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CASE REPORT

Hand/Peripheral Nerve

Novel Treatment of a Malignant Peripheral Nerve Sheath Tumor of the Median Nerve

Eirini Liodaki, Msc
Sandra Robiller, MD
Eike Wenzel, MD
Peter Mailaender, Prof.
Felix Stang, MD

Summary: Malignant peripheral nerve sheath tumors are rare, associated with a poor prognosis and uncertainty regarding the appropriate management. We report a novel oncologic and reconstructive treatment of a young patient with a malignant peripheral nerve sheath tumor of the median nerve of the left hand. The patient underwent a wide local excision, an opponensplasty, a nerve reconstruction by nerve allografts followed by brachytherapy treatment. Two years later, the patient remains disease free with preserved function of her hand. *(Plast Reconstr Surg Glob Open 2018;6:e2011; doi: 10.1097/GOX.0000000000002011; Published online 14 December 2018.)*

Malignant peripheral nerve sheath tumors (MPNST) account for about 5% of malignant soft-tissue sarcomas[1] and derive from neuroepithelial tissue.[2] The variety of MPNST (epithelioid, with mesenchymal differentiation, melanotic, and with glandular differentiation[3]) makes the establishment of standardized treatments and the development of modern molecular targeted therapies difficult.[4]

Unfavorable prognostic factors are large tumor size, truncal localization, prior irradiation, local recurrence, metastasis, and high tumor grade.[2,4,5] The presence of neurofibromatosis (NF)-1 is a debatable unfavorable prognostic factor as there are also data showing no significant difference in outcome for patients with and without NF-1.[2,5]

Prognosis for patients with MPNST is poor[2] due to their high metastatic potential, according to Collin et al.,[6] they even have the highest local recurrence rate of any sarcoma.

Tumor-free surgical margin status is believed to have the most beneficial impact on long-term survival and possible cure.[7,8] Distant metastases are common (40–80%), most frequently located in the lung, liver, and lymph nodes with a high frequency of local recurrence (in 22–45% of cases).[2,9,10] Patients with positive surgical margins bear a 2.4-fold risk of developing a local recurrence and a 1.8-fold risk of dying of disease.[1] Overall median survival ranges from 44 to 66 months.[7,8,11] Standards in the therapy of MPNST are lacking due to their low incidence and the treatment of these patients in different departments (plastic surgery, hand surgery, neurosurgery).[2,7]

Our aim is to report a case and describe an innovative and successful oncologic and reconstructive treatment method to an isolated median nerve MPNST in a young patient with no history of NF-1.

To our knowledge, there is a case report describing a similar treatment of a MPNST of a median nerve in a 73-year old man. A nerve reconstruction by 4 sural nerve grafts, opponensplasty, a latissimus dorsi flap, intraoperative brachytherapy (BT) radiation of 31 Gy and an additional external beam radiation therapy of 36 Gy[12] were performed.

CASE

A 30-year-old female patient referred to our department of plastic surgery with a history of carpal tunnel surgery of the left hand performed by a qualified hand surgeon. Intraoperatively, a solid mass originating from the median nerve was found and an incisional biopsy was made. Histopathologically, a MPNST of the median nerve was diagnosed. No comorbidities existed.

One month after the first surgery, a radical excision of the tumor with wide resection of the median nerve and partial resection of the thenar muscles was performed (Figs. 1, 2) with a maximum defect of 7 cm of the median nerve with 1 proximal ending and 4 distal endings (Fig. 3). The MPNST was classified as pT1b G2 R0 L0 V0.

Two weeks later, an opponensplasty using the flexor digitorum superficialis IV-tendon was performed, as well as a nerve reconstruction of the median nerve using 4 decellularized, processed human nerve allografts (Avance nerve graft, Axogen, Fla.). The length of the nerve allografts were 6 cm (from median nerve to N1/N2), 7 cm (to N3), 5.5 cm (to N4/N5) and 5 cm (to N6/N7). Finally, 5 BT tubes were inserted (Fig. 4). A primary closure was possible.

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Dr. Liodaki and Dr. Robiller contributed equally to this work. Department of Plastic, Hand Surgery and Burn Care Unit, University Hospital Schleswig-Holstein, Campus Lübeck, Germany.
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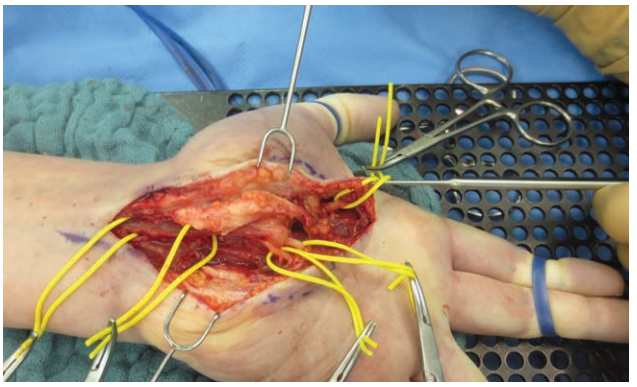

Fig. 1. The intraoperative finding of the tumor. A radical excision of the tumor with wide resection of the median nerve and partial resection of the thenar muscles was performed.

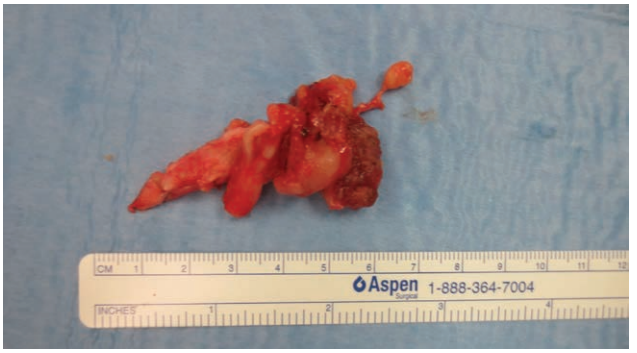

Fig. 2. Tumor tissue.

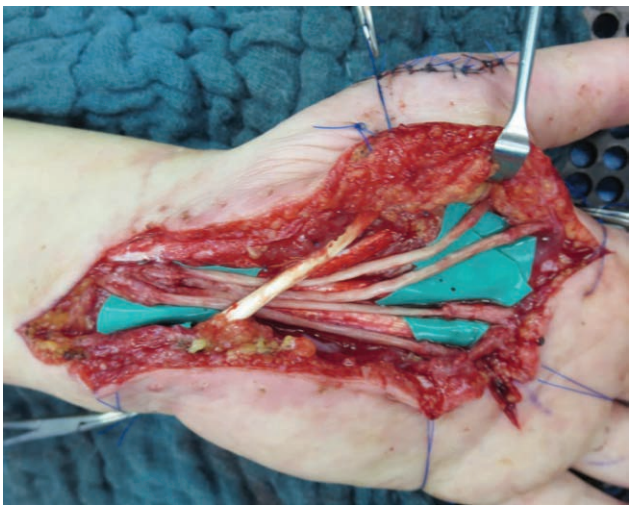

Fig. 3. It came to a maximum defect of 7 cm of the median nerve with 1 proximal ending and 4 distal endings.

Treatment commenced with interstitial BT with a total dosage of 30 Gy, fractionated in 2 × 2.5 Gy daily starting 4 days after surgery. No complications occurred postoperatively, the tubes were removed upon completion of BT.

After 12 days of hospitalization, the patient was released. Three months postoperatively, a complex regional pain syndrome occurred. This was treated conservatively with intensive physical and occupational therapy as well as pain medication.

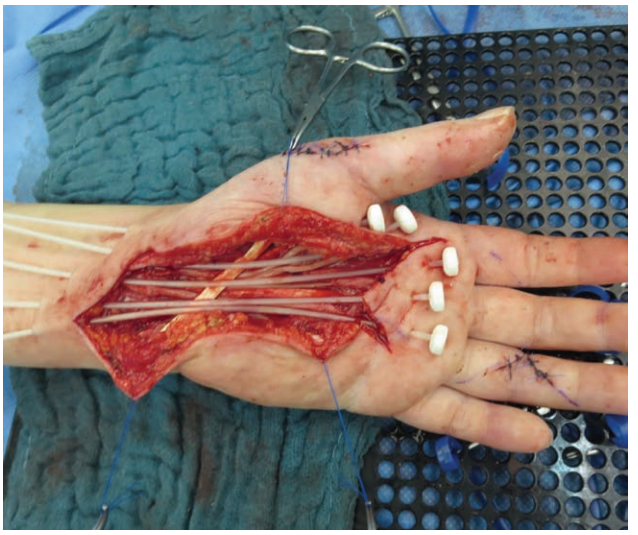

Fig. 4. An opponensplasty using the flexor digitorum superficialis IV-tendon was performed, and a nerve reconstruction of the median nerve using 4 decellularized, processed human nerve allografts. Five BT tubes were inserted. A primary closure was possible and there was no necessity for a flap.

On follow-up 1 year postoperatively, the patient reported cold intolerance, a feeling of stiffness and hyperhidrosis of the left hand.

The physical examination 2 years after surgery showed no significant changes compared with the year before, thus an active limitation of finger flexion existed (the distance from fingertip and palmar crease was 2.5, 2, 0, and 0 cm from DII to DV). Passive range of movement was complete. Opposition of the thumb remained restricted in the left hand (Kapandji score 8) compared with the right hand (Kapandji score 10). The sensibility of the hand was examined by static 2-point-discrimination (2PD) and presence of protective sensitivity. Protective sensitivity was given except for the digital nerves N4 and N5 of the left hand. The difference of the static 2PD of injured and uninjured contralateral digit (Δ2PD) in N1–N7 was 29–32 mm. In the area of the nerve reconstruction, the Hoffmann-Tinel-sign was positive.

Examination of pinch strength of the left hand with a pinch meter revealed 2 kg in contrast to 6 kg of the right hand. Strength of the hand measured by Jamar dynamometer was 5 kg in the left and 20 kg in the right hand. Hyperextension of the proximal interphalangeal joints and flexion of the distal interphalangeal joints of the fourth and fifth finger were noted, and hypotrophy of the thenar musculature. We also performed a DASH-questionnaire, analysis showed a result of 61. The patient's main limitations turned out to be tasks requiring physical strength like carrying heavy bags rather than manual dexterity.

Thirty months after initial diagnosis, there is no evidence for recurrence of the patients' disease.

DISCUSSION

There are contradictory data regarding the appropriate treatment of MPNST suggesting that further investigation of this tumor entity and its therapy is needed.[5] Big randomized and recent trials comparing different treatment strategies are lacking because of the low incidence of this disease.[2]

Regarding the surgical tumor resection, the usual treatment fields included the primary tumor sites with 3–5 cm margins, and no removal of regional lymph nodes is necessary.[9]

Adjuvant chemotherapy is controversial, as it has not significantly altered survival in patients with MPNST.[11] The role of radiation therapy in MPNST is expanding, and many authors support the use of adjuvant radiation therapy despite having clear surgical margins. A disease-free survival using combined surgical and radiation therapy for MPNST was demonstrated in 56% of the cases.[11]

Anghileri et al.[1] made the recommendation to consider radiation therapy for all operated patients, as they were able to show the beneficial impact of radiation therapy on local control rates. Advantages of BT are short treatment times and targeted dose distribution to the tumor bed.[13] It is a well-accepted treatment strategy on local control of MPNST, especially since wide local excision has replaced amputation for extremity sarcomas to preserve the structure and function of the affected body part, if possible. Important limitations of BT are the risk of wound healing complications and inhibition of nerve regeneration.[2,13]

In conclusion, a reconstructive approach combined with BT is a safe and successful oncologic treatment method. It offers a good local control and may preserve the function of the affected body part or even prevent amputation.

Eirini Liodaki, MSc
Department of Plastic, Hand Surgery and Burn Care Unit
University Hospital Schleswig-Holstein, Campus Lübeck
Ratzeburger Alle 160
23538 Lübeck
E-mail: liodaki.eirini@gmail.com

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