Eklem Hastalıkları ve Cerrahisi
Joint Diseases and Related Surgery

Case Report / *Olgu Sunumu*

Joint Dis Rel Surg
2007;18(3):130-133

A case of primitive neuroectodermal tumor in the distal phalanx

El distal falanksında primitif nöroektodermal tümör: Olgu sunumu

Onur Hapa, M.D., Halil Yalçın Yüksel, M.D., Hasan Hilmi Muratlı, M.D.,
Levent Çelebi, M.D., Ali Biçimoğlu, M.D.

Department of Orthopedics and Traumatology, Ankara Numune Education and Research Hospital

Primitive neuroectodermal tumor of the hand is extremely rare. To our knowledge, only four cases have been reported. A 13-year-old girl presented with pain, tenderness, and swelling 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. 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.

Elde primitif nöroektodermal tümör gelişimi son derece nadir bir durumdur; bugüne kadar dört olgu bildirilmiştir. On üç yaşında bir kız, sağ el orta parmak distal falanksında ağrı, hassasiyet ve şişlik yakınmalarıyla başvurdu. Fizik muayenede, orta parmağın distal volar ucunda, 2x1 cm büyüklükte, hassas, eritematöz nodüler bir kitle görüldü. Parmak röntgeninde falanksta yumuşak doku şişliği izlendi; manyetik rezonans görüntülemede, kemiğe infiltrasyon göstermeyen, ekstraosseöz yumuşak doku kitlesi gözlendi. Kemik sintigrafisinde, sadece distal falanksta olmak üzere artmış tutulum vardı. Yapılan insizyonel biyopsinin histopatolojik inceleme sonucu tümoral doku olarak bildirildi. İmmünhistokimyasal çalışmalar sonucunda lezyona primitif nöroektodermal tümör tanısı kondu. Tümör, dijital arter ve sinir korunarak marjinal rezeksiyonla çıkarıldı. Cerrahiden iki hafta sonra hastaya, altı ayda sekiz kür şeklinde olmak üzere kombine kemoterapi uygulandı. Kırk iki aylık takip süresi boyunca hastada nüks gözlenmedi; el fonksiyonu normal ve tam güçte idi.

Anahtar sözcükler: Kemik neoplazileri/cerrahi; el/patoloji; nöroektodermal tümör, primitif/cerrahi.

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.[2-5]

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. There was no sign of lymphadenopathy. A radiogram of the finger demonstrated

• *Received: February 6, 2007 Accepted: July 12, 2007*

• Correspondence: Onur Hapa, MD. Çelikkent Sitesi A/2, No: 6 Çayyolu 06530 Ankara, Turkey.
Tel: +90 312 - 240 32 70 *Fax:* +90 312 - 314 30 60 *e-mail:* onurhapa@gmail.com

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 T_1- and T^2-weighted images (Fig. 1b). Her white blood count (4,500/mm^3) 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.[9,10] Histological findings lack specificity; thus, differ-

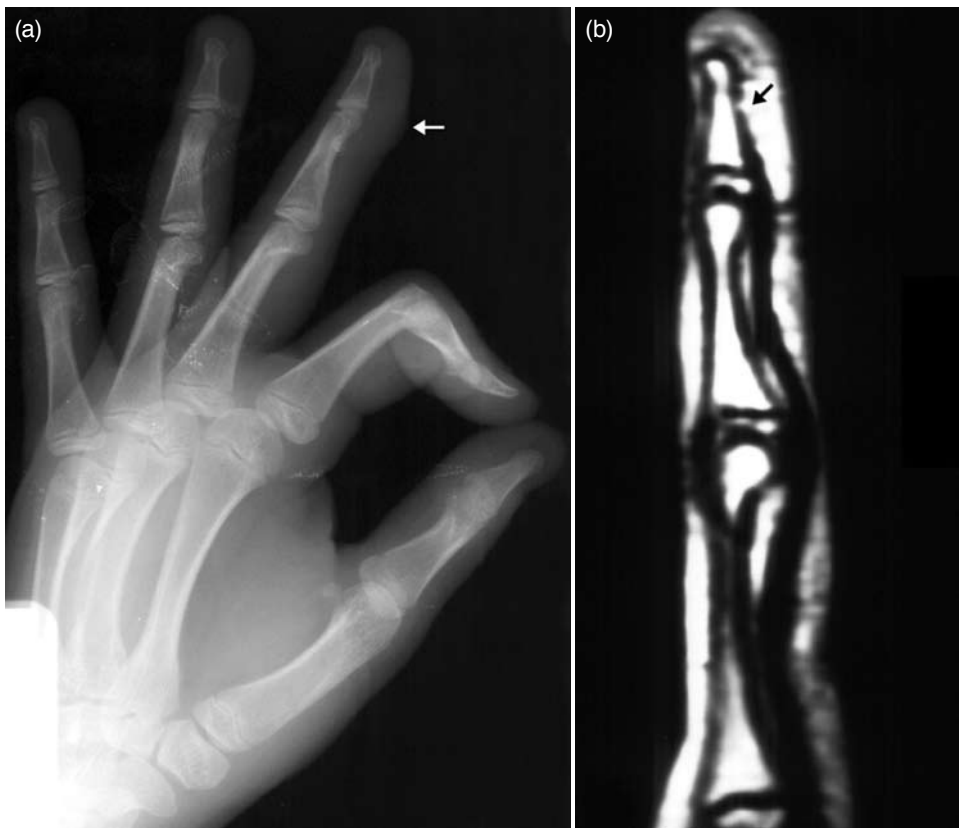

Fig. 1. (a) A preoperative radiogram of the patient showing a soft tissue swelling (arrow); **(b)** a magnetic resonance scan of the finger showing the extra-osseous tumor.

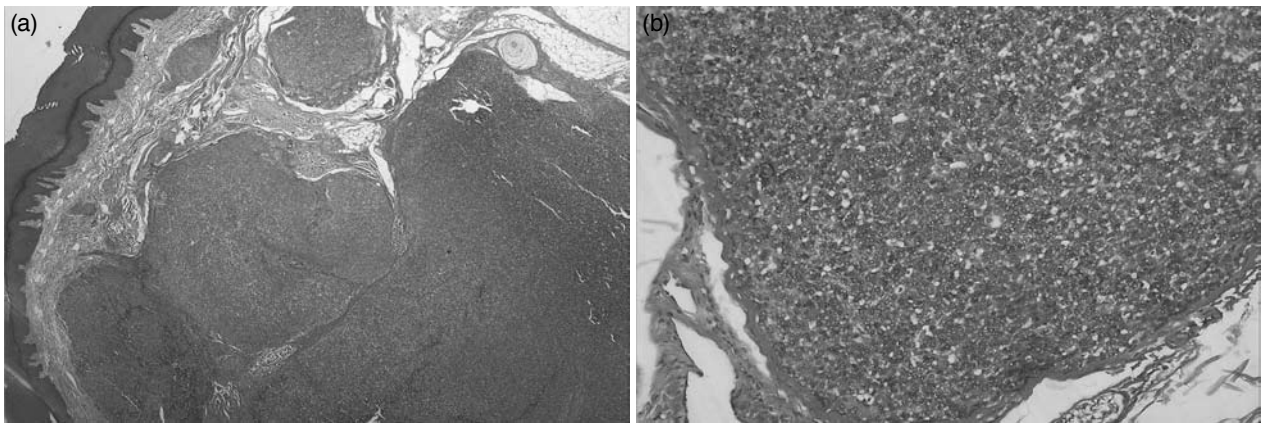

Fig. 2. (a) Tumoral tissue arranged in a lobulated pattern separated by fibrous septas (H-E x 10). **(b)** Ovale or round tumor cells with vesicular nuclei and narrow eosinophilic cytoplasm, some having an eccentric location (H-E x 40).

ential diagnosis is made by immunohistochemical studies. However, immunohistochemical staining required for diagnosis are often not definitive.[11]

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 congenial 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

1. Cebrian JL, Ibarzabal A, Garcia-Crespo R, Marco F, Ortega L, Lopez-Duran L. Peripheral primitive neuroectodermal tumor after radiotherapy. Clin Orthop Relat Res 2003;(413):255-60.
2. Harder Y, Buechler U, Vögelin E. Primitive neuroectodermal tumor of the thumb metacarpal bone: a case report and literature review. J Hand Surg [Am] 2003; 28:346-52.
3. Daw JL, Wiedrich TA, Bauer BS. Congenital primitive neuroectodermal tumor of the hand: a case report. J Hand Surg [Am] 1997;22:743-6.
4. El Hayek M, Trad O, Islam S. Congenital peripheral primitive neuroectodermal tumor refractory to treatment. J Pediatr Hematol Oncol 2004;26:770-2.

5. Erdmann D, Brown RE, Rumbolo PM. Congenital neuroepithelioma in an infant hand. J Hand Surg [Br] 1996;21:117-20.
6. Eggli KD, Quiogue T, Moser RP Jr. Ewing's sarcoma. Radiol Clin North Am 1993;31:325-37.
7. Roessner A, Mittler U, Rose I, Radig K, Grote H. Pathology of Ewing sarcoma. Pathologe 1996;17:6-17. [Abstract]
8. Price CH, Jeffree GM. Incidence of bone sarcoma in SW England, 1946-74, in relation to age, sex, tumor site and histology. Br J Cancer 1977;36:511-22.
9. Llombart-Bosch A, Contesso G, Peydro-Olaya A. Histology, immunohistochemistry, and electron microscopy of small round cell tumors of bone. Semin Diagn Pathol 1996;13:153-70.
10. Schmidt D, Herrmann C, Jurgens H, Harms D. Malignant peripheral neuroectodermal tumor and its necessary distinction from Ewing's sarcoma. A report from the Kiel Pediatric Tumor Registry. Cancer 1991;68:2251-9.
11. Malone M. Soft tissue tumors in childhood. Histopathology 1993;23:203-16.
12. Terrier P, Henry-Amar M, Triche TJ, Horowitz ME, Terrier-Lacombe MJ, Miser JS, et al. Is neuro-ectodermal differentiation of Ewing's sarcoma of bone associated with an unfavourable prognosis? Eur J Cancer 1995;31A:307-14.
13. Jurgens H. Interdisciplinary therapy of Ewing sarcoma. Schweiz Rundsch Med Prax 1995;84:1005-9. [Abstract]
14. Bacci G, Ferrari S, Bertoni F, Donati D, Bacchini P, Longhi A, et al. Neoadjuvant chemotherapy for peripheral malignant neuroectodermal tumor of bone: recent experience at the Istituto Rizzoli. J Clin Oncol 2000;18:885-92.
15. Marina NM, Etcubanas E, Parham DM, Bowman LC, Green A. Peripheral primitive neuroectodermal tumor (peripheral neuroepithelioma) in children. A review of the St. Jude experience and controversies in diagnosis and management. Cancer 1989;64:1952-60.
16. Kushner BH, Meyers PA, Gerald WL, Healey JH, La Quaglia MP, Boland P, et al. Very-high-dose short-term chemotherapy for poor-risk peripheral primitive neuroectodermal tumors, including Ewing's sarcoma, in children and young adults. J Clin Oncol 1995;13:2796-804.