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Synchronous presentation of extremity STS with the pulmonary metastasis- a case review

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IN BRIEF

Not avilable

INTRODUCTION

Soft tissue sarcoma (STS) encompasses an anatomically and histologically diverse group of neoplasms, most of which arise from mesodermal origin. Complete surgical resection is the cornerstone of treatment and prognosis is associated with tumor size, depth, histologic grade and anatomic site of the primary lesion [1]. Retroperitoneal/visceral (RP/V) sarcoma has a worse prognosis than extremity/trunk (E/T) lesions, and this is likely due to its frequent presentation with large size, precluding complete surgical resection [2]. Multimodality therapy with adjuvant radiation has been shown to improve local control in resected high risk extremity sarcoma [3, 4]. The data on the usefulness of adjuvant chemotherapy for high-risk STS is conflicting, but a meta-analysis of the published literature suggests a marginal benefit in disease-specific survival, but not overall survival [5]. Most histologic subtypes have the capacity to spread hemato-genously and regional lymph node metastases are rare. Pulmonary metastasis is the most common site of distant failure, but complete surgical resection of pulmonary metastasis has resulted in long-term survival in select patients [6].

The results of surgical therapy for pulmonary metastasis from STS have been reported by several groups [6, 7,8]. Careful patient selection is critical and only patients who have pulmonary metastasis amenable to complete resection, who have no evidence of extra-thoracic disease, and who are medically fit enough to tolerate a thoracotomy and resection should be considered. No randomized trial comparing surgery to observation or systemic therapy has been performed, but in several series the five year survival rate has been reported to be 20–40%, which is better than expected when compared to historical controls treated with chemotherapy or observation. [9]

In a large, recently updated series, patients treated with complete resection had an improved median survival of 33 months and a three-year actuarial survival of 46% [6] over previously reported three year survival of 23% [15] and is likely due to better patient selection and shorter follow-up with actuarial survival projection. Similar results were seen in a multi-center European trial in which disease-free post-metastasectomy survival rates were 42% at three years and 35% at five years [10]. In both studies, a long disease-free interval and complete resection were the most powerful, favorable prognostic indicators, underscoring the fact that careful patient selection is critical.

Due to the relative resistance of sarcoma to either chemotherapy or radiotherapy, compared to other solid tumors, surgical management of pulmonary metastases has been a pivotal therapy in this disease.[11]

Although no prospective, randomized trials have evaluated the efficacy of surgical resection of pulmonary metastases from soft-tissue sarcomas, multiple retrospective studies support the use of metastasectomy in selected patients. Many single-institution series report five year actuarial survival rates of 15% to 35% after complete resection. However, survival data are usually presented without the true denominator, and patients with limited disease amenable to resection are likely to have favorable tumor biology.[12]

Recently, these results have been verified with the publication of two large multi-institutional international retrospective databases: the European Organization for Research and Treatment of Cancer (EORTC) Soft Tissue and Bone Sarcoma Study Group [10] and the International Registry of Lung Metastases.[13]

A prospective database from Memorial Sloan-Kettering Cancer Center [6] outlined disease patterns in patients with soft-tissue sarcomas and pulmonary metastases. The median overall follow-up was only 9.7 months, and most patients received optimal multimodality therapy. In 18% of all patients, the pulmonary metastases presented synchronously with the primary tumor, whereas in 38% of cases the metastases developed metachronously. Soft-tissue sarcomas of the extremity and trunk accounted for 65% of all lung metastases. This distribution is similar to data from the Roswell Park Cancer Institute.[14]

According to the Memorial Sloan-Kettering and EORTC databases, the frequency of histopathologic subtypes of soft-tissue sarcoma that metastasize to the lung is as follows: leiomyosarcoma (19% to 21%), malignant fibrous histiocytoma (18% to 24%), liposarcoma (12%), synovial cell sarcoma (14% to 23%), fibrosarcoma (10% to 12%), and undifferentiated sarcoma (9%).[6,10] The incidence of pulmonary metastasis correlates with the incidence of high-grade differentiation within each histologic group. The majority (90%) of all lung metastases develops in patients whose primary tumor was high grade; 10% are of low-grade origin.

CASE SUMMARY

Our patient presented with 12x10 cm ulceroproliferative, friable, bleed-to-touch growth over medial aspect of right foot extending onto the dorsum for the past one and a half years with history of on and off bleeding.

No history of fever, significant weight loss, anorexia.

Patient is not a known case of tuberculosis, diabetes mellitus, hypertension, asthma with no past surgical history.

No h/s/o metastasis.

Performance status 70 by Karnofsky with poor nutritional status. Pallor present. No neurovascular deficit.

Hemoglobin level of 5.0 g% with rest of the investigations WNL.

X-ray chest was within normal limits. No pleural effusion or cannon ball opacities at the time of presentation.

Incisional biopsy showed growth to be a biphasic synovial sarcoma

Five units of blood was transfused and patient was built up. Since the limb salvage surgery was not possible, below-knee amputation was done when hemoglobin levels reached 10.0 g%. Post-op histopathology report showed synovial cell sarcoma with margins positive.

After four weeks patient presented with cough and bilateral cannon ball opacities on CXR PA view, s/o bilateral lung metastasis.

CONCLUSION

A high degree of suspicion is required while treating patients with STS for distant metastasis, especially pulmonary, as this is the most common site of distant failure, according to literature, and can present synchronously, as in our case, as well as metachronously with the primary.

Incidence of pulmonary metastasis is more in certain types like leiomyosarcoma (19% to 21%), malignant fibrous histiocytoma (18% to 24%) & synovial cell sarcoma (14% to 23%) which is the histological variant in our case.

Surgery plays a pivotal role in the management of pulmonary metastases, according to literature, but is not possible in all cases, as it is not in our case.

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