

# Aggressive intramuscular hemangiomas in the upper extremity

## A case report

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### Abstract

**Introduction:** Intramuscular hemangioma (IMH) is a rare congenital soft tissue tumor. Here, we report a case of IMH patient who had undergone several surgeries and other treatments that were all ineffective before he visited us.

**Clinical Findings:** This IMH patient was a 16-year-old male who was born with a tumor of unknown size in his right hand and forearm. On physical examination, the tumor and skin flap complex was seen with a size of 14 cm\_12 cm in his right hand, and the multiple postoperative scars were shown on his right hand and forearm. The patient was not able to raise his right shoulder, and the ranges of motion of his right elbow, wrist, and finger were almost zero degrees.

**Interventions:** Considering that the tumor had been surgically excised for several times and the multiple recurrences had affected adversely his daily life, an amputation of his right hand, forearm, and the part of his right arm was performed.

**Diagnoses:** The pathological examination confirmed the diagnosis of IMH.

**Outcomes:** After the amputation surgery, the patient gained a functional recovery and the tumor did not recur during the 2 years after the surgery.

**Conclusion:** A treatment of choice should be personalized according to an IMH patient's overall situation. For an IMH patient like our case with a history of multiple tumor recurrences, we suggest that an amputation surgery should be performed as early as possible to avoid the repeated, but ineffective surgical excisions and the unnecessary sufferings.

**Abbreviation:** IMH = intramuscular hemangioma.

**Keywords:** amputation, intramuscular hemangiomas, recurrence

Editor: Shizhang Ling.

*Consent:* The written informed consent was obtained from the patient for publication of this case report and all the accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal upon request. The ethical review has been approved by our ethics committee.

*Authorship:* HL conceived of the study, participated in the design and coordination of the study, and drafted the manuscript. HY and HS participated in the design of the study and performed the statistical analysis. All authors read and approved the final manuscript.

*Funding/support:* This study was supported by Zhejiang Traditional Chinese Medicine Research Program (grant number 2016ZA124, 2017ZB057), Zhejiang Medicine and Hygiene Research Program (grant number 2016KYB101, 2015KYA100), and Zhejiang Medical Association Clinical Scientific Research Program (2013ZYC-A19, 2015ZYC-A12).

The authors have no conflicts of interest to disclose.

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Medicine (2017) 96:3(e5939)

Received: 28 June 2016 / Received in final form: 25 December 2016 /

Accepted: 27 December 2016

<http://dx.doi.org/10.1097/MD.0000000000005939>

## 1. INTRODUCTION

Intramuscular hemangioma (IMH) is a rare soft tissue condition. Liston<sup>[1]</sup> reported the first case of IMH in 1843. It differs from the infantile hemangioma simply because it occurs in patients older than children.<sup>[2]</sup> It is generally considered that IMH is a congenital soft tissue disease. IMH can occur in a patient of any age, but it is commonly diagnosed in the early adulthood. Its incidence is not known yet, because most patients are clinically asymptomatic, so its diagnosis can be easily missed.<sup>[3,4]</sup> Surgical excision, sclerotherapy, and angiographic embolization are among the common treatment options currently available. However, surgical excision is the most preferred treatment of choice for this disease. Here, we report a case of an IMH patient who was treated by several surgeries and other treatments in the past, but all treatments were ineffective until we treated him.

## 2. CASE REPORT

A 16-year-old male who was born with a tumor of an unknown size in his right hand and forearm, which was diagnosed as a hemangiomatous tumor back then. However, none of his parents, siblings, or relatives has this disease. When he was 5 months old, the tumor was first partially excised in a hospital in Shanghai (Fig. 1). Although no postoperative infection occurred, the volume of the tumor was increased rapidly shortly after this surgery. A recurrence was suspected, but no any further treatment was attempted until the patient received an injection sclerotherapy at the age of 41 months old in a hospital in



**Figure 1.** It shows that the patient had a tumor in his right hand before the 1st partial surgical excision.

Zhengzhou. Unfortunately, the sclerotherapy was also ineffective, because the size of the tumor was still gradually increased with age. At the age of 7, he underwent a complete tumor excision surgery and an abdominal pedicle flap surgery to heal his right hand in a hospital in Beijing (Fig. 2). One year after the surgery, his right hand and fingers were separated from the abdominal pedicle flap. Surprisingly, during the separation procedure, the recurrent tumor was found around the margins of the skin flap and it had even progressed rapidly from his right forearm to the distal part of the right arm. Since then, 6 partial excisions were performed to stop the tumor growth in a hospital in Wenzhou, but they all failed (Table 1). Since then the recurrent tumor had grown so huge and heavy in his right hand so that the patient was not able to get up or walk independently; therefore, he depended on his family members for his daily activities. On his visit to us, the physical examination showed a huge “tumor and skin flap complex” of 14cm × 12cm in size in his right hand and the multiple postoperative scars on his right forearm. This patient was not able to raise his right shoulder on his own, but his right shoulder retained the normal range of passive movement. The deltoid muscle in his right arm had atrophied, consistent with the disuse muscle atrophy. The ranges of motion of his right elbow, wrist, and finger were almost zero degrees. The abdominal scars were the donor site complications caused by his previous surgeries (Fig. 3). Laboratory studies were unremarkable. X-ray imaging demonstrated the soft tissue swelling and the associated osteoporosis in his right hand (Fig. 4). A biopsy from the tumor was obtained and the pathological examination confirmed the diagnosis of IMH. Considering the facts that the tumor cannot be completely removed by a surgery due to its anatomical location, his daily life is adversely impacted by this ever recurring and fast growing tumor, and the patient will not tolerate more surgeries in the future, we reasoned that an amputation of his right upper limb to the middle of the right arm above the elbow should be the



**Figure 2.** It shows that the patient had the complete tumor excision and abdominal pedicle flap surgery at the age of 7.

best treatment of choice. The patient and his family member agreed with us about this treatment of choice.

The multiple IMHs were visible during the operation. Gross examination found some thrombi in the intramuscular arteries and malformed intramuscular veins in the segments of skeletal muscle. Pathological examination found the chronic inflammation in the soft tissue (Fig. 5A–D), the hyperpigmentation below the basal layer of the skin (data not shown), parakeratosis of the skin (data not shown), intramuscular lipoangioma (Fig. 5A), the dilated, thin-walled, sponge-like hemangiomas (Fig. 5B), the sac-like hemangiomas (Fig. 5C), and some thick-walled veins in the tumor (Fig. 5D). The patient is able to go back to school and lives independently after the amputation surgery. No evidence of local tumor recurrence was found at the 3-year follow-up visit (Fig. 6A). However, 1 year after the amputation surgery, the

**Table 1**

**The previous surgeries of patient.**

Visit	Age	Place	Therapeutic method	Result
1	5 months	Shanghai	Partial resection	Recurrence, increase of tumor size
2	41 months	Zhennzhou	Sclerotherapy	Recurrence
3	7 years	Beijing	Complete resection + abdominal pedicle flap	No evaluation
4	8 years	Beijing	Separation	Recurrence, increase of tumor size
5	8–16 years	Wenzhou	Partial resection	Recurrence, increase of tumor size



**Figure 3.** (A) The physical examination showed that there were multiple postoperative scars on the right forearm and a huge “tumor and skin flap complex” in his right hand. (B) Abdominal scars as the complications of the flap donor site.

patient got a prosthetic right upper limb and he started to exercise his atrophied muscles in his residual right upper limb by strenuously and excessively doing push-ups on the first day. In the meanwhile, a mass was found in the right supraclavicular fossa during this strenuous exercise (Fig. 6B). A magnetic resonance imaging showed a mass in the subclavian artery (Fig. 6C). However, this mass did not seem to cause any nerve compression symptoms, and it disappeared after a 2-week immobilization treatment. Therefore, this mass was suspected of a subclavian artery hematoma.

### 3. DISCUSSION

Hemangioma is a common soft tissue tumor, but IMH is a rare tumor constituting less than 1% of all hemangiomas.<sup>[5,6]</sup> IMH tends to occur in the deep fascia and the skeletal muscle and more often in the lower extremity.<sup>[5]</sup> Although IMH is a benign tumor in a deep soft tissue, it is locally aggressive.<sup>[7]</sup> Because it often invades its surrounding tissues, it is difficult to excise the tumor completely. In addition, it easily has a local recurrence. Its local recurrence rate is 18% to 61%.<sup>[5,6,8,9]</sup> IMH is composed of ectatic blood channels, so the differential diagnosis is mainly the intramuscular venous malformations. The intramuscular venous malformations are the most common lesion in the skeletal muscles and consist of dysplastic vessels that tend to grow because the vessels progressively dilate.<sup>[10,11]</sup>

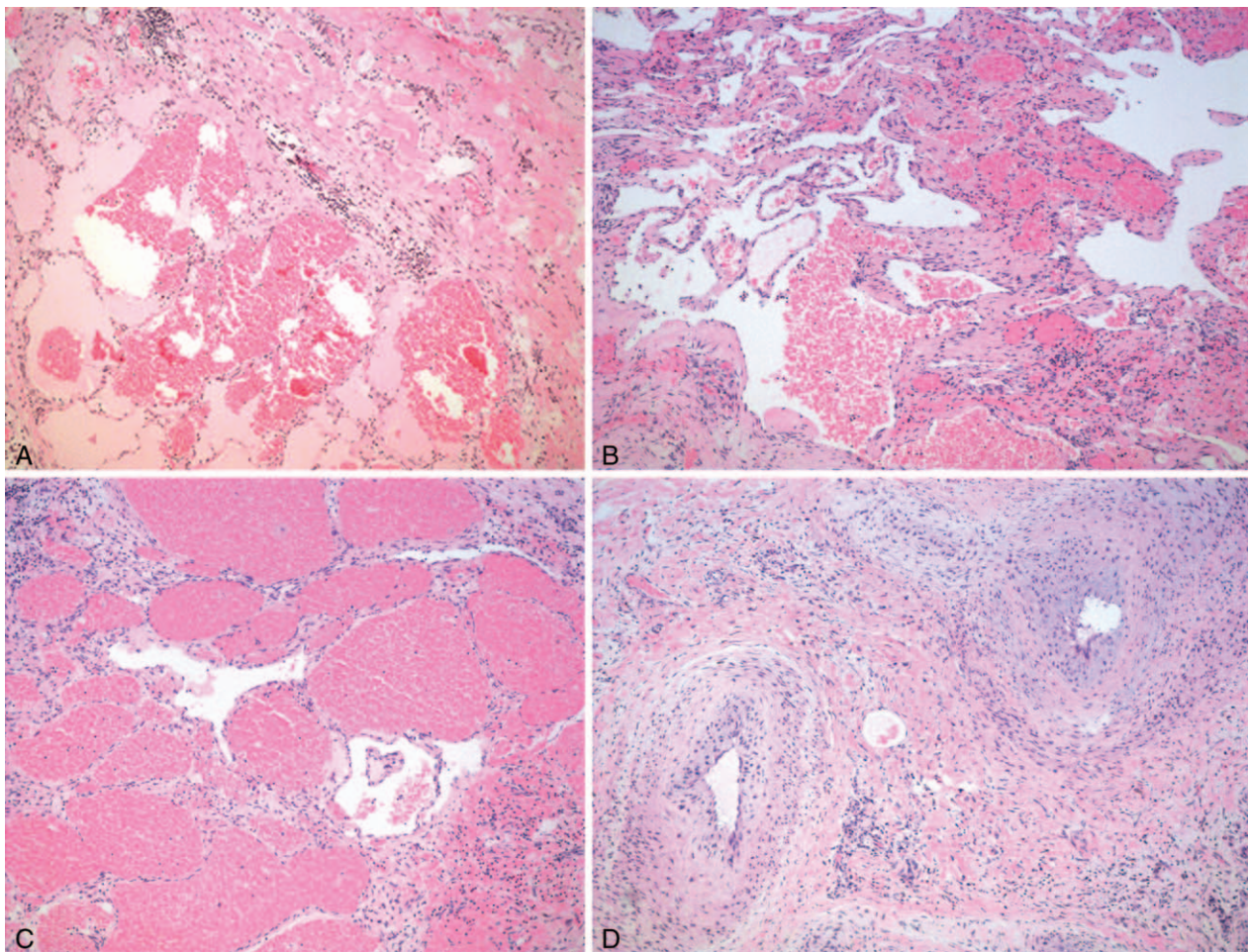
Surgery is the common treatment of choice for IMH. En-bloc resection of the tumor with the muscle involved is now the treatment of choice. Surgical margin is the major determinant of local recurrence-free survivorship for patients with IMH.<sup>[12]</sup> Due to the nerves, tendon, vascular tissue, and other essential structures surrounding the tumor, it is easy to cause an unintended physical disability if the tumor is excised completely. The main purpose of the surgery is to alleviate the local symptoms caused by progressive increase in size of the tumor such as pain, nerve entrapment, disturbance of the blood circulation of limbs, and functional impairment.<sup>[13–15]</sup> The patient in this case underwent multiple operations that all failed to stop the tumor growth, but caused the rapid progression of the tumor and the unnecessary injuries such as the abdominal postoperative scars in the donor site.

Chemoembolization, sclerotherapy, angiographic embolization, and combination therapy are the other treatment options.<sup>[16–18]</sup> No similar cases like ours with such a huge tumor have been reported in the literature. Unlike our case, the patients in the previous reports all had IMHs of a relatively small size. Therefore, we do not think that there exists a universal treatment of choice for all IMH patients, especially for our patient. Since the IMH tumor always recurred and grew faster after each surgery in

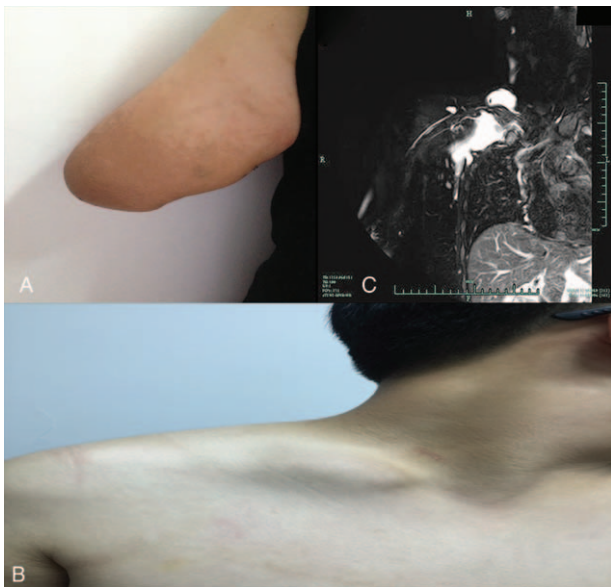


**Figure 4.** X-ray radiography demonstrated the swelling of soft tissue and the associated osteoporosis of the right hand.





**Figure 5.** Pathological microphotographs show the chronic inflammation in the soft tissue (A–D), the intramuscular lipoangioma (A), the dilated, thin-walled, sponge-like hemangiomas (B), the sac-like hemangiomas (C), and some thick-walled veins (D) and (hematoxylin-eosin staining, original magnification,  $\times 100$ ).



**Figure 6.** No local tumor recurrence was seen on the residual forearm (A). A mass was found in the right supraclavicular fossa during a strenuous exercise (B). Magnetic resonance imaging (MRI) imaging showed a subclavian artery hematoma (C).

the past, it was self-evident that more operations in the future would not help our patient at all; therefore, amputation surgery is the last resort to completely remove this tumor, to prevent our patient from suffering any ineffective surgeries in the future, and to eventually allow our patient to live a peaceful life.

#### 4. CONCLUSION

A personalized treatment of choice should be chosen according to an IMH patient's overall condition. For an IMH patient with a history of multiple tumor recurrences, we suggest that an amputation surgery should be performed as early as possible to avoid the repeated, but ineffective surgical excisions and the unnecessary sufferings.

#### Acknowledgments

The authors thank Dr Qiang Chen, Dr Hu Yang and Dr Hui Shen who have provided valuable assistance in the every stage of writing this manuscript. The authors also thank Zhejiang Traditional Chinese Medicine Research Program (grant number 2016ZA124, 2017ZB057), Zhejiang Medicine and Hygiene Research Program (grant number 2016KYB101, 2015KYA100), and Zhejiang Medical Association Clinical Scientific Research Program (2013ZYC-A19, 2015ZYC-A12) for the support. Last

but not least, I'd like to thank all of my friends and especially my lovely wife for her encouragement and support.

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