

Unusual Presentation of Peripheral Primitive Neuroectodermal Tumor of the Maxilla

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Abstract: The peripheral primitive neuroectodermal tumor (pPNET) is a rare and highly malignant soft tissue neoplasm in children and young adults. Fewer than 10 reported cases of pPNET of maxilla are available in the English literature. A 28-year-old woman was presented with the pPNET of the maxilla and metastasis. Two years after diagnosis, she experienced diplopia, and then magnetic resonance imaging was done, which showed a mass in the optic chiasma and parasellar region. The typical appearance resembled large noncalcified soft tissue masses in the magnetic resonance image and computed tomographic scan of the maxilla. Diagnosis was established by immunohistochemical features. She was treated with surgery, chemotherapy, radiation therapy, and gamma knife. She was under close observation since then (approximately 8 mo), and there has been no recurrence of tumor up to now.

Key Words: pPNET, maxilla, head and neck, diplopia

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Primitive neuroectodermal tumor (PNET) comprises small round cells and develops mainly in the central nervous system and soft tissue. These tumors are generally manifest in infancy or early childhood.^{1–3} Primitive neuroectodermal tumor outside the central nervous system is called peripheral primitive neuroectodermal tumor (pPNET) developing from migrating embryonal cells of the neural crest.² Although pPNET is exceedingly rare, its presence in the chest wall, abdomen, extremities, posterior mediastinum, myocardium, kidney, vagina, bladder, parotid, and even in the orbit has been reported.^{1–4} Fewer than 10 reported cases of pPNET of maxilla are available in the English literature.² The typical appearance resembled large noncalcified soft tissue masses with cystic or necrotic. Magnetic resonance imaging (MRI) and computed tomography (CT) are useful in predicting resectability, in detecting distant metastases, and in evaluating

response to treatment.⁵ Diagnosis can be established by immunohistochemical (IHC) and electromicroscopic features that demonstrate various degrees of neural differentiation.^{6,7} However, treatment of PNET in extracranial sites in adults is not clearly defined in the literature. Surgery is performed in all cases, either for definitive diagnosis or for therapy.^{4,7,8} The following article reports an unusual case of PNET in a young woman's maxilla because of its location, clinical presentation, and metastasis to parasellar region.

CLINICAL REPORT

In this study, we present a case of pPNET of the maxilla with metastasis in a 28-year-old woman. She was referred to us on May 2006 when she already had the left maxillary tumor enlarging since 1 year previously (Fig. 1). She had no noteworthy medical family history or past history in this case. Results of her examinations showed a firm nontender fixed mass (3 cm × 2 cm) on the left maxilla extending to the alveolar process and gingiva. The neck was clinically uninvolved, and chest radiograph and abdominal ultrasound were also normal. Before her referral, she had had 2 times shaving biopsy from the left maxilla, and her pathologic report showed a malignant, small, round, cell tumor suggesting a mesenchymal chondrosarcoma. According to this pathologic report, the left hemimaxillectomy was done on August 2006 (Fig. 2). After that, the patient was followed up with serial clinical examinations and CT scan every 6 months. The examinations continued without any sign of recurrence until July 2008 that she presented diplopia. Magnetic resonance imaging was performed, which showed a mass in the optic chiasma and parasellar region (Figs. 3 and 4). Then, neurosurgical consultation was done, and gamma knife (radiation) was performed. On September 2008, her diplopia improved. On November 2008, a CT scan showed small masses on the site of the maxillectomy and right maxilla. Afterward in December 2008, excision of tumors at the site of the previous maxillectomy (maxillofrontal process) and right maxilla was done. Her pathologic report indicated lymphoma or undifferentiated malignant tumor. Afterward, the pathologists suggested IHC for a definite diagnosis. According to IHC, CK and LCA were negative. Vimentin was moderately positive, S-100 was scattered in the cell, and Mic-2 (CD99) was positive. On the basis of these findings, the lesion was diagnosed as a pPNET of the maxilla. After that, in March 2009, radiotherapy (80 g) after 3 courses of chemotherapy (vincristine, actinomycin D, and ifosfamide) was administered. It was in this phase that she experienced severe leucopenia, and hence, she refused to continue this treatment. She was under close observation since then (approximately 6 mo), and there has been no recurrence of tumor up to now.

DISCUSSION

Peripheral primitive neuroectodermal tumors are uncommon tumors that are mainly seen in the trunks and extremities of children

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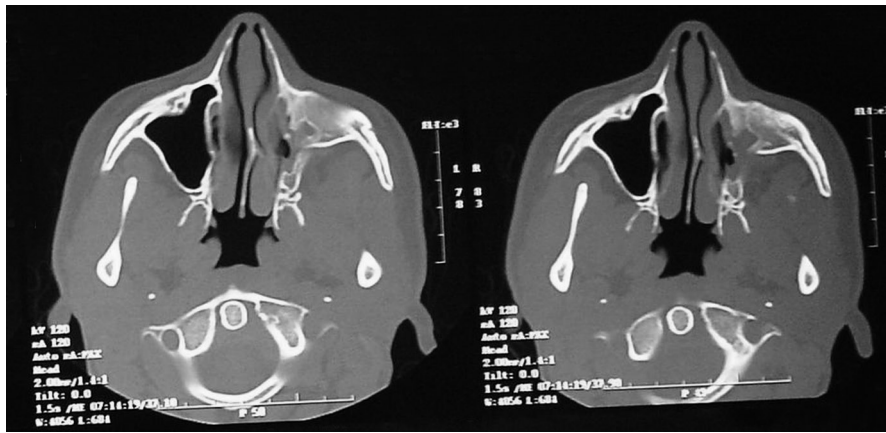


FIGURE 1. A large tumor in the left maxilla.

and young adults. Occasional clinical reports have described pPNET at unusual locations like the head and neck region and retroperitoneum. Ewing tumor families of pPNET are primitive, small, round cell tumors (SRCTs) of the bone and soft tissue. Peripheral primitive neuroectodermal tumors of the maxilla are extremely rare disease entities.⁹

Pathologically, PNETs are believed to represent a transition between neoplastic Schwann cells, neuroblasts, and perhaps paraganglionic elements.^{10,11} It is important that this diagnosis be considered in adults who present it with bone and soft tissue SRCTs. Also, appropriate biopsy specimens should be collected for molecular analysis at the time of diagnosis. However, pPNETs only rarely enter the differential diagnosis of bone and soft tissue SRCTs in adults. The differential diagnosis of small round cell tumors in the head and neck includes malignant lymphoma, leukemia, neuroblastoma, leiomyosarcoma, rhabdomyosarcoma, undifferentiated carcinoma, and pPNET-Ewing sarcoma.^{7,12,13} The final diagnosis of pPNET in the present case was based on immunohistochemistry.

Radiologic workup included standard radiographs, CT, MRI, and bone scintigraphy. The radiographic appearance of these tumors was not specific for differentiation of pPNETs from other types of bone and soft tissue tumors.⁵ The typical appearance resembled large, noncalcified, soft tissue masses with cystic or necrotic areas. Heterogeneous enhancement with intravenous contrast agents was evident on CT scan, as was an intermediate signal intensity on T1-weighted

images and hyperintense signal on T2-weighted images.^{5,7,8} Imaging in cases of pPNET maxilla have shown local bone destruction with invasion of the surrounding walls.^{5,7,9,12} Magnetic resonance imaging and CT were useful in predicting resectability, in detecting distant metastases, and in evaluating response to treatment. Peripheral primitive neuroectodermal tumors progress rapidly and have often metastasized at the time of diagnosis. Incidence of distant metastases to lung, liver, bone, and lymph node may be high.^{1,8,12} There was an optic chiasma and parasellar region metastasis in our patient, which presented itself as diplopia.

Because of the rare occurrence of pPNET, optimal therapy is challenging, particularly if they occur in the head and neck. In many studies of such patients, aggressive local treatment in terms of surgery followed by adjuvant radiotherapy to a dose of 45 to 70 Gy and multiagent chemotherapy has been described.^{1,3,8,9,13,14} However, Mohindra et al reported that an 8-year-old boy was treated with multiagent combination chemotherapy followed by definitive radiation. So pPNET treatment is similar to Ewing sarcoma.¹³ We believe that conducting surgery followed by chemotherapy and radiation therapy has better survival and aesthetic result in the maxilla. Close cooperation between surgeons and their oncologist and radiotherapist

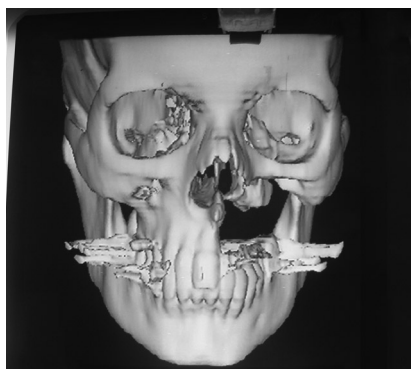


FIGURE 2. Left hemimaxillectomy was done and shown three-dimensionally.



FIGURE 3. Magnetic resonance image showing a mass in the optic chiasma and parasellar region.



FIGURE 4. Magnetic resonance image showing a mass in the optic chiasma and parasellar region.

colleagues is obligatory when treating pPNET. A close follow-up with regular radiographic examination for at least 5 years is mandatory.

CONCLUSIONS

Maxilla as a site of origin of pPNET is rare.⁹ Hence, the differential diagnosis of pPNET is very important. Peripheral primitive neuroectodermal tumor often metastasizes at the time of diagnosis. A combination of organ preserving surgery, chemotherapy, and adjuvant radiation therapy has been the recommended treatment of choice.

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