A case of primitive neuroectodermal tumor in the distal phalanx

El distal falanksında primitif nöroektodermal tümör: Olgu sunumu

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Primitive neuroectodermal tumor (PNET) is one type of malignant small-cell tumor. Other diseases of this family include neuroblastoma, Ewing’s sarcoma, embryonic rhabdomyosarcoma, and lymphoma. These tumors are generally distinguished with immunohistochemical findings.1

Primitive neuroectodermal tumor generally involves the central nervous system, kidney, pelvis, and the chest wall.2 In this paper, we report a case of PNET of the middle finger. To our knowledge, only four cases of typical PNET have been reported, involving the hand.2,3

CASE REPORT
A 13-year-old girl presented with pain, tenderness, and swelling of gradual onset at the distal phalanx of her right middle finger. On physical examination, there was a tender, swollen, erythematous nodular mass, 2x1 cm in size, at the distal volar tip of the middle finger. On physical examination, there was a tender, swollen, erythematous nodular mass, 2x1 cm in size, at the distal volar tip of the middle finger. A radiogram of the finger demonstrated a soft tissue swelling over the phalanx. Magnetic resonance imaging revealed an extra-osseous soft tissue mass without infiltration to bone. Bone scintigraphy showed increased uptake only in the distal phalanx. Histological examination of an incisional biopsy showed cellular tumoral tissue. The lesion was diagnosed as primitive neuroectodermal tumor based on immunohistochemical studies. Surgical treatment was performed with marginal resection of the tumor site and preservation of the digital artery and nerve. Two weeks after surgery, combined chemotherapy was administered at eight cycles over a time period of six months. Forty-two months after surgery, the patient was free of tumor. She had normal function of the hand with full strength.

Key words: Bone neoplasms/surgery; hand/pathology; neuroectodermal tumors, primitive/surgery.

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A soft tissue swelling over the phalanx with no change in bone (Fig. 1a). Magnetic resonance imaging revealed an extra-osseous soft tissue mass without infiltration to bone on both T1- and T2-weighted images (Fig. 1b). Her white blood count (4,500/mm³) and erythrocyte sedimentation rate (6 mm/hr) were within normal limits. A chest radiograph and computed tomography scan showed no metastatic lesions. A ⁹⁹Tc-methylene diphosphonate bone scan showed increased uptake only in the distal phalanx. An incisional biopsy was obtained through a longitudinal incision over the lesion. Histological examination showed cellular tumoral tissue of lobulated pattern separated by hyalinized fibrous septas.

Tumoral cells showed an alveolar and solid pattern (Fig. 2a). Cells were oval or round. Some were eccentrically located, with a vesicular nucleus and narrow eosinophilic cytoplasm (Fig. 2b). Immunohistochemical studies revealed positive results for vimentin, CD99, chromogranin, and negative results for para-aminosalicylic acid, cytokeratin, desmin, synaptophysin, NCAM, epithelial membrane antigen, myoglobin, leukocyte common antigen, S-100 protein, and CD3-20-34-68. The tumor was diagnosed as PNET.

Surgical treatment comprised of marginal resection of the tumor site with preservation of the digital artery and nerve. Two weeks after surgery, multiagent chemotherapy (vincristin, dactinomycin, and cyclophosphamide) was administered at eight cycles over a time period of six months.

Forty-two months after surgery, the patient was free of tumor clinically. There was no recurrence. Magnetic resonance imaging of the hand and bone scintigraphy were normal. She had normal function of the hand with full strength.

**DISCUSSION**

Malignant small-cell tumors represent 6% to 10% of all malignant bone tumors, ranking in the sixth place.[6,7] They are usually seen between the ages of 10 to 15 years and rarely seen over the age of 30 years.[8] Ewing’s sarcoma usually involves the metaphyses or epiphyses. Primitive neuroectodermal tumors predominantly involve the diaphyses of the long bones together with central and axial skeleton.[8,10] Histological findings lack specificity; thus, differ-

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**Fig. 1. (a)** A preoperative radiogram of the patient showing a soft tissue swelling (arrow); (b) a magnetic resonance scan of the finger showing the extra-osseous tumor.
ential diagnosis is made by immunohistochemical studies. However, immunohistochemical staining required for diagnosis are often not definitive.[14]

To our knowledge, only four cases of PNET have been reported, involving the hand.[2-5] A consensus on the treatment of PNET has not been established. A review of the literature yields different treatment strategies. Terrier et al.[12] treated 47 cases of PNET of bone with a combination of radiotherapy and chemotherapy. Jurgens[13] reported on patients treated by chemotherapy followed by surgery and/or radiotherapy, whose mean survival rate was 66% over a two-year follow-up period. Schmidt et al.[10] treated 24 patients with chemotherapy and/or radiotherapy depending on the localization and reported a survival rate of 37.5% at the end of a seven-year follow-up. Bacci et al.[14] reported the five-year survival rate as 54% in a series of 44 cases treated with neoadjuvant chemotherapy followed by surgery, or surgery and radiotherapy, or radiotherapy alone. Cebrian et al.[1] treated a 41-year-old male patient with PNET of the ankle with wide local excision followed by chemotherapy and external radiotherapy at 5,000 cGy. Although PNET seems to be radiosensitive, radiotherapy is not the preferred method of treatment.[2] Based on the findings of 26 patients with PNET, Marina et al.[15] advocated aggressive surgery as the first line treatment and recommended radiotherapy only in the presence of microscopic disease. According to Kushner et al.[16] radiotherapy should only be used to prevent recurrences of poor-risk axial PNET. The authors recommended a treatment regimen composed of initial surgical excision followed by adjuvant chemotherapy, and radiotherapy only to ablate the residual disease. Finally, based on the evidence obtained from a review of the literature, Harder et al.[2] concluded that wide local excision followed by chemotherapy constituted the mainstay of treatment.

Since congenital PNET is more aggressive, its treatment is usually unsuccessful and the use of chemotherapy is limited due to increased toxicity in neonates.[4,6] We used combined chemotherapy (vincristin, dactinomycin, and cyclophosphamide) in our patient after the excision of the tumor. In contrast to previous reports, we did not perform a wide local excision, or radiotherapy, and did not amputate the distal part of the affected finger. Despite this limited surgery, the patient remained disease-free during 42 months after surgery.

In conclusion, PNET of the hand is very rare, highly aggressive with a high recurrence rate, and more reports are necessary to establish a treatment strategy against this tumor.

REFERENCES


Fig. 2. (a) Tumoral tissue arranged in a lobulated pattern separated by fibrous septas (H-E x 10). (b) Oval or round tumor cells with vesicular nuclei and narrow eosinophilic cytoplasm, some having an eccentric location (H-E x 40).
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