

LEIOMYOSARCOMA PRESENTING AS A RECURRENT HEMATOMA OF THE DELTOID REGION. A CASE REPORT

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LEIOMYOSARCOMA PRESENTING AS A RECURRENT HEMATOMA OF THE DELTOID REGION (Abstract):

Leiomyosarcoma is a rare cancer of smooth muscle. The prognosis depends on the patient's age and the size, histologic grade and stage of the tumor. CASE REPORT: The case of a 63-year-old man diagnosed with a leiomyosarcoma of the deltoid region is reported. The patient was previously admitted twice in our surgical unit with a large recurrent posttraumatic hematoma of the right deltoid region which was incised and drained. One month later the patient was readmitted under the suspicion of malignancy. Following the large excision of the lesion, pathology and immunohistochemistry report showed a poorly differentiated leiomyosarcoma. One year later the recurrency of the tumour in the upper arm and axillary region imposed a large excision of the tumour of the upper arm and the axillary tumour, followed by a latissimus dorsi miocutaneous flap reconstruction. 4 months later multiple pulmonary metastases were documented at chest X-ray and chemotherapy with doxorubicine was commenced. A new axillary recurrence occurred 9 months later and was resected with clear margins followed by radiotherapy. In conclusion, the natural history and clinical picture of leiomyosarcoma may lead to diagnosis delay, poor local control of the disease with multiple recurrences and repeated surgery.

KEY WORDS: LEIOMYOSARCOMA; DELTOID REGION; HEMATOMA; RECURRENCE; SURGERY

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INTRODUCTION

Leiomyosarcoma is a rare cancer of smooth muscle accounting for 5–10% of soft tissue sarcomas [1]. The best outcome occurs when it can be removed surgically leaving clear margins. The prognosis depends on the patient's age and the size, histological grade and stage of the tumor. Patients older than 60 years of age, tumors larger than 5 centimeters or high-grade tumor histology are associated with a poorer prognosis.

While low-grade tumors are usually treated by surgery alone, higher grade sarcomas are associated with higher local recurrence rates and increased metastatic risk.

Patients with high-grade tumors, greater than 5 centimeters in size have the greatest tendency for disease to metastasize and are eligible for prospective clinical trials of adjuvant chemotherapy.

Doxorubicin alone or with dacarbazine is considered the best chemotherapeutic regimen for advanced sarcoma.

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CASE REPORT

A 63 year-old male patient, was admitted on January 2011 for a recurrent leiomyosarcoma of the right upper arm and axillary region. The history of the patient consisted of several hospital stays, initially being operated in another surgical unit for a hematoma of the right deltoid region following a contusion; incision with drainage was performed at that time. Further on, in January 2010, the patient was admitted on emergency basis in our surgical department for a recurrent large, under tension, hematoma of the right deltoid region (Fig. 1). Except for arterial hypertension under treatment, the general physical examination revealed no significant pathological conditions



Fig. 1 Recurrent hematoma of the right deltoid region

Lab tests (including coagulation profile) were within normal range.

Under the suspicion of a soft tissue tumour, the patient was operated on under general anesthesia. Following a large incision and evacuation of blood, the exploration of the wound revealed a cystic tumour arising from the deltoid muscle.

A large excision was performed, as frozen section examination from the tumour wall showed malignant cells (Fig. 2).

Definitive pathology report of the tumour described round epithelioid and fusiform tumour cells with a marked pleiomorphism and frequent mitoses with area of hemorrhagic necrosis.

The tumour was delineated peripherally by a conjunctive tissue, invaded in some areas by tumour.



Fig. 2 Wound aspect following large excision and drainage



Fig. 3 Tumour recurrence in January 2011



Fig. 4 Reconstruction with a latissimus dorsi flap

Immune-histochemistry diagnosis was poorly differentiated leiomyosarcoma (h-caldesmon+, smooth muscle actina+, vimentin+, citokeratin AE1/AE3 -, Myo D1, HMB45, S100 -).

Adjuvant radiotherapy was indicated by the oncologists but the patient neglected this treatment. At the current admission, the physical examination and ultrasonography revealed a large recurrence of the tumour (Fig. 3).

The tumour was excised and the large soft tissue defect of the upper arm was covered with a latissimus dorsi miocutaneous flap. The flap had a proximal pedicle of 10 cm length and was translated under a fascio-cutaneous bridge over the posterolateral arm defect (Fig. 4).

The pathology report from the both tumours showed leiomyosarcoma. The margins of resection seemed to be tumour-free.

On April 2011 he was admitted to the Oncology Unit when the pulmonary metastases were obvious of the chest X-ray and there were no signs of local tumour recurrence.

Chemotherapy with doxorubicine was well tolerated. In august 2011 palliative radiotherapy (TD of 8 Gy on the right arm and 16 Gy for the axillary recurrence) was performed. However, on September 2011, a new recurrence of the axillary tumour occurred (Fig. 5).

A large excision of the recurrent tissue, in the proximity of the right axillary vein was performed. Pathology report showed scanty foci of tumour cells. The postoperative recovery was uneventful. The patient is actually under the oncologist follow-up, with no signs of local recurrence, although complaining of moderate shoulder pain and impaired motility of the right arm (Fig. 6). No significant progression of the pulmonary metastases was recorded.

DISCUSSIONS

Soft tissue leiomyosarcomas are malignant tumours arising from the smooth muscle cells lining small blood vessels. They

have been classically been subdivided into three groups for prognostic and treatment purposes: leiomyosarcoma of somatic soft tissue, cutaneous leiomyosarcoma and leiomyosarcoma of vascular origin [1-3].



Fig. 5 Axillary recurrence



Fig. 6 No signs of local recurrence

There are no specific clinical features diagnostic of leiomyosarcoma of soft tissue, the disease typically occurring in the 5th and 6th decades of life. Initial imaging should include plain radiographs of the affected area, an MRI of the lesion and a chest CT-scan to evaluate for the presence of metastatic disease in the lungs. The presented patient was initially misdiagnosed due to misleading past history of a blunt trauma and due to lack of information consisting with a tumour at ultrasound scan of the soft tissues. Repeated hematoma

formation drew attention to a potential serious lesion, therapeutic decision being wound exploration, frozen section from the suspected areas and large excision. Staging of leiomyosarcoma is important both in guiding treatment and in providing prognostic information. The most commonly used system is the AJCC system (Table I) [2].

Table I AJCC staging system

Stage	Grade	Size	Location*	M**
IA	Low	< 5cm	Superficial or Deep	No
IB	Low	≥ 5cm	Superficial	No
IIA	Low	≥ 5cm	Deep	No
IIB	High	< 5cm	Superficial or Deep	No
IIC	High	≥ 5cm	Superficial	No
III	High	≥ 5cm	Deep	No
IV	Any	Any	Any	Yes

* relative to fascia; ** systemic or metastatic disease present

The reported case had, at the moment of first excision, a stage III leiomyosarcoma arising from the right deltoid muscle. Although adjuvant radiotherapy was indicated the patient refused it and after 12 months a tumour recurrence occurred and was excised. Then, ten months later, multiple pulmonary metastases were detectable on chest-x ray, corresponding to stage IV of the disease.

Due to the rarity of these tumors and the need for a multi-specialty treatment team, treatment is best carried out in a specialized center with expertise in sarcoma care [4]. A treatment plan can be formulated based upon the input from orthopedic and general surgeons, musculo-skeletal radiologists, pathologists, medical and radiation oncologists.

Patients older than 60, tumors larger than 5 centimeters or high-grade tumor histology are associated with a poorer prognosis.

Patients with high-grade tumors (grades 3 or 4) and larger than 5 centimeters in diameter have the greatest tendency for disease to metastasize and are eligible for

prospective clinical trials of adjuvant chemotherapy.

Doxorubicin alone or with dacarbazine is considered the best chemotherapeutic regimen for advanced sarcomas [5]. Although the patient had poor prognostic factors: age over 60, tumour size more than 5 cm, poorly differentiated tumour, hemorrhagic necrosis, stage IV of the disease due to pulmonary metastases, his biological condition is good. Stage and grading are the two most important prognostic factors for recurrence / metastasis [6-8].

Radiation therapy is an important additional treatment for improving rates of local control when surgical margins are close, especially in high-grade sarcomas.

CONCLUSION

The natural history and clinical picture of leiomyosarcoma may lead to diagnosis delay, poor local control of the disease with multiple recurrences and repeated surgery.

CONFLICT OF INTERESTS

None to declare.

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