Recurrent intramuscular myxoma: Review of the literature, diagnosis and treatment options

NILGÜN ÖZBEK¹, MURAT DANACI², BARIŞ OKUMUŞ¹, BİLGE GÜRSEL¹, ŞABAN ÇAĞIR¹, NEVZAT DABAK³, FILİZ KARAGÖZ⁴

Ondokuz Mayıs University, School of Medicine, Departments of ¹Radiation Oncology, ²Radiology, ³Orthopedic Surgery and ⁴Pathology, Samsun-Turkey

ABSTRACT
Intramuscular myxoma is a very rare benign soft tissue tumor. Local recurrence has not been reported after resection. We treated a patient with intramuscular myxoma by radiotherapy due to three recurrences after operation to reduce the tumor volume, to achieve local control and organ preservation. But we cannot obtain sufficient treatment response in our patient. Surgical excision seems to be the most effective treatment. After surgical excision, if recurrence occurs, histopathologic diagnosis should be reviewed. [Turk J Cancer 2006;36(2):75-78].

INTRODUCTION
Intramuscular myxoma (IMM) is a rare benign soft tissue tumor. The incidence varies between 0.10 and 0.13 per 100,000 (1). The majority of lesions present in fourth to sixth decade of the life and there is a slight female predominance (2). Clinically, tumor presents as a painless, palpable mass and its symptoms are dependent on size and site of the mass (3). Tumor may occur as an isolated lesion or associated with fibrous dysplasia or Albright syndrome (1). It is not possible to diagnose this tumor prior to histopathologic examination (2).

Plain radiography may be normal or may show nonspecific soft tissue mass. On computed tomography, IMM is a well-defined, homogenous, low-density mass within muscles. On magnetic resonance imaging (MRI), tumor has homogenous low signal intensity on spin echo T1 weighted images and homogenous bright signal intensity on spin echo T2 weighted images (4). In a patient with IMM, angiography is not proposed because this tumor is relatively avascular.

Differential diagnosis includes sarcomas, metastasis and benign intramuscular tumor such as lipoma, hemangiomma, hematoma, desmoid tumor.

Local recurrence has not been reported after resection in several cases reported in the literature. We treated a
patient with IMM by radiotherapy due to three recurrences after operation to reduce the tumor volume, to achieve local control and organ preservation. We present the results of treatment and discuss its histopathologic diagnosis, the differential diagnosis and the treatment modalities in this rare tumor.

CASE REPORT

A 70-years-old female patient presented with a mass and swelling in her right thigh and gluteal region. The patient had been operated two times due to the mass in that region in 1997 and 1998. Histopathologic examination proved IMM both in 1997 and 1998. The patient was admitted to our hospital due to third recurrence in 1999. On physical examination a 20x10 cm mass was detected under incision scar. On magnetic resonance imaging (MRI), the mass has low signal intensity on spin echo T1 weighted images and high signal intensity on fast spin echo T2 weighted images. The tumor heterogeneously enhanced after contrast administration (Figure 1). The patient was re-operated but the mass could not be excised completely. Pathological examination revealed spindle and satellite cells in an abundant myxoid stroma lobulated by fibrous bands of variable thickness and sparse vascular structures. The tumor further infiltrated adjacent striated muscle. No lipoblast could be identified by oil red-O stain. Immunohistochemically, the cells stained positively for vimentin, while s-100, myoglobulin and smooth muscle actin were all negative (Figure 2). A course of radiotherapy was planned in order to achieve local control with organ preservation, because the mass extended from perineal region to the knee. In the treatment of radiotherapy, two sets of treatment fields were used. The original volume encompassed the residual palpable tumor with 10 cm margin which correlated approximately 5 cm margin to preoperative MRI findings. The gross volume got a dose of 5000 cGy (200 cGy/day) and final tumor boost dose of 2000 cGy (200 cGy/day) applied which cones down on the residual tumor with 2 cm margin. Total dose was 7000 cGy. On control MRI performed 2 months later, the mass volume was the same, but necrotic areas were detected on contrast study. Tumor progression was observed on follow-up (Figure 3). Amputation was proposed to the patient, but she did not accept the operation and medical support. The tumor size increased on follow-up. The patient died due to vascular obstruction and infection in March 2001.

Fig 1. Sagittal, gadolinium enhanced spin echo T1 weighted MR image, a heterogeneously enhanced tumor is seen in right thigh. Non-enhanced areas within the tumor suggest necrosis

Fig 2. The tumor is composed of stellate-shaped cells and abundant myxoid stroma with scarce vascular structures (H&E, x100)

Fig 3. Sagittal, T2 weighted MR image, tumor progression is seen 9 months later
DISCUSSION

In the treatment of IMM, simple or wide local excision may be performed (3). In several cases published in the literature, recurrence has not been reported after resection (2,3,5). On contrary to known, recurrence has been detected three times in our patient and patient was operated three times due to recurrence. Recently, it has been reported that intramuscular low-grade myxoid neoplasm (cellular myxoma) is a soft tissue tumor with histology intermediate between intramuscular myxoma and low-grade myxofibrosarcoma or myxoid malignant fibrous histiocytoma. In contrast to intramuscular myxoma, it is characterized by the potential to recur locally (6).

Recurrence may probably be due to insufficient resection of the tumor or the tumor may have a different histology such as low-grade myxoid neoplasm in our case.

Primary concern of radiotherapy is to treat patients with malignant tumors. Even with recognition of the risks of late skin injury, carcinogenesis, leukemogenesis, and genetic damage from all ionizing radiation; radiation therapy also continues to be the accepted treatment for benign diseases that do not respond to other methods of therapy (7). Preservation of organ function and local control are important for benign tumors and somewhat similar to the indication for using radiotherapy on malignant and benign disorders. Radiotherapy offers therapeutic options for non-malignant disorders where other treatments are not as effective, induce more side effects or are less practical.

In the literature, radiotherapy with 60Cobalt device has not been used for the treatment of this tumor up to now. We treated the patient to achieve local control, to preserve organ function and to reduce tumor volume. But we cannot obtain sufficient treatment response in our patient.

Because of large radiotherapy volumes and high doses in our case, we also evaluated the radiotherapy side effects. The classic concepts of radiation pathophysiology are based on the concepts of the normal anatomic-physiologic or functional unit of an organ. Probably the most important modulators of radiation effects are the total radiation dose and fraction size, the duration of time during which the course of radiation delivered, the rate at which the radiation was given, the specific organ being irradiated, and the volume (8). The treatment field of our patient encompassed skin, bone, cartilage and muscle. The tolerance doses of bone and cartilage are greater than 70 Gy. The risk of necrosis rises with increasing volume and increasing dose. Near-tolerance doses typically used in curative radiation therapy result in changes that occur over a long period (8). Because of high radiotherapy tolerance doses of structures such as muscle bone cartilage in the radiotherapy field, we didn’t observe any life-threatening complication. This may be associated with the short survival of the patient due to her illness as well.

In conclusion, for the treatment of IMM, radiotherapy is not a suitable alternative treatment method. Surgical excision seems to be the most effective treatment. After surgical excision, if recurrence occurs, histopathologic diagnosis should be reviewed. The tumor may be low-grade myxoid neoplasm and treatment plan may be changed.
References


