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# Computed tomography and magnetic resonance imaging findings of peripheral primitive neuroectodermal tumors of the head and neck

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#### ABSTRACT

*Purpose:* We aimed to analyze the computed tomography (CT) and magnetic resonance imaging (MRI) findings of peripheral primitive neuroectodermal tumor (pPNET) of the head and neck. *Methods:* Eight patients with pPNET of the head and neck confirmed by histopathological examination were analyzed retrospectively.

Results: The average patient age was 8 years. The tumor location in the 8 patients was as follows: maxillofacial region (right, 2; left, 1), left maxillary sinus (1), right masticator space (1), left carotid space (1), right infratemporal fossa (1), and left parotid gland (1). All 5 patients who underwent CT demonstrated ill-defined soft masses and no calcification. Three patients with tumors in the maxillofacial region showed homogeneous small masses and a mild enhancement. The patient with left maxillary sinus tumor showed a heterogeneous mass with patchy, necrotic foci and mild heterogeneous enhancement. The patient with right masticator space tumor showed a heterogeneous mass, and marked heterogeneous enhancement. The T1-weighted images of the patients with right infratemporal fossa, left carotid space, and left parotid gland tumors were isointense. The T2-weighted images were heterogeneous and mildly hyperintense in 2 patients and hyperintense in 1 patient. Heterogeneous intermediate enhancement was demonstrated in 2 patients and mild ring enhancement in 1 patient.

Conclusion: The imaging features of pPNET of the head and neck are non-specific. An ill-defined, aggressive mass and variable enhancement on CT and MR images may suggest the diagnosis of pPNET. Peripheral PNET should be included in the differential diagnosis of children and adolescents' regional tumors.

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#### 1. Introduction

Primitive neuroectodermal tumor (PNET) is a kind of small round cell tumor with neuroectodermal origin [1]. This highly malignant tumor can further be divided into central PNET (cPNET) and peripheral PNET (pPNET). Peripheral PNET occurs outside the central and sympathetic nervous system and expresses high amounts of the CD99 and exhibits high characteristic chromosomal translocation t(11;22)(q24;q12)[1-4]. Peripheral PNET often occurs in the thoracopulmonary region (Askin tumor), retroperitonium, and extremities and rarely in the head and neck [5]. In recent years, there were a few radiological reports on pPNET arising in the thoracopulmonary region and abdomen, and few case reports focused on peripheral PNET of the head and neck [6–20]. Here, we conducted a retrospective radiological review of 8 patients with pPNET of the head and neck.

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#### 2. Materials and methods

We searched the database of our hospital and identified patients who were treated at our hospital between January 2000 and December 2008. In all, 12 patients with pPNET located in the head and neck were found. Eight patients with complete clinical and imaging data were included in this study, the other 4 patients without complete imaging data were ruled out. An institutional review board exemption and a waiver for the requirement of the written informed consent were obtained to perform this retrospective study.

Of the 8 patients, 5 had been examined with computed tomography (CT) and 3 had undergone magnetic resonance imaging (MRI). CT imaging was performed using a Twin Flash (Elscint, Haifa, Israel) or a Brilliance  $TM^{16}$  (Philips Medical Systems, Best, The Netherlands) helical scanner. The scan parameters were as follows: 5 mm slice thickness reconstructions, 23 cm field of view, 120 kV voltage, 200–300 mA current, and  $256 \times 256$  matrix. An intravenous bolus dose of 100 mL of a non-ionic iodinated contrast agent (iopromide; Ultravist; Schering) was administered at a rate of 2.5 mL/s to the patients who underwent contrast-enhanced CT.

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**Fig. 1.** In patient 1, the right maxillofacial region tumor shows homogeneous enhancement with absorption of the anterior wall of the right maxillary sinus.

Contrast-enhanced CT scans were started 50–60 s after the administration of the contrast agent.

MRI was performed using a 1.5 T MRI unit (Gyroscan Intera; Philips Medical Systems, Best, The Netherlands) and a head and neck coil (Philips Medical Systems, Best, The Netherlands). The MRI protocol included unenhanced axial and coronal T1-weighted sequences, axial and sagittal T2-weighted sequences, and contrast-enhanced axial, sagittal, and coronal T1-weighted sequences. The parameters for these sequences were as follows: T1-weighted spin echo (SE) sequence (TR/TE, 530/15 ms; slice thickness, 5.0 mm; field of view, 380–520 mm; matrix scan, 256 × 256) and T2-weighted turbo-spin echo sequence (TR/TE, 4800/120 ms; slice thickness, 5.0 mm; field of view, 300–380 mm; matrix scan, 256 × 256). An intravenous dose of 0.1–0.2 mmol/kg of contrast agent (Gadolinium-DTPA; Magnevist; Schering) had been administered to the patients undergoing contrast-enhanced MRI.

#### 3. Results

#### 3.1. Clinical data

The study group consisted of 4 men and 4 women with a mean age of 8 years (range 3–13 years). Of the 8 patients, the tumors were located in maxillofacial region (2 in the right and 1 in the left), left maxillary sinus (1), right masticator space (1 patient), right infratemporal fossa (1), left carotid space (1), and left parotid gland (1). Of the 8 patients, 6 presented with maxillofacial swelling, progressive growing masses and pains, 1 presented with a progressive growing mass in the left upper neck, and 1 presented with progressive growing tubercle in the left parotid gland.

#### 3.2. CT findings

The radiological findings from the eight cases are summarized in Table 1. The unenhanced CT images showed 3 small homogenous masses in the patients with maxillofacial region tumors (Fig. 1), a heterogeneous mass with patchy, necrotic foci in the patient with left maxillary sinus tumor, and a bulky heterogeneous mass in the patient with right masticator space tumor (Fig. 2).

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Radiological findings and prognoses of 8 patients with head and neck pPNET.

Case no./age (years)/sex	Site	Size	Margin	Density/intense	Enhancement	Prognosis
1/13/M	Right maxillofacial region	4.8 × 2.0cm	III-defined	Homogeneous	Mild homogeneous	Remission
2/3/F	Left maxillofacial region	$2.0\times1.5cm$	III-defined	Homogeneous	Mild homogeneous	Remission
3/5/F	Right maxillofacial region	$3.2\times1.8cm$	III-defined	Homogeneous	Mild homogeneous	Remission
4/7/F	Left maxillary sinus	$2.5\times1.8cm$	III-defined	Heterogeneous	Mild heterogeneous	
5/11/M	Right masticator space	$9.0\times7.6cm$	III-defined	Heterogeneous	Marked heterogeneous	Died
6/13/M	Right infratemporal fossa	$5.6\times7.8cm$	III-defined	Isointense (T1WI), heterogeneous mild hyperintensity (T2WI)	Intermediate heterogeneous	Died
7/13/M	Left carotid space	$2.7\times1.5cm$	Well -defined	Isointense (T1WI), heterogeneous mild hyperintensity (T2WI)	Mild ring	Remission
8/3/F	Left parotid gland	$4.5 \times 4.0$ cm	Well-defined	most isointense(T1WI), heterogeneous mixed hyperintensity (T2WI)	Intermediate heterogeneous	Remission

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