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Treatment of Giant Intramuscular Hemangioma: A Multistep Approach in Three Patients

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Abstract. *Background: Giant intramuscular hemangioma (GIH) is a rare, progressively enlarging benign tumor, characterized by variable presentation and usually initially diagnosed in childhood. Large volume, rapid enlargement and particular radiologic imaging create suspicion of malignancy. Radiologic investigations and needle or small excisional biopsy are not always reliable for an accurate diagnosis; therefore, histology on a large surgical specimen is often requested. The timing and modality of treatment of these tumors is a matter of debate. Patients and Methods: Data on 3 patients with GIH of the upper trunk and neck are reported. Associated vascular anomalies were found in all patients. All 3 patients had surgery because of the effect of the growth on their function, the severe symptoms and suspicion of malignancy. Results: A one-step excision of an enormous tumor was carried out in one patient, who died from severe postoperative complications. A second patient was successfully treated by a multistep surgical and multidisciplinary approach. An uneventful removal of part of the tumor was performed on the third patient, who is currently in follow-up for completion of treatment. Conclusion: Surgery remains the most effective mode of treatment for GIH and often results in permanent cure. The authors suggest performing the surgical removal of these tumors at first diagnosis, when their smaller size requires less demanding procedures, presents lower rates of morbidity and offers a better chance of complete excision.*

Intramuscular hemangiomas, first reported by Liston in 1843 (1), are rare benign neoplasms most frequently arising within the skeletal muscles and trunk extremities; they account for about 1% of all hemangiomas. Congenital, traumatic and hormonal theories have been proposed to explain the etiology of these growths (2-4). Any muscle can be affected, but those of the lower extremities are more frequently involved (5-7). Intramuscular hemangiomas may be divided on the basis of histologic features into capillary and cavernous, although mixtures of both histologic types have been described (8-10). These lesions are seldom correctly clinically diagnosed until they reach considerable size, because of their rarity, variable presentation and deep location (10). Clinical presentation, biologic behavior and recurrence rate vary according to histopathologic subtypes (9). The clinical history in the capillary variant is often short, characterized by a small swelling. The cavernous type is usually characterized by larger lesions and a longer clinical history (10). Pain and the presence of swelling are cardinal symptoms (11). Hemangiomas may proliferate and alternatively involute or increase dramatically in size (7, 9, 11, 12). Because many intermediate biologic and histologic types exist between benign hemangiomas and anaplastic angiosarcoma , histologic examination of a biopsy specimen is mandatory to evaluate the exact nature of the mass (7, 13-17). However, the results of a needle or a small excisional biopsy are frequently unreliable (15, 17) and, therefore, a large surgical biopsy is indicated for a definitive diagnosis (15). Further indications for surgery include rapid increase of tumor growth, control of bleeding, intractable pain, functional impairment, local skin necrosis, thrombocytopenia and cosmetic deformity (7, 12). The surgical treatment of such overgrown tumors may be extremely demanding and still not achieve complete removal. We report the clinical history, diagnosis and treatment of giant intramuscular hemangiomas (GIHs) in 3 patients.

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Patients and Methods

From January 2000 to December 2002, 3 cases of GIH of the thoracic wall and neck were identified from the files of the First Department of Surgery of the University of Rome "La Sapienza"

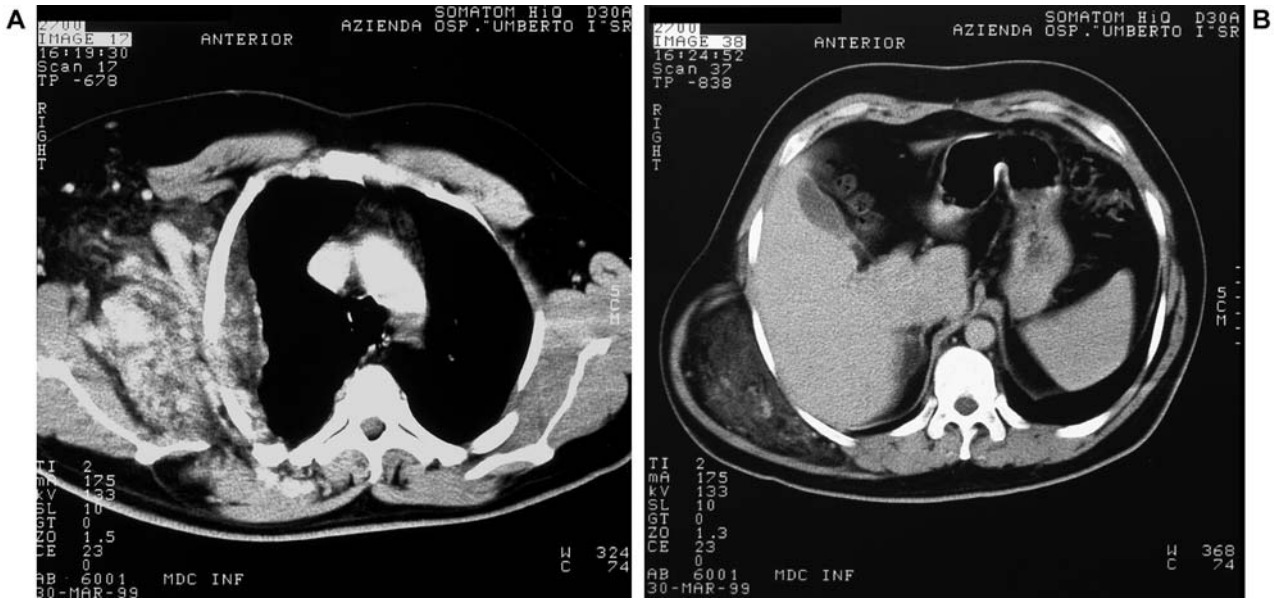

Figure 1. *A. Case 1. CT scan. The large mass involves the posterior right hemithorax of the thoracic wall with extension in the right pleural cavity. B .Case 1. The distal extension of the mass.*

Medical School, Italy. Clinical presentation, patient demographics, histology, treatment and outcome were recorded. Follow-up information was obtained by contacting the referring or primary care physicians.

Case 1. A 38-year-old man, affected by a congenital vascular tumor of the right posterior trunk, had undergone, at age 16, an aneurysmectomy of an ipsilateral subclavian branch; at the age of 37, he underwent a partial surgical removal of the tumor. He was admitted to our institution because of rapid growth of the tumor to a huge size, severe pain and hindered movement of the right upper limb. On clinical examination, a large tumor involving the posterior right hemithorax, the right supraclavicular region and the homolateral arm was observed. At angiography and computed tomography (CT) scanning, the mass was shown to be vascularized by anomalous, large branches arising from the subclavian, axillary and intercostal arteries. The presence of numerous, high-flow arteriovenous shunts described the hemangiomatous nature of the mass and indicated that a preoperative embolizing treatment would probably be ineffective. Furthermore, the CT scan showed an extension of the growth into the right pleural cavity (Figure 1). Intervention was planned to remove the extrathoracic part of the tumor and to reduce the blood flow to the intrathoracic extension. Removal of the intrathoracic part of the tumor would have been the object of subsequent surgery. A skin "U" incision, including almost all the right hemithorax up to the superior limb root, with a large flap, was performed. Complete replacement of almost all regional muscular structures, not involving the rib wall, was observed. A wide *en bloc* excision of the entire tumor mass was carried out, but was hindered by continuous blood loss which, despite attempts at adequate hemostasis, continued throughout the 8-hour operation. Residual tumor, close to the scapula, was left to avoid compromising the function of the superior limb and in the hope that further surgery or alternative treatments might be attempted. The pathologic examination of the specimen (7 kg, 30x21x10 cm) confirmed the cavernous hemangiomatous feature without areas of malignancy. Severe bleeding in the immediate postoperative period required reoperation. Oozing hemorrhage from the wide tumor bed was controlled. However, neurologic brain damage, sustained by hypotension, occurred. Furthermore, wound infection and the outbreak of general sepsis, supported by the large tissue detachment, led to the patient's death within a few days.

Case 2. A 21-year-old man, with a previous diagnosis of Budd-Chiari syndrome, was admitted to our department because of a huge tumor that had grown progressively since infancy and affected his right shoulder, lateral and supraclavicular regions of the neck and nuchal region. Magnetic resonance imaging (MRI) and CT scanning disclosed the involvement of the right brachial plexus, clavicle, scapula and lateral neck regions (Figure 2). A pseudo-capsule and an evident compression on the closer muscles were also observed. The most proximal part of the growth extended into the parapharyngeal space and infratemporal fossa, displacing the pharynx to the left. The jugular vein and carotid artery were displaced as well, whereas subclavian vessels were included within the growth. Neither bone and vascular involvement, nor intrathoracic extension were seen. Increasing enhancement after contrast infusion was apparent. Neither CT scanning, nor MRI could exclude malignancy. Large branches arising from the right subclavian artery, with arteriovenous shunt, nourishing the tumor, were observed at arteriography. Histology performed on an excisional biopsy showed features of intramuscular hemangioma. Because of the clinical behavior, radiologic features and increasing complaints from the patient,

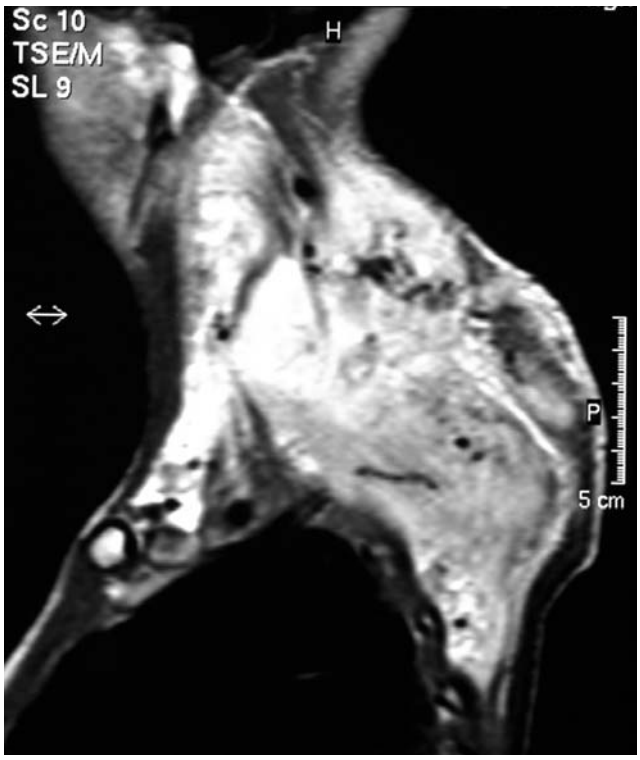

Figure 2. *Case 2. A lateral view of the chest MR image shows a large mass of the chest and neck.*

surgery was planned. A preoperative attempt at embolization of the tumor-feeding arteries (thyrocervical trunk, thoracoacromial artery, circumflex scapular artery, acromial branch) failed because of the high flow of the arteriovenous shunt. At surgery, infiltration to surrounding muscles appeared more severe than results on a previous excisional biopsy would have suggested. Surgery was, therefore, suspended to inform the patient about the need for a wide demolishing procedure and the related possible risk of loss of upper right limb function. A new intervention was carried out a few days later. The cervical and supraclavicular portion of the tumor were removed *en bloc* with the omohyoid and scalene muscles; the vagus and long thoracic nerve and brachial plexus were preserved by careful and accurate dissection. Carotid, vertebral and subclavian vessels were all removed, while segmental resection of the internal jugular vein and interruption of vessels supplying the tumor were performed. The tumor was classified at histology as an intramuscular mixed hemangioma. Two weeks later, through a posterior access, a third operation was performed and wide muscular destruction (deltoid, rhomboid minor and major, levator scapulae, splenius capitis and cervicis, lungus capitis) was performed. Histologic examination confirmed the previous diagnosis of mixed-type intramuscular hemangioma. The patient's immediate postoperative outcome was uneventful. A CT scan performed on postoperative day 45 showed the presence of a small tumor residue behind the pharynx. Nevertheless, the patient was discharged because local disease control and remission of symptoms were considered acceptable.

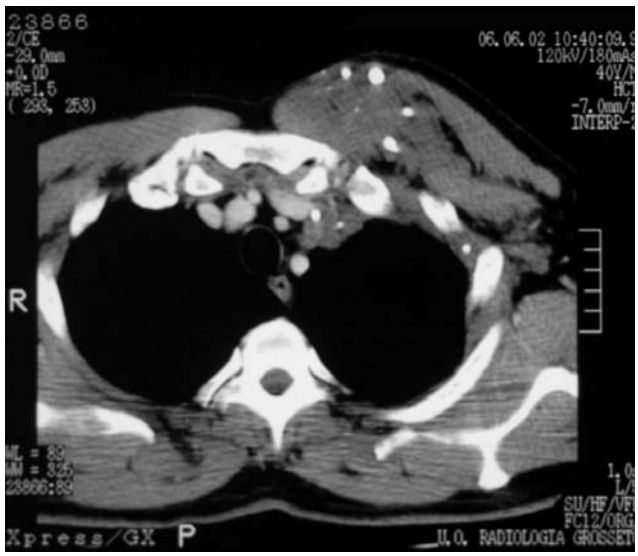

Figure 3. *Case 3. Chest CT scan.*

A further CT control, 3 months later, showed an increase in size of the residual mass in the pharyngeal and cervical region and a little regrowth close to the right scapula. A fourth operation to remove both lesions was completed by embolization of the vessels directed to the supraclavicular right region. At 6 months, the patient was well and disease-free, based on imaging results.

Case 3. A 44-year-old man had had a partial excision of a congenital intramuscular hemangioma of the left anterior hemithorax at age 14. He was admitted to our department because of pain and deformity from a large, slowly growing swelling at the site of the previous surgery. At CT scanning, calcification and muscular deformity were apparent inside the mass situated in the left pectoral region and protruding posteriorly into the left pleural cavity and anterior mediastinum (Figure 3). Here, a close adherence was observed to the ascending aorta and supra-aortic vessels; the most cranial portion of the tumor reached the supraclavicular fossa. No branch from the left subclavian vessels supplying the tumor and no left internal mammary artery could be seen at angiography. Complete removal of the superficial part of the tumor and reduction of the blood flow to its inner part were the aims of surgery. Through a wide "inverted L" incision on the left anterior hemithorax, along the parasternalis and the subclavicular lines, after interrupting the vascular supply to the tumor, a muscular flap was created with the major pectoralis muscle. The tumoral mass, replacing most of the intercostal muscles, consisted of several large, thin-walled dilated vessels connected with those nourishing the portion of tumor situated in the internal surface of the thoracic wall. Tearing on the parietal pleura, caused by interruption and stitching of these vessels, required the insertion of an intrathoracic drain. Tumor isolation and dissection of axillary vessels induced severe bleeding, which was controlled by packing and stitches. Histologic examination of the specimen (14x9x4.5 cm) revealed hemangiomatous features of arterio-venous and intramuscular mixed type without malignancy. The

postoperative outcome was uneventful and the patient was discharged on postoperative day 8. Four months later at CT scanning, no overgrowth of the residual intrathoracic tumor was seen. Removal of the residual tumor was, therefore, deferred and a further period of close follow-up decided on. Currently, local control of the disease seems acceptable and the patient is well 12 months after surgery.

Discussion

There is no agreement about the etiology of GIH, which is, however, mostly considered congenital (2-4, 6, 18-21). From an embryologic standpoint, these tumors represent mesodermal remains of malformed tissue that arrested in the endothelial stage of development, resulting in a meshwork of endothelial "lakes" (9, 22, 23). In the current series, vascular anomalies, found in all cases associated with the tumor, may be considered as supporting the congenital etiology. GIHs are often misdiagnosed, because of the depth of the tumor, the fiber thickening, or the fibrosis of surrounding muscles. The diagnosis of these tumors is more clearly established by imaging. The probability increases if the mass has a hyperechoic structure on ultrasound examination (5, 24). Plain radiographs and CT scanning are superior to MRI in identifying calcified thrombi, whereas MRI is preferred for defining the local extent of GIHs (14-16, 25, 26). The use of fine-needle aspiration biopsy more frequently results in inconclusive findings because of an excessively bloody specimen (5, 27).

The modality for treatment of these tumors has been a matter of debate for many years. The results of radiation therapy have been disappointing, because of side-effects consequent to the high doses necessary to obliterate the hemangioma (9). Cryotherapy and sclerotherapy have usually been ineffective, because they achieve only partial and temporary regression of the mass and relief of pain and compression symptoms (5, 28). The use of lasers is currently controversial (11), but may gain greater clinical application as favorable data accumulate. Biologic suppression with antiangiogenic agents may also become an alternative mode of therapy (11). However, all of these procedures should be, at least for now, considered as palliative and surgery remains the most effective mode of treatment offering the chance of permanent cure (7, 12, 18, 29). Incomplete excision, being associated with a high risk of relapse, cannot be recommended (7, 19); it may, however, represent a stage of a multistep treatment possibly associated with previous embolization, as shown in one of our patients. The current accepted treatment to minimize complications, as well as to halt destruction of the surrounding tissues, is total excision with a margin of normal muscles (5). When planning the surgical removal of GIH, angiography should precede the intervention because CT scanning and MRI only rarely provide details about the vascular supply to the tumor (7, 30). However, unfortunately, even arteriography in giant cavernous hemangiomas may not demonstrate direct feeding vessels (9, 10). Intra-arterial embolization combined with prompt surgery has been reported to give acceptable results because it lessens the risk of bleeding, which should be feared as a severe intraoperative complication (13, 31). Because of the infiltrative nature of GIH, the removal of normal muscle may be required well beyond the limits of the tumor to prevent local recurrence (5, 6, 8, 32). If complete excision is impossible because of extensive infiltration or the likelihood of permanent deformity or functional impairment, radiotherapy or embolization may be beneficial (5, 18). In these cases, residual tumor should be monitored with close long-term follow-up to diagnose and treat overgrowths in a timely fashion (6, 18, 32).

In all cases reported in the literature, including those of our report, the diagnosis of muscular hemangioma was initially made in childhood on much smaller lesions when compared with those treated in deferred surgery. No treatment is adopted at this point in the hope of an unlikely spontaneous regression until, finally, the dimension of the growth and the relevance of its complications make treatment necessary. Extremely demanding surgical procedures, invariably burdened by relevant complications and occasional mortality and meaningful anatomic and functional mutilations, are the outcomes of this "waiting" approach. The need for some change in this strategy seems desirable.

Our present conviction is that it would be considerably advantageous if surgery were performed at the initial diagnosis of these tumors. At this stage, the smaller size and less widespread local infiltration should significantly reduce the burden of surgery, musculoskeletal destruction and consequent permanent mutilations. However, were those tumors observed initially as giant lesions, a one-step aggressive treatment should be abandoned and a multistep approach preferred to lessen morbidity and avoid mortality.

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