Two case reports of fair lower limb function after sciatic nerve sacrifice for the treatment of a malignant peripheral nerve sheath tumor

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Abstract

Wide resection for malignant soft tissue tumors may require resection of a major nerve. When a limb salvage procedure is selected in these cases, the anticipated functional outcome of the procedure must be clearly discussed with the patient, as recovery of normal function cannot be expected. In this report, we describe our surgical management of two cases of malignant peripheral nerve sheath tumor, a 58-year-old man and a 10-year-old girl, requiring a limb salvage procedure with sacrifice of the sciatic nerve. Although both patients required a short-leg brace for walking, because of a drop foot, both patients reported satisfactory functional results and were able to perform their activities of daily living with only slight difficulties. Based on our experience, limb salvage surgery can be considered for the treatment of malignancies involving the sciatic nerve, with fair functional outcomes expected.

Keywords MPNST · Limb salvage surgery · Sciatic nerve · Function

Introduction

Wide resection for malignant soft tissue tumor may require sacrifice of a major nerve. Particularly, in patients with malignant peripheral nerve sheath tumor (MPNST), sacrifice of the nerve trunk for curative treatment may be required. In these cases, it is important to achieve a superior activity of daily living (ADL) than at baseline when deciding to proceed with resection of a major nerve, such as the sciatic nerve, as part of the treatment for a malignant neoplasm. Although the number of limb salvage procedures performed has increased in recent years, there is a little literature regarding postoperative functional outcomes in these cases.

Our aim in this report was to describe the clinical features and postoperative functional outcomes for two patients with a MPNST treated with a limb salvage procedure requiring

sacrifice of the sciatic nerve, where functional outcomes were evaluated using the method of Enneking et al. [1] and knee range of motion.

The patients and/or their families were informed that the data from the case would be submitted for publication, and we received their consent.

Case reports

A summary of these two cases is presented in Table 1. Enneking et al.'s scale for assessment of limb function includes the following six factors, each scored on a 5-point scale, with better function indicated by a higher score to a maximum score of 30 points: pain, function, need for external support, walking ability, gait, and satisfaction (Table 2).

We evaluated each case of functional outcomes using the scale 2.5 years after the operation on case 1, and 1 year after the operation on case 2.

Case 1

A 58-year-old man presented with a complaint of left hip and thigh pain that had persisted for over 1 year. His family and medical history gave no indications of a genetic

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Table 1 Summary of the two cases

Case	Age	Sex	Histology	Site	Chemotherapy	Follow-up (years)	Prognosis
1	58	Male	MPNST	Left hip	ADR + IFM	3	CDF
2	10	Female	MPNST	Right thigh	None	1.5	CDF

Age is expressed in years

Dosage, [adriamycin (30 mg/m² × 2 days) and ifosfamide (2 g/m² × 5 days)] × 4 courses

MPNST malignant peripheral nerve sheath tumor, ADR adriamycin, IFM ifosfamide, CDF continuous disease-free period

Table 2 Functional evaluation of Enneking

Score	Pain	Daily function	Support	Walking ability	Gait	Satisfaction
5	No medication	No disability	No supports	Same as preoperative	No alteration	Would recommend to others
3	Non-narcotic analgesics	Recreational restriction	Use of brace	Significantly less	Cosmetic alteration	Would do again
1	Intermittent narcotics	Partial occupational restriction	Use of one cane	Unable to walk outside	Minor functional deficit	Would repeat reluctantly
0	Continuous narcotics	Total occupational restriction	Use of two canes	Can walk only with assistance or wheelchair bound	Major functional deficit	Would not repeat

Pain includes both the intensity and the effect of pain of daily function, and is quantified by the type of analgesic medication required by the patient for pain relief. Function assesses restrictions in activities (actual or prohibitive) and the effect of these restrictions on a patient's life style, and requires knowledge of the patient's pretreatment occupation and the degree of occupational disability caused by the physical restrictions. Supports refers to the type and frequency of use of external supports that are necessary to compensate for weakness or instability during standing and/or walking, and considers both the type and frequency of use. Walking ability refers to the limitations imposed on walking by the procedure, and requires knowledge of the maximum distance walked and the type of limitations (as for example, indoors and/or outdoors and managing inclines and stairs). Gait refers to the presence or absence of gait alterations and the effect of these alternations on function and restrictions in activities. Satisfaction assesses patient satisfaction with the procedure, and includes both the emotional reaction to the procedure and the subjective perception of functional outcomes

predisposition to malignant tumors. His family doctor had observed a soft tissue tumor in his left hip, which prompted a referral to our center. Magnetic resonance (MR) imaging revealed an isointense mass (on T1-weighted sagittal image; Fig. 1a), with heterogeneous hyperintensity (on T2-weighted axial image, Fig. 1b, c), that was contiguous to the sciatic nerve. A needle biopsy was performed, with a pathological diagnosis of probable leiomyosarcoma. Two cycles of chemotherapy (combination of adriamycin and ifosfamide) were administered. After chemotherapy, we performed a wide resection of the tumor, which included the sciatic nerve. The definite pathological diagnosis was MPNST (Fig. 1d). Postoperatively, the patient completed two cycles of adjuvant chemotherapy. The patient has remained disease-free for 3 years.

With regard to functional outcome, the Enneking score totaled 20 points (67%), with the following distribution: pain score of '1', with the patient requiring pregabalin 75 mg/day and tramadol 112.5 mg/day for left leg pain; function score of '3', due to occasional stumbling on stair ascent; support score of '3', with the patient requiring a short-leg brace for a foot drop; gait score of '5' points, with no significant gait

deviation or limitation, with the exception of the short-leg brace; and satisfaction score of '3' points, as the patient had hoped to be free of leg pain. He got sensory loss under his left knee. As for the MMT of his left lower leg, iliopsoas and quadriceps were 5, tibial anterior muscle, extensor hallucis longus, flexor hallucis longus, and gastrocnemius muscle were 0–1. However, he recovered an active and passive knee range of motion of '0' to 140° either, and he was able to walk independently (with the short-leg brace) while carrying a load.

Case 2

A 10-year-old girl presented with a 6-month history of right hip pain. Her family history had no indications of a genetic predisposition for soft tissue tumors. However, she had previously been diagnosed as a sporadic case of neurofibromatosis type1 (NF1). Her family doctor had identified a soft tissue tumor in her right posterior thigh, prompting a referral to our center. MR imaging revealed an isointense mass (T1-weighted sagittal image, Fig. 2a), with heterogeneous hyperintensity (T2-weighted axial image, Fig. 2b), which

Fig. 1 Case 1: 58-year-old man. We marked sciatic nerve with an arrow in (a–c). **a** Sagittal T1-weighted magnetic resonance (MR) image, showing an isointense mass which is contiguous to the sciatic nerve. **b** Sagittal T2-weighted MR image, showing a heterogeneous, high signal intensity mass which is contiguous to the sciatic nerve. **c** Axial T2-weighted MR image showing a heterogeneous, high signal intensity mass which involves the sciatic nerve. **d** Pathological examination of the biopsy specimen, with confirmation of a malignant peripheral nerve sheath tumor; magnification $\times 200$ (**d**)

was contiguous to the sciatic nerve. On positron emission tomography-computed tomography (PET-CT), the maximum standardized uptake value (SUV) within the tumor lesion was 7.4 (Fig. 2c), with no abnormal uptake at other site. A diagnosis of probable MPNST was made, based on the pathological assessment of needle biopsy. We performed a wide resection of the tumor, including resection of the sciatic nerve. Considering the concurrent diagnosis of NF1, chemotherapy was not recommended following tumor resection as the efficiency of chemotherapy in these cases is only estimated at about 50%. A definite diagnosis was MPNST was confirmed (Fig. 2d). The patient remained disease-free 1.5 years after surgery. With regard to functional outcome, the Enneking score totaled 28 points (93%), with the following distribution: pain score of '5', with the patient not requiring any medication; function, walking and gait scores of '5', with the patient being able to run; support score of '3' as she requires a short-leg brace for a foot drop; and satisfaction score of '5' as she does not report an inconvenience in her activities of daily living. She got sensory loss under

her right knee. As for the MMT of her right lower leg, iliopsoas and quadriceps were 4, tibial anterior muscle, extensor hallucis longus, flexor hallucis longus, and gastrocnemius muscle were 0–1. However, she recovered a knee active and passive range of motion of '0' to 140° either, and she was able to jump rope and ride a bicycle using a short-leg brace.

Discussion

We report two cases of MPNST treated using a limb salvage procedure that required resection of the sciatic nerve. Functional outcome in these two cases was evaluated retrospectively. MPNSTs comprise ~2% of all sarcomas, representing a small fraction of a group of cancers, with an annual incidence rate of 5 cases per million [2]. The most common sites of involvement are the nerve roots and bundles in the extremities and pelvis, particularly the sciatic nerve [3]. Half of MPNSTs are associated with NF1, which is an autosomal dominant condition, with an incidence rate of 1 per 3000

Fig. 2 Case 2: 10-year-old girl. We marked sciatic nerve with an arrow in (a, b). Sagittal T2-weighted magnetic resonance (MR) image showing a heterogeneous, high signal intensity mass which looks contiguous to the sciatic nerve (a). Axial T2-weighted MR image showing a heterogeneous, high signal intensity mass which involves the sciatic nerve (b). Positron emission tomography-computed tomography (PET-CT) showing a mass in the right posterior thigh, with a maximum SUV of 7.4 (c). Pathological examination of the biopsy specimen, with confirmation of a malignant peripheral nerve sheath tumor; magnification $\times 200$ (d)

live births, representing the most common genetic predisposition to cancer genetic in humans. Several large series report significantly worse outcomes for MPNST arising in setting of NF1 compared with sporadic disease, with inferior responses to cytotoxic chemotherapy and 5-year survivals that are up to 50% worse [4, 5].

Limb salvage surgery as a treatment for soft tissue tumor, with neoadjuvant and adjuvant therapy, has become the treatment of choice for most patients [6–8]. The decision to proceed with limb amputation or reconstruction should largely be based on the need for local or a wider extent of treatment [8], as well as on the anticipated function after surgery. Several authors [8] have recommended hip disarticulation or hindquarter amputation over limb salvage when peripheral nerves are involved based on the assumption that an extremity without tactile sensation after complete resection of sciatic nerve is not worth saving. Nambisan et al. [9] reported one case of limb-sparing surgery performed for a neurosarcoma of the sciatic nerve, which resulted in

moderate functional impairment, but with an overall acceptable functional result. Fuchs et al. [10] reported on the outcomes of 10 patients who underwent a lower limb salvage procedure which included resection of the sciatic nerve, with patients recovering the ability to perform more than 70% of their daily activities with a little or no difficulty. We achieved a comparable outcome with both patient able to perform their daily activities with slight difficulties. These outcomes are supported by a study which reported a higher level of disability after lower limb amputation than reconstruction [11].

Even if limb salvage surgery is justified oncologically and is feasible technically, the anticipated functional outcome must be discussed with the patient. As an example, in our two cases, the loss of motor and sensory supply to the foot after the sacrifice of the sciatic nerve required that both patients use a short brace to correct for a drop foot during gait. Therefore, the loss of function will need to be compensated with the use of an external device for the rest of the

patient's life. Both patients were satisfied with the surgery with regard to function, with the lower satisfaction score in case 1 being related to the persistence of pain after the limb salvage procedure.

There is a difference of functional result between our two cases. Case 1 was scored as 20 points, and it looks fair. On the other hand, case 2 was scored as 28 points, and it is excellent. The indexes of support, walking ability, gait are same points because both patients could walk outside same as preoperative with short-leg brace. There are some possible explanations for the differences of other score. Case 1 patient's occupation was transportation, so he had to carry burdens. It might to be influenced leg pain and the pain might lead decreasing a score of satisfaction. Bickels et al. reported patients who had resection of the nerve at a lower anatomic level had better functional outcomes than did patients whose resections involved a higher level because the innervation to the semimembranosus, semitendinosus, and long head of the biceps femoris is preserved in lower-level resections [12]. Case 2 was performed nerve resection slightly lower-level than case 1. It might be influenced the outcome. Moreover, younger age also affected for good functional outcome of Case 2.

Although we only report on two cases in our report, our findings do support limb salvage with sacrifice of the sciatic nerve as a feasible procedure for the treatment of a MPNST involving the sciatic nerve, with good functional outcomes expected. Our cases can also serve as an important teaching tool to help patients to understand the anticipated impact of the surgery on their function and ADL postoperatively.

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Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with human participants performed by any of the authors.

Informed consent The patients and/or their families were informed that data from the case would be submitted for publication and provided their consent.

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