The Type of this Study of the EBM Level was Level IV: This is a Case Study. And Evidence Obtained from Multiple Time Series with the Intervention

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Abstract

Background: Neurofibromas are benign peripheral nerve sheath tumors. Solitary neurofibromas may be quite large. They are radioresistant and have poor sensitivity to chemotherapeutic drugs. Surgical resection, the remaining option, can be difficult due to the infiltrating nature of the tumor and the risk of massive hemorrhage.

Case Presentation: We report a patient with a massive (>80 kg) neurofibroma who was successfully managed with preoperative DSA (Digital Silhouette Angiography) and embolization of feeder arteries followed by surgical resection. After removal of the tumor of neurofibroma, the clinical course results are stable in the 2-year follow-up.

Conclusion: We found that preoperative feeder arteries branches of the left lumbar artery and the internal and external iliac artery were the main vessels supplying the tumor. DSA and embolization is technically feasible and speculated that it will be part of the perioperative plan for management of solitary peripheral neurofibromas. And future studies are recommended for determining the effectiveness of the treatment.

Keywords: Giant neurofibroma; Resection; DSA; Embolization

Introduction

Neurofibroma is a benign, heterogeneous peripheral nerve sheath tumor composed of fibroblast proliferations and Schwann cells [1]. Clinically it may appear as a hard and tough cylindrical or fusiform mass, as multiple small nodules, or as a localized lipoma-like mass. The latter are usually associated with neurofibromatosis type I [2]. Neurofibromatosis type can be divided into 3 types [3], the Neurofibromatosis type 1 (NF1) was accounted for 90% of the total incidence [4], it was an autosomal dominant genetic disease, and it was often accompanied by skin pigmentation, was characterized by skin milk coffee spots and freckles blue rubber-bleb nevus, also can cause bone, heart, kidney and other visera damage and dysfunction body show multiple or single neuroma. Giant neurofibromas of the trunk often require surgical treatment due to their severe impact on the appearance and function of patients. Neurofibroma weighing >60 kg has not been reported in the literature to date. In this report, we present a rare case of a massive (80 kg) neurofibroma that was successfully resected.

Case Presentation

A 34-year-old man with a history of neurofibromatosis type I, hypothyroidism and hashimoto’s thyroiditis presented with progressive enlargement of a tumor in the lower back and right leg over the last 3 months. The tumor was so large that he could not stand without support or lie in the supine position. The mass extended from the level of the 10th rib to the back of right knee. It was ~60 cm wide, ~30 cm thick and ~230 cm long. The overlying skin, which was pigmented dark brown, was soft in the region of the trunk but coarse and leathery distally. Sensation was reduced. The perianal skin was also affected, but penile and scrotal skin was normal. A second large tumor was present in the region of the nose. The total nasal volume was increased, and the skin surface was uneven. This tumor protruded into the right nasal cavity, affecting respiration. In addition, the patient had multiple small (0.5 cm × 0.5 cm to 2 cm × 1 cm) soft non-tender subcutaneous tumors and café au lait spots (0.5 cm × 0.5 cm to 2 cm × 2 cm) scattered all over the body. The clinical picture was characteristic of neurofibromatosis type I. There was no family history of similar
lesions. The patient was 150 cm tall and weighed 161 kg. On clinical examination his blood pressure was 135/85 mmHg; pulse rate, 87/min; respiratory rate, 18/min; and body temperature, 36.6°C. Chest radiography revealed scoliosis convex to the right. There was no spina bifida and no myelocele. B-mode ultrasound examination of the mass revealed many veins and venous sinuses within the tumor, with the largest vein being 1 cm in diameter. CT and MRI were not performed because the tumor was too large to fit into the scanner.

We decided to excise the tumor. Preoperative intravascular embolization also makes it easy for the surgical procedure of neurofibroma [5]. Intravascular embolization should be performed for aneurismal lesions once they are found [6]. DSA and embolization of feeder arteries was performed 2 days before the operation. Branches of the left lumbar artery and the internal and external iliac artery were the main vessels supplying the tumor. Punctured the right femoral artery (Figure 1 left figure arrow shows), introduced the guidewire (Figure 1 middle figure arrow shows), and placed the catheter tip in the right internal iliac artery, external iliac artery and lumbar artery angiography. DSA showed that the left lumbar artery branch, the right internal iliac artery and the right external iliac artery branch (upper femoral artery) were involved in the blood supply of the huge tumor. Blood vessels thickened and tortuous, typical tumor stain, rich blood supply 20 spring coils (Figure 1 right figure arrow shows) were given through the catheter (size: eleven 5 mm to 5 mm, Nine 5 mm to 8 mm.) It was shown that these vessels were embolized successfully after re-embolization angiography (Figure 1); the tumor supplying blood vessels were almost disappeared.

Resection of the mass was performed under general anesthesia. Mark the incision line down the iliac waist to the lateral side of the right thigh. Continuous suturing of all layers and ligation of feeding vessels was performed to reduce intra-operative blood loss, and suture of great vessels, was performed to reduce bleeding. Nerve monitor is used during the resection of tumor by electric knife so as not to damage the peripheral nerves of the tumor. The tumor tissue was removed within the marked line of the waist and hip. The skin tissue of the right thigh was formed into a local skin flap, closed part of the wound, and a drainage tube was placed. The left wound of the right hip joint was about 40 cm × 20 cm in size. The full-thickness skin graft of corresponding size was taken from the normal skin area of resected tumor body with new dermatome, and transplanted into the right hip joint to seal the wound. Total intraoperative blood loss was ~6000 mL. Blood transfusion included ~1200 mL of preoperatively reserved autologous blood and 1500 mL of intraoperatively salvaged blood plus ~2000 mL allogeneic blood. Total operative time was 15 h. The excised tumor weighed 80 kg as was shown in Figure 2. Differential diagnosis of sarcoma, neurofibroma, hemangioma and angiolipoma was made but biopsy confirmed the diagnosis of neurofibroma. Postoperatively, the patient recovered well, MRI did not show any recurrence so far during the 8-year follow-up.

**Discussion and Conclusion**

NF1, formerly known as von Recklinghausen disease, is an autosomal dominant neurocutaneous disorder with an estimated incidence of 1 in 3000 births [7,8]. Peripheral neurofibromas have traditionally been managed by surgery because these tumors are radioresistant and their slow growth rate diminishes their sensitivity to chemoradiotherapy [9].

NF1 can be classified into isolated, diffuse and plexiform forms. In this paper, the plexiform NF1 with rapid growth, large tumor, severe impact on the body appearance and the impact of tumor dropping on daily life is called giant neurofibromatosis. Typical plexiform neurofibromatosis is present at the birth of the child, with no symptoms at first or local skin spots and fat thickening. As the tumor grows with age, it will gradually grow up and show corresponding clinical symptoms [10]. NF1 is not sensitive to chemoradiotherapy, and surgery is the only effective treatment for this disease at present [11].

However, surgical resection can be difficult, and especially so when the tumor is large [12,13]. The two main challenges during surgery are the infiltrating nature of the tumor (which results in a high rate of tumor recurrence) and the risk of massive hemorrhage [13].
To ensure the success of the operation, we made extensive preparations, with several multidisciplinary consultations, and finally formulated a detailed standard operating procedure. Incidence tumor was giant, adopt extended resection ideas: (1) to reduce intraoperative bleeding, as is known to all, neurofibroma tumors had brittle, tumors had blood sinus of common sizes and sparse cellular organization, it is sometimes gray mud, rich blood supply and lack of smooth muscle, causes inelasticity in the lumen, intraoperative hemostasis was difficult, sometimes hemorrhage volume is bigger. DSA that the author’s team used was performed to realize the situation of traffic vessels, and catheter embolization was performed to reduce intraoperative bleeding, which were very convenient in both electrocoagulation and suture hemostasis. Nerve fibroma tumors contains a large number of blood sinus, blood sinus stored a lot of blood, intraoperative excision directly will lose excessive blood, operation needs a large amount of blood transfusion, although it has autologous blood reservation ,the resource waste were still larger. Neurofibroma tumors are lobulated, blood in blood sinus clean first, adopts the hinged joint firm can also reduce the risk of intraoperative hemorrhage. Meanwhile, intraoperative controlled hypotension, injection of ice saline and embolization of the main blood vessels are all effective methods to reduce intraoperative bleeding. (2) Selection of reverse skin grafting of tumor tissue and free skin graft transplantation: For the smooth, uniform and soft surface of tumor, we choose reverse skin grafting of tumor tissue, which can reduce secondary damage and problem of donor scar. (3) After tumor resection, vacuum sealing drainage therapy was performed in the operative area to increase the survival rate of skin grafting.

This method can maximally remove tumor tissue, preoperative feeder arteries embolization reduce the risk of surgery, and reduce the advantages of postoperative donor injury is worthy of clinical promotion, and speculated that it will be part of the perioperative plan for management of solitary peripheral neurofibromas. And future studies are recommended for determining the effectiveness of the treatment. As these tumors have high risk of recurrence, long-term follow-up is essential.

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