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Original Article

Surgical management of giant neurofibroma in soft tissue: a single-center retrospective analysis

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Abstract: Neurofibroma, a common benign tumor in soft tissue, continues to grow, and often appears to be giant. In this study, we retrospectively analyzed the surgical treatment of 26 patients with giant neurofibromas in our clinic in the past 10 years from Jan. 2004 to Dec. 2013. The tumors were located in the head (n = 10), trunk (n = 9), limbs (n = 5), and multi-sites (n = 2). According to the location and extent of the lesion, as well as the adjacent anatomy, surgical management was performed to partially (n = 15) or almost completely (n = 11) resect the tumor. The wounds were repaired by skin flap or skin graft. Among them, one child with a giant tumor in the scalp underwent three times of skin expander treatment, and acquired complete removal of the tumor finally without baldness. Eleven cases underwent the interventional embolization of tumor's nutrient arteries, which successfully reduced the bleeding in operation. Most of the skin flap and skin graft survived well. After operation, the appearance of the patients and the function of the limbs were improved largely. In conclusion, for the giant neurofibroma, surgical treatment effectively reduces the tumor burden, rehabilitates the appearance and function, and so improves the quality of life. Skin expandor and interventional embolization of nutrient artery can be used when appropriate.

Keywords: Neurofibroma, operation, interventional embolization, expander

Introduction

Neurofibroma, a common benign tumor, may occur anywhere in the body, including skin and soft tissue, nervous system, muscle, skeleton, and visceral organs. Most of neurofibromas are solitary [1]. Multiple neurofibromas are called neurofibromatosis, including three types, type 1 neurofibromatosis (NF1), type 2 neurofibromatosis (NF2), and schwannomatosis [1, 2]. NF1 often affects soft tissue. The tumor continues to grow and expand so it often appears to be giant, which is a big burden for patients. The prolonged ulcer and repeating bleeding of the tumor may distress patients and impact their quality of life. Unfortunately, although we have known something about its pathogenesis [3-6], and found a few of drugs which had potential therapeutical effect [7-11], surgical management is still the most effective method to control or even cure this tumor [12, 13]. For giant neurofibroma, it is always a challenge due to the risk of excessive bleeding in the operation and the difficulty of repairing the huge wound. In this study, we made a retrospective analysis of the patients with giant neurofibroma in soft tissue who underwent surgical management in our clinic in the past ten years. The results would be helpful for the clinical practice in the future.

Patients and methods

This research was approved by the Committee on Clinical Investigation of Jinling Hospital. Informed consent was provided for the patients, according to the Declaration of Helsinki. All the patients meet the diagnostic criteria for neurofibroma [1]. From Jan. 2004 to Dec. 2013, 26 patients (15 males and 11 females) with giant neurofibroma in soft tissue were enrolled into the study. Ages of patients at admission ranged from 8 years to 60 years old. The tumors were located in the head (n = 10), trunk (n = 9), limbs (n = 5), and multi-sites (n = 2, both trunk and limbs). 14 patients had solitary lesions, and 12 patients were diagnosed as NF1.

Figure 1. Case1. A 9-year-old girl had a giant neurofibroma in the right scalp. The tumor was completely removed by two times of skin expandor treatment (A and B). And the remnant scalp scar was resected in the third treatment of skin expander (C).

All patients underwent lab examinations including blood routine, coagulation function, comprehensive metabolic panel, and other necessary examinations. Electrocardiogram, sternum, ultrasonography of abdominal organs, and magnetic resonance imaging (MRI) scanning of the tumor were also performed in every patient. With above examinations, the systemic

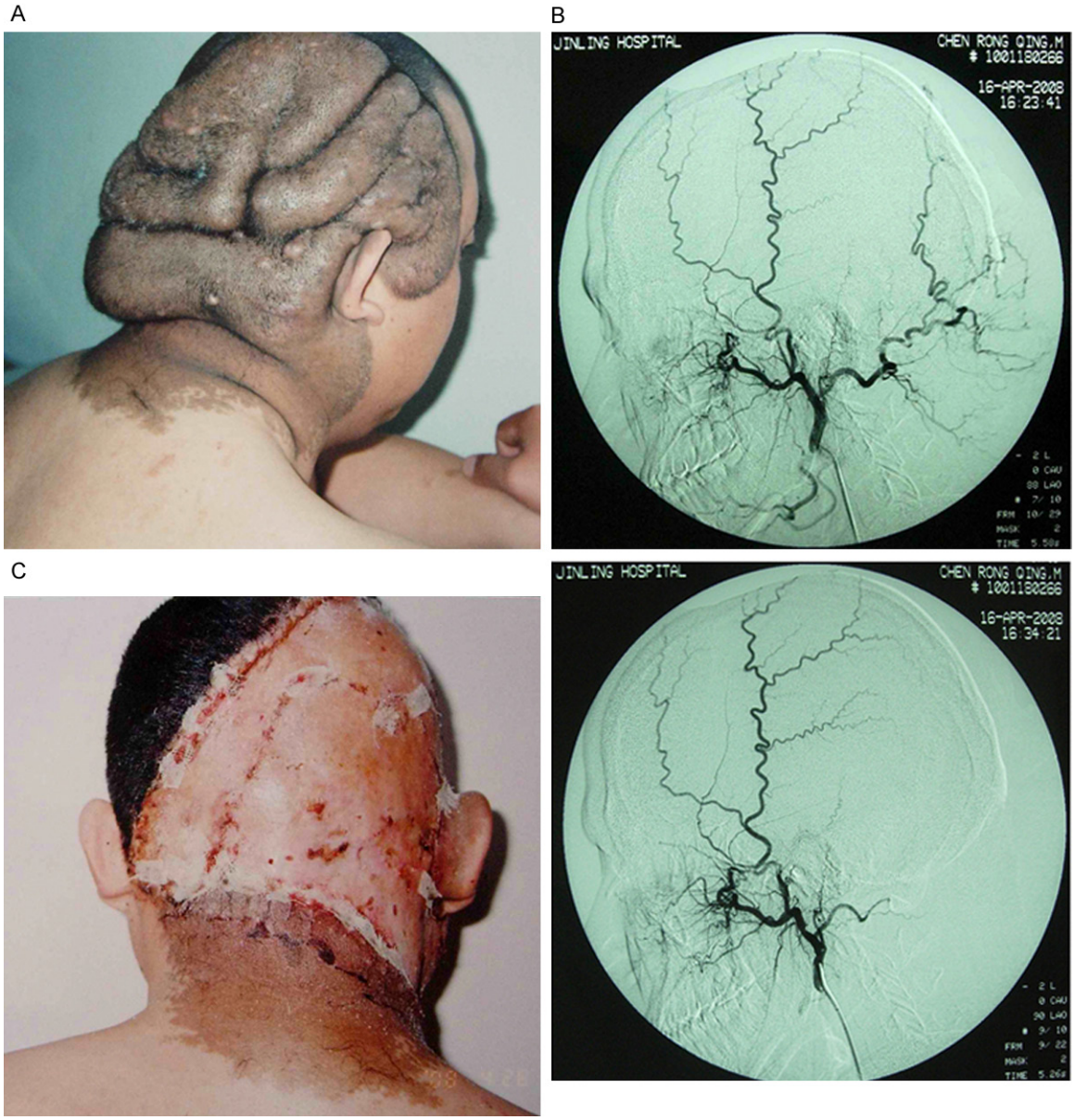

Figure 2. Case 2. A 43-year-old man had a giant neurofibroma in the right scalp (A). DSA showed that the right occipital artery was the main nutrient artery (B, above image). Selective embolization of nutrient artery and its branches was performed. And repeated angiography confirmed that most of the branches in the tumor were not displayed (B, below image). The tumor was resected and the wound was repaired by skin graft (C).

condition of the patient was showed. The extent of the tumor and the adjacent anatomy around it were displayed. Several patients with anemia and malnutrition received appropriate treatment before the operation.

The procedure of surgical management was set up based on the information from above examinations. For the tumor in the trunk or limbs, the aim of surgical management was to reduce the tumor burden and recover the function so we removed the tumor partially and repaired the wound by flap or skin graft. For the tumor in the head and neck, the appearance was very concerned by patients so we tried to make a complete removal of the tumor as far as possible. Skin expander should be used when possible to avoid the baldness. In order to reduce the bleeding in operation, a number of patients underwent the interventional embolization of

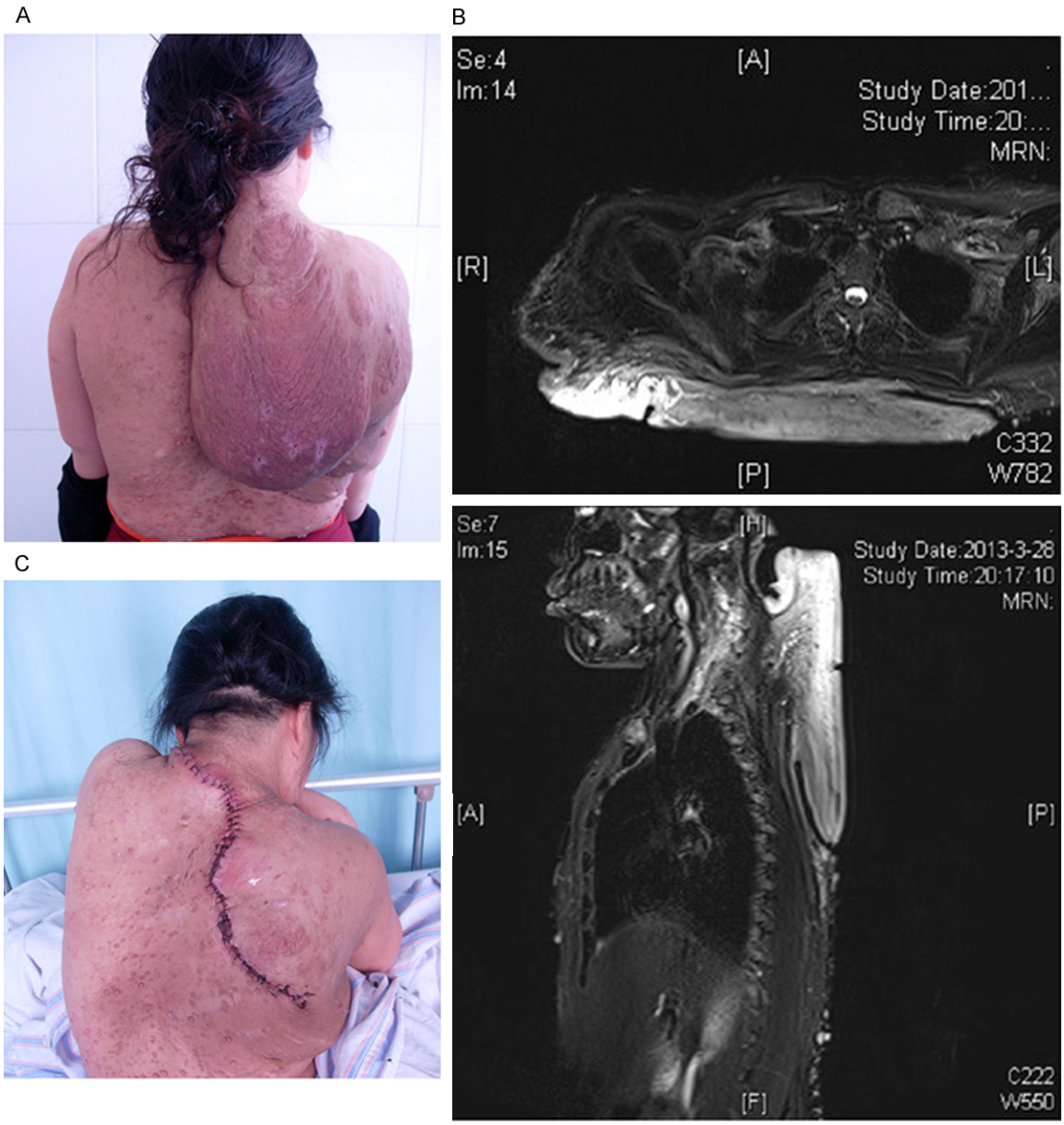

Figure 3. Case 3. A 40-year-old woman had a giant neurofibroma in the neck and back (A). MRI showed that the tumor invaded into the deep fascia and muscles with ill-defined boundary (B). The tumor was partially resected and the wound was repaired by skin flap (C).

tumor's nutrient arteries. Three days before the operation, digital subtraction angiography (DSA) was performed to show the nutrient arteries of tumor, and the embolization of the arteries and their branches in tumor was made with gelatin sponge particles. In operation, allogeneic blood transfusion or autologous blood transfusion was used when necessary. After operation, vital signs were monitored until the patients recovered well. The management of the incision, flap, or skin graft was done routinely.

Results

All of 26 patients underwent successful surgical management. Partial resection of the tumor was performed in 15 cases. And complete resection in 11 cases. The wounds were repaired by the flap or skin graft. Among them, one girl with a neurofibroma in the scalp was treated by skin expander for two times, with the complete removal of tumor finally; and the remnant scalp scar was resected in the third treatment of skin expander. Nine adult patients with

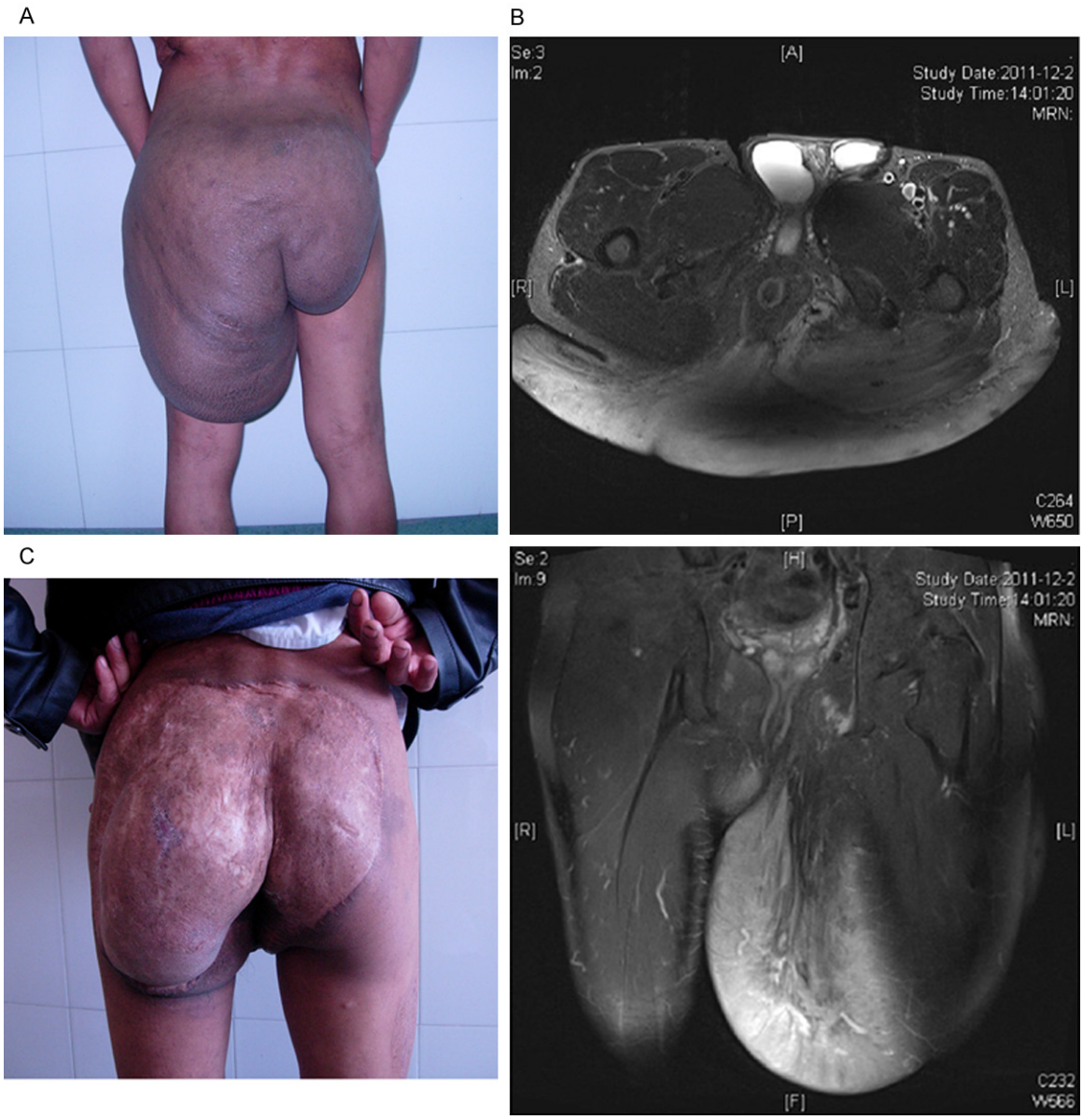

Figure 4. Case 4. A 55-year-old man had a giant neurofibroma in the hips (A). MRI showed that the tumor was located on the gluteus maximus muscle (B). The tumor was almost completely resected and the wound was repaired by skin graft (C).

neurofibroma in the scalp underwent interventional embolization of tumor's nutrient arteries and the complete removal of tumor with the wound repaired by skin graft. Two patients with very huge tumors in both trunk and legs also underwent the preoperative interventional embolization of nutrient arteries and the tumors were removed partially with the wound repaired by flaps. In our cases, most of the flaps and skin grafts survived very well. Skin necrosis occurred in the margin of flap in two cases; seroma under the flap in one case; and partial necrosis of skin graft in one case. The delayed healing of above incisions or wounds was acquired with several times of changing dressing carefully. Finally, all patients acquired an obvious improvement of their appearance and the function of the limbs. And so the quality of life was improved tremendously.

Figures 1-5 show five typical cases. In case 1, the neurofibroma in the scalp of a young girl was removed completely by two times of skin expandor treatment. In case 2, the neurofibro-

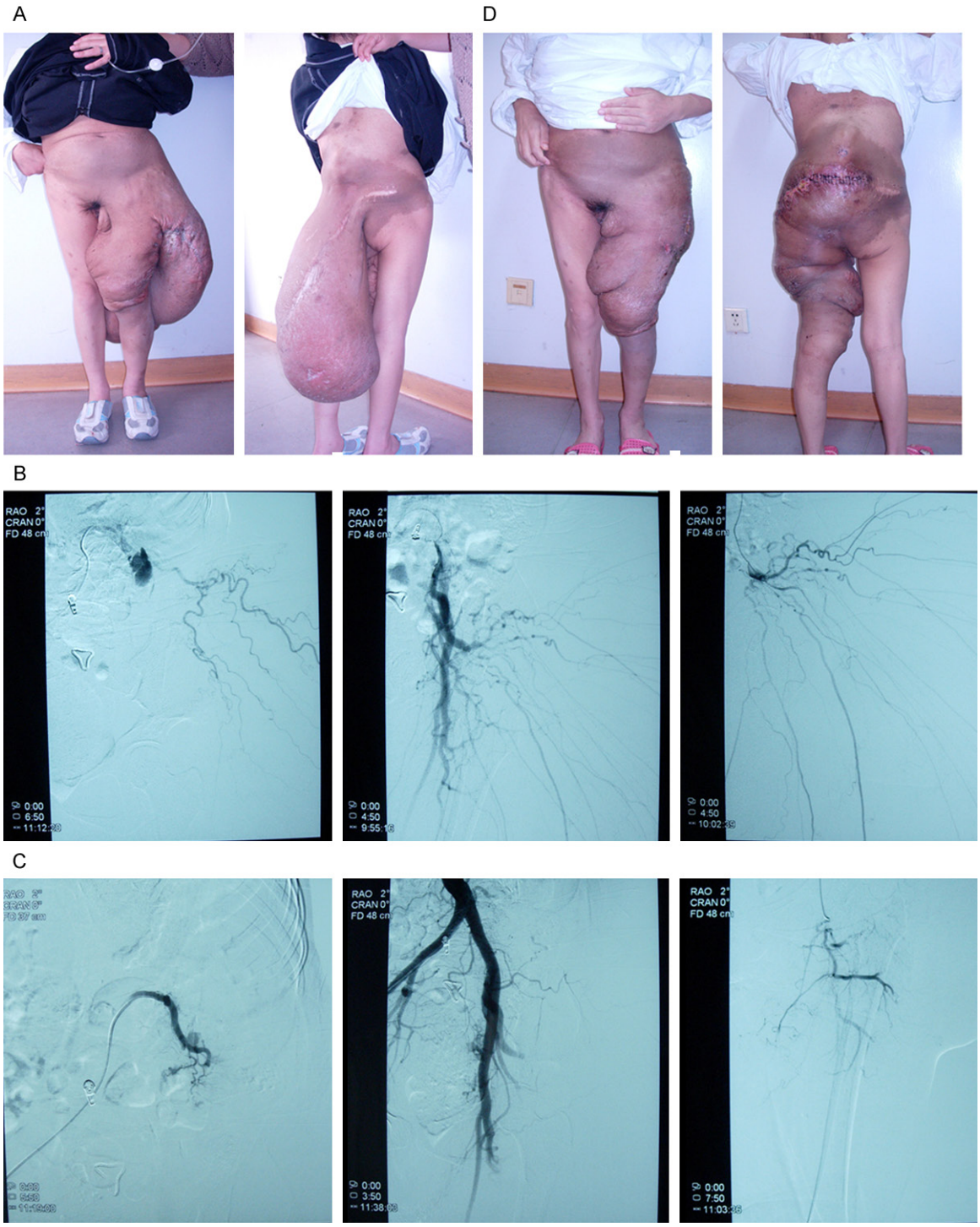

Figure 5. Case 5. A 46-year-old woman had a huge neurofibroma in the left hip and lower limb (A). DSA showed that the fifth lumbar artery (B, left image), superior gluteal artery (B, middle image), inferior gluteal artery (B, middle image), and lateral femoral circumflex artery (B, right image) were the main nutrient arteries. Selective embolization of the branches of above arteries was performed. And repeated angiography confirmed that most of the branches in the tumor were not displayed (C). A huge volume of tumor was resected and the wound was repaired by skin flap (D).

ma in the scalp of a 43-year-old man was removed after the interventional embolization of nutrient artery and its branches, and the wound was repaired by skin graft. In case 3, the

neurofibroma in the neck and back of a 40-year-old woman was resected partially, and the wound was repaired by skin flap. In case 4, the neurofibroma in the hips of a 55-year-old man was resected almost completely, and the wound was repaired by skin graft. In case 5, a huge neurofibroma in the left hip and lower limb of a 46-year-old woman was partially resected after the interventional embolization of nutrient arteries and their branches, and the wound was repaired by skin flap.

Discussion

Most of neurofibromas are sporadic. Systemic multiple neurofibroma, neurofibromatosis (NF), is an autosomal dominant disorder that arises from the abnormal differentiation of neural crest cells. NF appears as three types, including NF1, NF2, and schwannomatosis [1, 2]. The genes mutations responsible for them were located in 17q11.2, 22q11.2-q12, and SMARCB1 respectively [14-16]. NF1, also called Von Recklinghausen disease or peripheral neurofibroma, always involves skin and soft tissue, affecting approximately 1/3000 individuals without sexual prevalence [17]. Twelve patients in our cases were diagnosed as NF1. NF2, also called bilateral acoustic neurofibroma, is symmetric, non-malignant brain tumors in the region of the cranial nerve VIII [18]. Schwannomatosis is characterized by the multiple neurofibromas along the peripheral nerves [2, 16].

Neurofibroma grows invasively with ill-defined boundary. It can't be treated radically. Surgical management is the most effective method to control or even cure it. The procedure of surgical management was set up base on the adequate knowing the adjacent anatomy of the tumor and the comprehensive communication with the patients and their family members. For the neurofibroma in the scalp, the complete removal of the tumor should be achieved as far as possible to avoid the recurrence. Skin expander can be used to avoid the baldness if the patients have enough time to accomplish the treatment (such as case 1). If the patient didn't care the baldness, removal of the tumor and repair of the wound with skin graft also was also feasible, which reduced the times of operation and decreased the financial burden (such as case 2). For the neurofibroma in the trunk, if it was too huge to be resected completely, the partial resection of the tumor was made and the wound was repaired by flap (such as case 3). And if it wasn't very huge, almost complete resection of the tumor was made and the wound was repaired by skin graft (such as case 4).

Abundant malformed vessels had been found in neurofibroma, such as abnormal venous sinuses, aneurism and arteriovenous fistula [19]. So, the risk of excessive hemorrhage in operation is a challenge for surgeon. The preoperative embolization of tumor's nutrient artery was first reported by Littlewood in 1983 [20]. With endovascular techniques and new embolic agents, preoperative embolization has become a useful adjuvant method for the surgical management of giant tumors [21, 22]. In this group, nine adult patients with giant tumor located in the scalp (such as case 2) and two patients with very huge tumor in the button and lower limb (such as case 5) underwent preoperative embolization of nutrient arteries, which effectively reduced the bleeding in the operation.

In summary, this retrospective study shows that individual surgical management of giant neurofibroma effectively reduces the tumor burden, rehabilitates the appearance and function, and so improves the quality of life. Skin expandor, interventional embolization of tumor's nutrient arteries, and other assistant techniques can be used when appropriate. Of cause, the secondary deformity, such as the baldness in some cases, need further treatment.

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Disclosure of conflict of interest

None.

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