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Primitive Neuroectodermal Tumour of the Maxilla Sinistra Region

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Abstract

Primitive neuroectodermal tumors (PNET) are a group of malignant tumors composed of small round cells of neuroectodermal origin that affect soft tissue and bone. PNET have a rare incidence, occurring usualy in children and adolescents, rarely in adults. In this case report are focus of Peripheral primitive neuroektodermal tumors. Peripheral primitive neuroectodermal tumors (PNETp) are a group of aggressive malignancies that most commonly present in multiple tissues and organs including kidney, adrenal, bladder, liver, small intestine, colon and rectum, with a preferred location within the chest area (thoracopulmonary region), in the limbs and around the spine, but rarely, in the head and neck area. Here we present a case of the maxilla sinistra PNETp in a 17-year-old girl and we discuss the clinical features, histopathological characteristics, diagnosis, and treatment.

Keywords: primitive neuroectodermal tumors, maxilla malignancy, immunohistochemical

INTRODUCTION

A primitive neuroectodermal tumor (PNET) has been known for a long time as soft tissue tumors that are often found in children and adolescents. These tumors belong to the Ewing sarcoma (ES) which is a group of malignant tumors consisting of small round cells, derived from neuroectodermal tissue that damages soft tissue and bone. Neuroectodermal tissue is a layer of outer nerve tissue originating from ectoderm embryonic tissue, which develops into the tissue forming the skin and outer nerves (neuroectodermal).[1]

Stout,[2] first reported a case of PNET in 1918, and this tumor is thought to originate from nerve tissue. Understanding the pathology and cytogenetics of these tumors has increased significantly in the last 25 years. Based on the molecular cytogenetic analysis, both ES and PNET types are known to have translocation abnormalities on the same chromosome, most commonly occurring on chromosomes 11 and 22. Advances in the immunohistochemical examination have helped in further differentiating between PNET and ES from small round tumor types and other undifferentiated tumors types. PNET often shows aggressive clinical behavior, with worse effects than other small round cell tumors.[3]

Primitive neuroectodermal tumors morphologically show a high degree of diversity in clinical manifesttations and similarity in histopathological features with other small round cell tumors. Batsakis (1996) divides PNET into three groups based on the origin of soft tissue, among others; primitive neuroectodermal tumors of the nervous system, i.e. tumors originating from the central nervous system. Neuroblastoma, a tumor originating from the autonomic nervous system and peripheral primitive neuroectodermal tumors, i.e. tumors originating from tissue outside the central and autonomic nervous system.[1,3,16]

Peripheral primitive neuroectodermal tumors (PNETp) which will be the focus of the discussion in this case

report, namely the case of PNETp maxillary sinistra region in girls aged 17 years. The incidence of this case is very rare, therefore for diagnosis and treatment it remains a challenge. That is why we present this topic, which focuses on case reports, specifically about the role of radiotherapy in this case. region in girls aged 17 years. The incidence of this case is very rare, therefore for diagnosis and treatment it remains a challenge. That is why we present this topic, which focuses on case reports, specifically about the role of radiotherapy in this case.

EPIDEMIOLOGY

The incidence of PNET is often not reported in the literature, because it is still difficult to distinguish from other types of small round cell tumors. However, from the current diagnostic progress, these tumors can be distinguished from small round cell tumors and other undifferentiated tumors. Although PNET is very rare and the annual incidence of this tumor is not greater than the type of Ewing tumor, which is only 1–4% of all cases of malignancy in soft tissue.[4,16] These tumors can occur at newborn age until the age of 20 years and peak at ages 10 to 15 years. The incidence is 2.9 out of one million inhabitants. From some of the literature reports PNET usually occurs in the teens, with a ratio of incidence between men and women 1.5: 1. From the data recorded, this tumor rarely occurs in children of Asian descent and American children of African descent, with the majority of cases worldwide most common in white children and adolescents.[3]

The PNET incidence rate in the sinonasal region that has been reported is around 4-17% of all cases of soft tissue tumors in children. Two PNET cases were reported by the International Society of Pediatric Oncology in 2002. Nine PNET cases in neonates were reported by Sabire et al in 2003.[4]

Most PNET manifests in the thoracopulmonary region (Askin tumor), pelvis, abdomen and extremities. In the 26 cases reported by Jones and McGill, 11 of the 26 patients with PNET cases were manifested in the head and neck region.[2] From other published case reports, there were still rare cases of PNET in the head and neck area. About 20% of cases involving the head and neck, the location of the affected area is very diverse and including the paranasal sinus area, jugular foramen, oral cavity, nasal cavity, neck, skull, lingual nerve, parotid gland, larynx, retropharyngeal space, maxilla, mandible, masseter muscle, temporal area, pterygomaxillary space, esophagus and orbit.[3,5]

CASE ILLUSTRATION

A 17-year-old girl patient, came to the Radiotherapy clinic at dr. Cipto Mangunkusumo National General Hospital (RSCM) on September 21, 2016, was referred from the RSCM tumor surgery department with complaints of a lump in the left maxillary area, which was originally as big as a peanut but was felt getting bigger and bigger since 10 months ago. Another complaint is the nasal congestion and nasal discharge, which has been felt for 3 months before entering the hospital. No previous headaches or nosebleeds. The patient brought the results of laboratory blood tests that were within normal ranges, radiological X- Ray examination of the thoracic within normal ranges, and CT scan of sinonasal on August 11, 2016, showing a solid mass filling the left sinonasal with an expansion of the left orbital cavity and left-sided oral cavity (figure 1). The biopsy results were said to be PNET, with differential diagnosis is neuroendocrine carcinoma and the results of the immunohistochemical examination were positive strong membrane expression of CD99 markers, positive synaptophysin, positive neuron-specific enolase (NSE), nonspecific epithelial markers (AE1 / AE3), positive S100, which in conclusion supported a PNET. The results of postoperative anatomic pathology examination subtotal sinistra maxillectomy are aligned with histologically of PNET (pT3), with the upper incision limit of the maxillary sinus containing tumor mass and found lymphovascular invasion.

Subtotal sinistra maxillectomy on August 30, 2016, and consigned to Radiotherapy for adjuvant postoperative radiation on September 21, 2016. External radiation was performed on the local tumor field with IMRT technique with a total dose of 66 Gy in 33 fractions, starting from November 3 to December 19, 2016. Currently, the patient controls routinely every month in the oncology department with the condition still attached NGT and left maxillary prosthesis without complaints that interfere with activity. Patients were continued for chemotherapy in pediatric medical oncology division as further therapy, and given vincristine (Completed in July 2017). And resumed maxillary reconstruction surgery in August 2017, and had a Percutaneous Endoscopic Gastrostomy tube (PEG) attached due to dyphagia, and was removed in March 2018. Currently, patients are controlled every 3 months in tumor surgery.

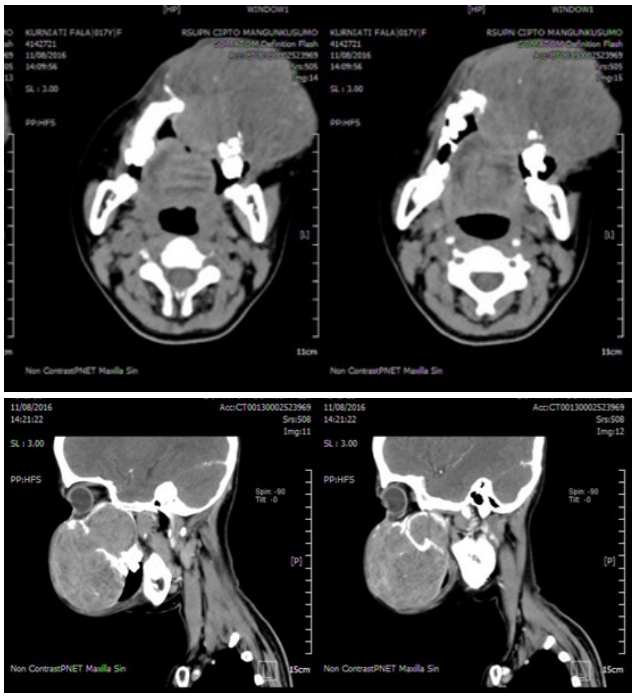

Figure 1. Axial and sagital CT Scans of RSCM patient, Ms. KF / 17 years old.

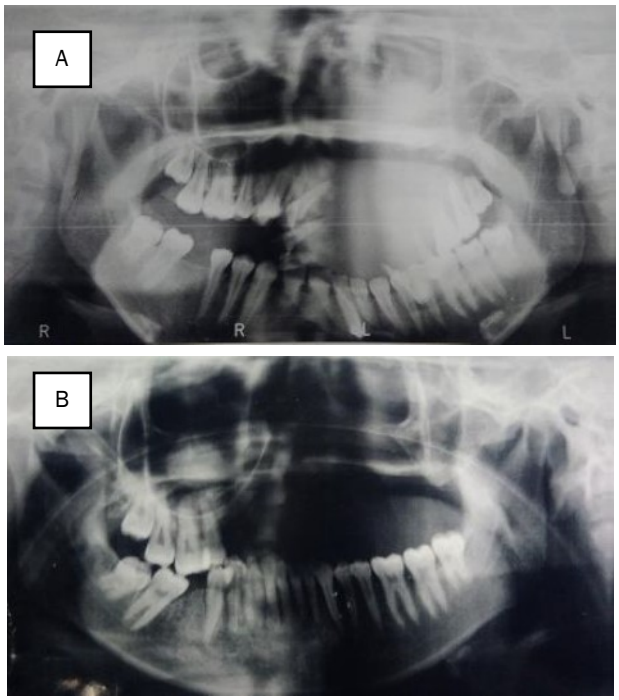

Figure 2. Panoramic photo of RSCM patient, Ms. KF / 17 years Old. On preoperative date 8/22/2016 (A) and one-year postoperative date 3/8/2017 (B).

DISCUSSION

PNET cases have been reported in children aged 17 years. Following the Lester report cited by Stafford, PNET generally occurs in children and young adults, can occur from newborns to the age of 20 years and most between the ages of 10 to 15 years. The clinical symptoms of PNET depended on the location of the affected organ but often caused pain and swelling in the structure in the area around the tumor. Other symptoms and signs that are often reported are specific to the location, in this case the clinical symptoms show a lump in the left maxillary area, nasal congestion and nasal discharge. According to the literature, tumors in the pediatric group are mostly occurred without typical symptoms and signs, which complicates the diagnosis and management. Furthermore, another literature supports that non-typical polymorphisms and clinical features are the main characteristics of this type of tumor, and the symptoms can differ according to tumor location.[4,7,8]

Physical examination revealed a polypoid shaped mass in the left maxillary sinus extending to the left maxillary region with a size of 5.5x8x7.5 cm, literature also informs that these tumors are usually polypoid, and can be quite large that can reach to 6 cm or more.[6] Computer tomography examination showed the presence of a left sinonasal malignant mass extending to the left protruding exophytic protrusion reaching the subcutis and cutis, accompanied by destruction of the sinus wall and left maxillary bone, no visible enlargement of the lymph node. PNET is a heterogeneous mass as in other small round cell neoplasms.

The result of Pathology Anatomy examination, based on morphological, shows that tumors have monotonous cells with a size almost as large as mature lymphocytes, pale cytoplasm and a few, hyperchromatin rounded nucleus, clear nucleolus and nucleus, possibly a picture of a small round cell tumor. From the immunohistochemical examination, it shows a positive membranous CD99 marker, positive synaptophysin, positive NSE, non-specific epithelial (AE1 / AE3) marker, S100 positive, concluded the PNET. This is consistent with the literature stating the Ewing's sarcoma or PNET is supported by strong immunoreactivity against CD99. Other tumor markers that have been detected by immunohistochemistry in this tumor type show a differentiation of neuroepithelial lines that include neuron-specific enolase (NSE), synaptophysin, S100 proteins, secretogranin II, vimentin and keratin.[9]

Management of this patient is surgical excision of the tumor and hemimaxillectomy. When mass surgery can be completely extrapolated, an anatomic pathology examination shows a picture of PNET with a left upper maxillary sinus incision boundary containing tumor mass and lymphovascular invasion, this patient is then

consulted to the Radiotherapy and Pediatric Medical Oncology Department to be advised to undergo Radiation and continued with chemotherapy. As performed by the patient at this time, resection and subtotal maxillectomy sinistra, followed by external radiation IMRT with a total dose of 66 Gy in 33 fractions. After the radiation, the patient continued chemotherapy with vincristine and adriamycin until July 2017 and performed palate reconstruction surgery again in August 2017. Until now, the patient controls routinely to the surgical department. The patient condition generally good without significant complaints until 2018. Based on the literature, stating that therapy requires resection with a combination of radiotherapy and chemotherapy to treat tumors that may remain or the presence of undetectable metastases.[10,11]

CONCLUSION

PNET does not have a definite protocol therapy, so it still refers to the management of Ewing's sarcoma, namely the act of resection with a combination of radiotherapy and or chemotherapy to treat possible tumors remaining or the presence of undetectable metastases.

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