremely unusual. Here we describe a rare case of 13-year-old female who presented with painful, progressively enlarging and swelling mass located in the right parotid gland.

Keywords: Primitive Neuroectodermal Tumor •Parotid Gland

Introduction

PNETs are rare small round cell tumors occurring in bone and soft tissues, characterized as a group by the presence of the typical translocation t (11; 22) (q24; q12) and its variants [1,2]. Outside the central nervous system PNET is called peripheral primitive neuroectodermal tumour (pPNET).They most commonly occur in the thoracopulmonary region (Askin ' s tumor), abdomen, pelvis, extremities and rarely in the head and neck [2].Diagnosis requires histopathological examination, immunohistochemistry and cytogenetics. Here, we report a case of pPNET of parotid in a 13-year-old female.

Case Report

A 13-year-old female with no noteworthy medical family history or past history consulted our department due to the appearance of a painful, progressively enlarging and swelling mass located in the right parotid gland that had been growing for 6 months. On physical examination, a firm nontender fixed mass (6 × 4 cm) was found on the right parotid gland extending to the retroparotid area, with a homolateral facial paralysis grade III according to House-Brackmann facial paralysis scale [3]. Multiple right laterocervical indurate lymph nodes were also identifed (Figure 1).

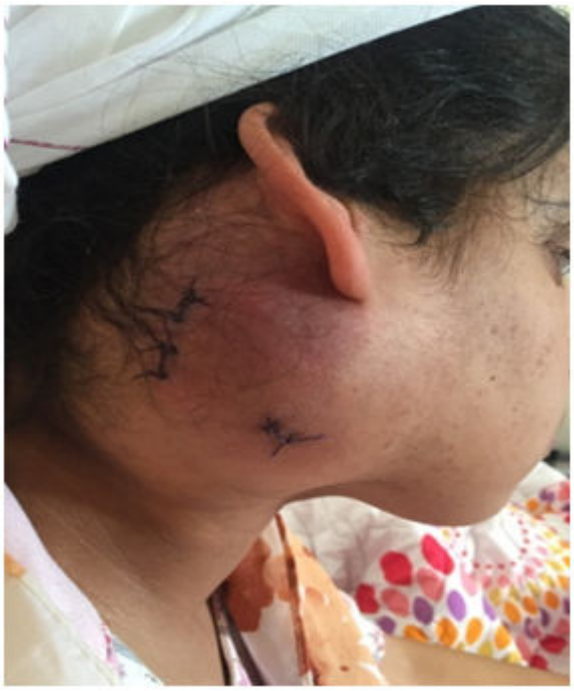

Figure 1: lateral view of the patient showing the mass of the right parotid gland extending to the retroparotid area.

A computerized tomography (CT) scan of the headneck was indicative of a relatively well-defined heterogeneous, moderately enhancing soft tissue mass of the right parotid gland (white star). The mass was ~6.2 × 4.2 × 5.8 cm, isodense to the muscle involving the

***Corresponding author:** Ait Taleb Oum' hand Hajar, ENT Department, Hospital of Speciality of Rabat, University Hospital Avicenne, Morocco; E-mail: dr.hajaraitaleb@gmail.com*

***Received:** April 28, 2020; **Accepted:** May 12, 2020; **Published:** May 19, 2020*

superficial and deep lobe of the gland without reaching the parapharyngeal space(red arrow). It infiltrates the retro-stylian-space, compresses the right jugular vein with an infiltration of the right carotid artery which remains permeable (blue arrow) and comes into contact with the mastoid with bone lysis (Figure 2).

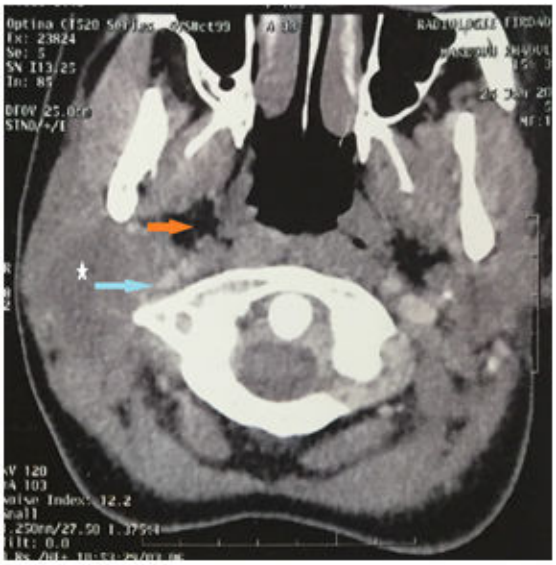

Figure 2: Contrast enhanced axial soft tissue window CT showing the mass involving the right parotid gland.

T1, T2, STIR and T1 FAT SAT weighted axial and coronal sequences with and without contrast MRI was done to ascertain the anatomical extent of the tumor and exclude intracranial involvement A large nodular mass of 61× 47 mm developed mainly at the external lobe of the right parotid, slightly extended to the internal lobe. The parotid lesion was intensified at the periphery with a central necrotic area (white arrow). There was no invasion of the right mandible, or intracranial extension neither the parapharyngeal space. Bilateral jugular lymphadenopathy have been also identified (Figure 3).

CT scans of the thorax and abdomen did not reveal any involvement of other sites.

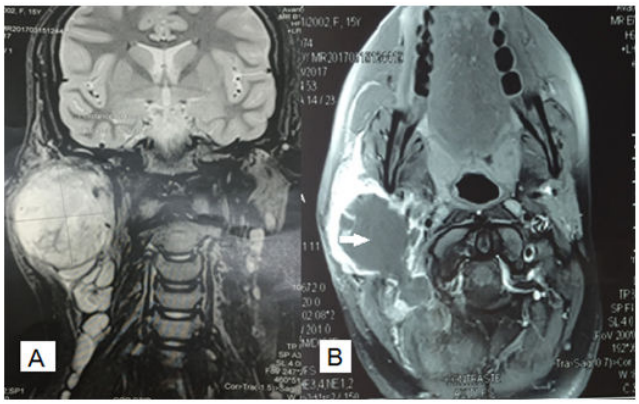

Figure 3: Post contrast T1 STIR coronal (A) and FAT-SAT axial (B) MRI showing a large heterogeneously enhancing mass with necrotic areas (arrow in B).

Owing to the requirement for obtaining a histopathological diagnosis, incisional biopsy was performed under local anesthesia. Histopathological examination showed a large necrotic round cell tumor proliferation. The round cells are of lymphocytoid type, with hyperchromatic nuclei (Figure 4). Mitoses and apoptotic bodies were also identified. Immunohistochemical study was done. The immunohistochemistry showed a Ki-67 Proliferation Index estimated at 40%, negative CD20 and CD3 and negative cytokeratin AE1/AE3, however strong positivity for CD99 was confirmed. On the basis of these findings, a final diagnosis of pPNET was established.

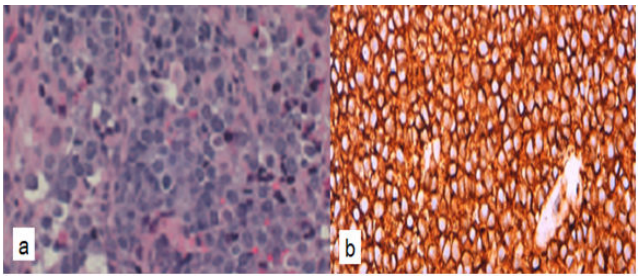

Figure 4: (a) view of the biopsy specimen showing a pleomorphic cellular infiltrate with hyperchromatic small cells with irregular nuclear contours and proeminent nucleoli (hematoxylin and eosin staining; original magnification, x4000) (b) Immunohistochemical staining showing strong positivity for MIC2 (CD99).

A multidisciplinary consultation meeting was carried out directly after the confirmation of the pPNET diagnosis. It was decided to treat the patient according to three phases: Induction chemotherapy followed by surgery then adjuvant chemotherapy. In order to receive the first phase of treatment the patient was referred to pediatric oncology center. Induction chemotherapy phase was based on 3 cycles of multidrug therapy (Vincristine-Cyclophosphamide-Doxorubicine-Actinomycine D).

At the end of the second chemotherapy session the patient had an acute chest pain associated with dyspnoea of recent onset, tachycardia: Pulse rate =120 beats/min and an excessive decreased of oxygen saturation.

Considering the high clinical probability of acute Pulmonary embolism (PE) anticoagulant treatment was initiated without delay based on a weight-adjusted bolus of Unfractionated Heparin (UFH). Subsequently, a CT angiography was performed confirming the diagnosis of massive bilateral pulmonary embolism. The patient was urgently scheduled for thrombolysis but she succumbed following a cardiogenic shock.

Discussion

Ewing's sarcoma (EWS)/peripheral primitive neuroectodermal tumour (pPNET) are small round cell tumours derived from tissues outside the central and autonomic nervous system [1,2]. pPNET was first introduced by Stout in 1918 [4]. It is a rare disease that comprise only around 1% of all the sarcomas, most common in the thoracopulmonary region (Askin's tumor), pelvis, abdomen and extremities with a slight male predominance [5,6]. Most studies have reported that the head and neck region is an infrequent site of involvement, in this area the most common location of occurrence is the orbit, followed by neck and then the parotid gland [7,8]. In a review of the English literature, it was found that pPNET of the parotid gland had only been reported in adults, aged between 15 and 60 years old [6]. Clinically they are usually rapidly growing, painful and locally aggressive tumors.

Despite the fact that the radiological features of the tumors are nonspecific and frequently cannot be differentiated from those of other types of bone and softtissue tumors. The primary role of computerized tomography (CT) and MRI is to estimate the extent of the disease and presence of adequate margins for resection. CT may show cortical bone involvement better, but MRI offers better soft tissue resolution and may be mandatory for intracranial extension and marrow infiltration. In fact PNETs are known to have internal hemorrhage and necrosis [9] in this patient, MRI images were corresponding to central necrotic area of the right parotid gland which was confirmed later at pathological examination.

Diagnosis requires histopathological examination, immunohistochemistry and cytogenetics. Microscopically, PNET are composed of uniform small round cells with round nuclei containing fine chromatin, scanty clear or eosinophillic cytoplasm, and indistinct cytoplasmic membranes[10]. The differentiation of PNET from Ewing's sarcoma is based on the presence of neural differentiation which is indicated by histological evidence of Homer-Wright rosettes. They are characterized by immunoreactivity for the surface antigen CD99/ MIC2, which is expressed in up to 97%of cases[11].Cytogenetic Studies showed that this family of tumors have a characteristic chromosomal translocation t(11; 22) (q24; q12) leading to the EWS gene mutation, resulting in EWS- FLI1 fusion protein formation [12,13].

Due to the rare occurrence of pPNET, optimal therapy is challenging particularly when the tumor is localized in the head and neck area. The use of a systematic multidisciplinary consultation meeting at each stage including before the biopsy is mandatory. Endeed successful treatment requires using a combination of therapeutic modalities including:

-Induction multidrug chemotherapy based on : Anthracyclines such as Doxorubicin, alkylating agents as Cyclophosphamide, ifosfamide, vincristine, dactinomycin, etoposide ...

-Surgical excision of the primary tumor if it's possible and if it doesn't delay the course of the chemotherapy (surgery planned in advance)

-Conventional or intensified consolidation chemotherapy [12,14].

Radiotherapy is discussed depending on the resection margins, the response to chemotherapy as well as the anatomical situation notably in cases with proximity of vital structures and in inoperable tumors [15].

In order to detect local or metastatic relapse at a time when treatment is possible and effective, it is essential to establish clinical and radiological tumor surveillance including a chest X-ray, CT scan of the lungs, a bone scan, and bone marrow aspiration [15,16]. Because of the tendency for recurrence and early distant metastasis to lung, liver, and bone marrow, the overall prognosis of PNET is poor. Its disease -free survival rate is less than 50% in 3 years and 30%–45% in 5 years. It seems that primary tumor location is an important prognostic factor [12,17]. In our case, the patient had a massive pulmonary embolism after her second session of chemotherapy. Despite emergency admission in intensive care and heparin administration, the patient presented a cardiogenic shock which led to her death.

According to medical literature tumor disease multiplies the risk of thromboembolic complications by 4 [18].If death is sometimes linked to the metastatic course of the cancer disease, it can occur early in as a result of massive pulmonary embolism (PE) or a serious anticoagulant accident which can lead the patient in intensive care [19].

The prevalence of PE is significantly higher in patients with cancer. A Norwegian study reporting the results of 6,197 autopsies of carrier patients of cancer found PE in 10.5% of cases (compared to 8.4% in the absence of cancer) [20].Chemotherapy was identified as a risk factor for Venous thromboembolism (VTE). In an American case-control study carried out in 2000, the risk of VTE was multiplied by 6.5 (95% CI: 2.1–20.2) in cancer patients receiving chemotherapy and 4.1 (95% CI: 1.9 – 8.5) in cancer patients not receiving chemotherapy compared to non-cancer patients [21]. The mechanisms evoked to explain this increased thrombotic risk are multiple and probably included: direct endothelial toxicity, apoptosis and cellular necrosis with expression of tissue factor, release of pro-inflammatory cytokines, platelet activation, induced hemostasis abnormalities (decrease in protein C, protein S, antithrombin III, increased PAI-1) with hypercoagulability [22,23].

Conclusion

PNET is a rare and aggressive tumor with poor prognosis, in particular due to complex diagnostic and multidisciplinary but not yet codified treatment combining most often surgery, radiotherapy and chemotherapy

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How to cite this article: Ait Taleb Oum'hand Hajar, Mahiou Noureddine , Belhaj Najoua and Nitassi Sophia et al. "Primitive Neuroectodermal Tumor (PNET) of the Parotid Gland in a 13-year-old Female: A Rare Case Report". *JCancer Clin Trials* 5 (2020) doi:10.37421/2577-0535.2020.5.47