Primitive neuroectodermal tumor of hand and forearm: A rare clinical entity

Raja Tiwari · Satya S. Tripathy · Ramesh K. Sharma

Published online: 23 June 2012
© American Association for Hand Surgery 2012

Introduction

Sarcomas of the small round cell type originate from neural ectodermal tissue. They are composed of small, blast-like round cells [19] and are classified into three subgroups: typical Ewing's sarcoma, atypical Ewing's sarcoma, and primitive neuroectodermal tumor (PNET). Peak incidence occurs in the second decade of life. An onset after the third decade of life is extremely rare. There is a slight male predominance [19].

The various sites of metastases include the lung (50 %), bone (25 %), and bone marrow (20 %). Metastases to other sites are rare. Radiotherapy and aggressive surgery independently or in combination have formed the hallmark of therapy in treating Ewing's sarcoma and related PNET.

With better prognosis, reconstructive surgery, and surgery that minimizes amputation have gained importance in the treatment protocol [7, 15].

Various reports and publications have dealt with PNET affecting mainly the central nervous system, kidney, pelvis, and chest wall. Only five reported cases of typical PNET have involved the hand [3, 5, 6, 16, 22].

Case Report

An otherwise healthy 36-year-old man presented with a history of recurrent pain and gradually increasing swelling of the thenar region of the right, dominant hand for the past 1 year. There had been no previous trauma. Clinical examination revealed a 7×5-cm nodular mass, over the proximal palm and occupying the thenar and hypothenar regions (Fig. 1). It was firm in consistency and slightly tender on touch. There was no sign of lymphadenopathy.

Fig. 1 Showing mass in the palm

Electronic supplementary material The online version of this article (doi:10.1007/s11552-012-9432-2) contains supplementary material, which is available to authorized users.

R. Tiwari · S. S. Tripathy · R. K. Sharma (✉)
Department of Plastic Surgery, Post Graduate Institute of Medical
Education and Research,
Sector 12,
Chandigarh 160012, India
e-mail: drsharmark@gmail.com

The magnetic resonance imaging (MRI) showed an extra-osseous soft tissue mass without infiltration to the bone, hyperintense on T2 and hypointense on T1-weighted images (Figs. 2 and 3). The lesion was confined to the subcutaneous plane; posteriorly it was abutting the flexor retinaculum and thenar and hypothenar muscles. However, there was no involvement of flexor tendons, nerves, or blood vessels in the region.

The white blood count (4,500/mm³) and erythrocyte sedimentation rate (6 mm/h) were within normal limits. A chest radiograph and computed tomography scan showed no metastatic lesions. A fine needle aspiration cytology was obtained, which showed high cellularity of small to large clumps of cells having plump oval monomorphic nuclei with scanty cytoplasm. These features are suggestive of low-grade malignant mesenchymal tumor.

Surgical treatment comprised of marginal resection of the tumor with enbloc resection of transverse carpal ligament, thenar, and hypothenar muscles (Fig. 4). The radial and ulnar arteries and median and ulnar nerves were preserved. The thumb was stabilized using a K-wire and skin cover was given using posterior interosseous artery flap (Fig. 5). A histopathology report confirmed the diagnosis of PNET. It showed cells having round to oval nuclei having fine nuclear chromatin, inconspicuous nucleoli with scanty pale cytoplasm, arranged in sheets and fascicles separated by capillaries (Fig. 6).

The lymphovascular emboli were seen on microscopic examination on immunohistochemistry, the tumor cells were positive for CD99, NSE, and S-100.

Three weeks after surgery, radiotherapy was administered, a total dose of 45 cGy over 8 weeks. There was no local recurrence over a period of 20 weeks postoperatively. He had complete wound healing with acceptable cosmesis of the hand (Fig. 7). There was weakness in opposition for which an opponensplasty was planned. Tc99 scan 3 months postoperatively documented no local or distant metastasis.

Discussion

Apart from leukemias and lymphomas, which are easily identified by immunohistochemical means, the term small round cell tumor includes neuroblastoma; Ewing's sarcoma; peripheral primitive neuroectodermal tumor (PNET); and rhabdomyosarcoma, which predominate in the pediatric age group; desmoplastic small round cell tumor; and some examples of poorly differentiated synovial sarcoma [8].

Malignant small-cell tumors represent 6 % to 10 % of all malignant bone tumors, ranking in the sixth place [4, 20]. They are usually seen between the ages of 10 and 15 years and rarely seen over the age of 30 years [18].

Fig. 2 MRI showing extraosseous soft tissue mass, hyperintense on T2 image

Fig. 3 MRI showing extraosseous soft tissue mass, hypointense on T1 image

Histological findings lack specificity; thus, differential diagnosis is made by immunohistochemical studies. However, immunohistochemical staining required for diagnosis are often not definitive [13].

Up to now, only five reported cases of typical PNET has involved the hand [4–6, 8, 16] in English literature. It is also well-known and has been previously published that patients

Fig. 4 Defect after excision

Fig. 5 Defect resurfaced with posterior interosseous artery flap

presenting with upper limb sarcomas require early diagnosis, careful preoperative assessment, and an appropriate operation [1, 9].

The clinical presentation of tumors of the ES family may be insidious, with pain and swelling as the most common complaints. MRI has become the standard in the diagnosis, primary local staging, and assessment of treatment response of these neoplasias [11, 17].

Although PNET seems to be radiosensitive, radiotherapy is not the preferred method of treatment [10]. Based on the findings of 26 patients with PNET, Marina et al. [14] advocated aggressive surgery as the first-line treatment and recommended radiotherapy only in the presence of

Fig. 6 Histopathological report

Fig. 7 Appearance at 20 weeks postoperative

microscopic disease to prevent recurrences of poor risk axial PNET.

Surgical techniques have changed substantially over time. Because of the improvements made in the diagnosis and the use of microsurgical techniques, the rate of amputations has decreased remarkably. Fortunately, limb salvage has not resulted in a worse prognosis in terms of survival [2].

In the hand, this frequently necessitates reconstruction of a significant soft tissue and/or bony defect. Although it is clearly the primary aim of the surgical team to completely resect the tumor and render the patient disease free, stable reconstruction to provide coverage, recover function, avoid immobilization, minimize the number of surgical procedures, and restore form are secondary but very significant concerns [12].

Various reconstructive alternatives are available, including primary closure; skin grafting; and local, regional, and free flaps. Talbot et al. gave an algorithm for choosing the appropriate site-specific coverage [21].

Sometimes excision of vital structures of hand results in functional deformities and procedures such as opponensplasty to be undertaken. According to the authors' recommendations, this should be done when the patient has completed the radiotherapy protocol and has been declared disease free.

Conclusion

PNET of the hand is a rare group of tumor of the hand, and more case reports are necessary to establish a protocol for early diagnosis and effective management.

Conflict of interest The authors declare that they have no conflicts of interest, commercial associations, or intent of financial gain regarding this research.

References

1. Brien EW, Terek RM, Geer RJ, Caldwell G, Brennan MF, Healey JH. Treatment of soft-tissue sarcomas of the hand. *J Bone Joint Surg (Am)*. 1995;77:564.
2. Daecke W, Ahrens S, et al. Ewing's sarcoma and primitive neuroectodermal tumor of hand and forearm. *J Cancer Res Clin Oncol*. 2005;131:219–25.
3. Daw JL, Wiedrich TA, Bauer BS. Congenital primitive neuroectodermal tumor of the hand: a case report. *J HandSurg*. 1997;22A:743–6.
4. Eggli KD, Quiogue T, Moser Jr RP. Ewing's sarcoma. *Radiol Clin North Am*. 1993;31:325–37.
5. El Hayek M, Trad O, Islam S. Congenital peripheral primitive neuroectodermal tumor refractory to treatment. *J Pediatr Hematol Oncol*. 2004;26:770–2.
6. Erdmann D, Brown RE, Rumbolo PM. Congenital neuroepithelioma in an infant hand. *J Hand Surg [Br]*. 1996;21:117–20.
7. Exner GU, von Hochstetter AR, Schreiber A, Sokolovic-Dalipagic E. Osteosarcoma and Ewing's sarcoma. Improved recovery chances in the past 25 years according to the experience of Balgrist Hospital. *Schweiz Rundsch Med*. 1996;85:706–13.
8. Fisher C (1999) Current aspects of the pathology of soft tissue sarcomas: seminars in radiation oncology. 9(4):315–327
9. Fleegler EJ. An approach to soft tissue sarcomas of the hand and upper limb. *J Hand Surg (Br)*. 1994;19:411.
10. 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.
11. Henk CB, Grampp S, Wiesbauer P, Zoubek A, Kainberger F, Breitenseher M, et al. Ewing sarcoma. Diagnostic imaging. *Radiologe*. 1998;38:509–22.
12. Him FP, Casanova R, Vasconez LO. Myocutaneous and fasciocutaneous flaps in the upper limb. *Hand Clin*. 1985;1:759.
13. Malone M. Soft tissue tumors in childhood. *Histopathol*. 1993;23:203–16.
14. 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.
15. McLean TW, Hertel C, Young ML, Marcus K, Schizer MA, Gebhardt M, et al. Late events in pediatric patients with Ewing sarcoma/primitive neuroectodermal tumor of bone: the Dana-Farber Cancer Institute/Children's Hospital experience. *J Pediatr Hematol Oncol*. 1999;21:486–93.
16. Onur Hapa MD, Halil Yalçın Yüksel MD, Hasan Hilmi Muratlı MD, Levent Çelebi MD, Ali Biçimoğlu MD. A case of primitive neuroectodermal tumor in the distal phalanx. *Joint Dis Rel Surg*. 2007;18(3):130–3.

17. Ozaki T, Hashizume H, Kawai A, Inoue H. Ewing's sarcoma of the hand: magnetic resonance images and treatment. *J Hand Surg.* 1995;20A:441–4.
18. 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.
19. Roessner A, Mittler U, Röse I, Radig K, Grote H. Pathology of Ewing sarcoma. *Pathologe.* 1996;17:6–17.
20. Roessner A, Mittler U, Rose I, Radig K, Grote H. Pathology of Ewing sarcoma. *Pathologe.* 1996;17:6–17.
21. Talbot SG, Mehrara BJ, Disa JJ, Wong AK, Pusic A, Cordeiro PG, et al. Soft-tissue coverage of the hand following sarcoma resection. *Plast Reconstr Surg.* 2008;121:534–43.
22. Yves Harder MD, Ueli Buechler MD, Esther Vögelin MD. Primitive neuroectodermal tumor of the thumb metacarpal bone: a case report and literature review. *J Hand Surg.* 2003;28A:346–52.