Local Recurrence of Low-Grade Myofibroblastic Sarcoma of the Chest Wall: Report of a Case and Literatures Review

Jie Wu¹
Gixun Chen²
Huinen Zhu²

¹Department of Thoracoabdominal Surgery;
²Department of Pathology, Zhejiang Cancer Hospital, Hangzhou 310022, Zhejiang Province, China.

Correspondence to: Jie Wu
E-mail: wujiephd729@126.com

KEYWORDS: myofibroblastic sarcoma, chest wall, recurrence.

Myofibroblastic sarcoma, composed primarily of myofibroblast, is a rare malignant tumor. Low-grade myofibroblastic sarcoma (LGMS) has been defined properly as a distinct entity in the 2002 WHO classification of soft tissue tumors[1]. Primary sarcoma of the chest wall is also a rare disease. This article describes a case of locally recurrent LGMS of the chest wall.

Case Report

A 71-year-old female with a history of a resected sarcoma of the chest wall presented for a follow-up examination 34 months after surgery. She had undergone wide excision of a soft tissue mass on the right chest wall in January 2004. The mass, together with involved intercostal muscles, ribs (fifth and sixth), and pleura was resected en bloc. Defects of the chest wall were repaired using Prolene mesh. Postoperative pathological diagnosis revealed low-grade fibroblastic/myofibroblastic sarcoma involving skeletal muscle, ribs, and pleura. On immunohistochemical staining, the tumor was negative for actin, desmin, CD34, cytokeratin and S-100, except for CD99. Her postoperative course was uneventful, and she received no postoperative adjuvant therapy.

At her follow-up visit in November 2006, she presented with a 3-month history of a progressively enlarging mass on the right chest wall. Physical examination was significant for a large mass measuring 10×7 cm on the right chest wall with a well-healed scar. CT revealed a large chest wall mass (maximum diameter=11 cm, Fig.1). Distant metastasis was excluded with a preoperative evaluation, and en bloc resection was then planned. Exploratory surgery revealed that the tumor had invaded the pectoralis major, ribs, intercostal muscle, pleura and lung. En bloc resection involving the pectoralis major, ribs four to seven, partial lung (middle and lower lobe) was performed. The chest wall was reconstructed with Marlex mesh and a latissimus flap. Pathological examination of the specimen revealed a LGMS involving skeletal muscle, ribs, pleura, and lung. Immunohistochemically, the tumor was positive for smooth muscle actin, desmin, CD34, CD99 and actin, but negative for cytokeratin, epithelial membrane antigen, fibronectin, and HBME1(Fig.2). The patient received no postoperative treatment and felt well 9 months after the second operation.

Discussion

LGMS is a rare soft tissue sarcoma and has generally been ac-
cepted as a unique tumor only in recent years\cite{1}. There have been 47 published cases summarized in a literature review\cite{1}. Among these cases, up to one third have a predilection for the soft tissue of the head and neck including the tongue, oral cavity and face. Others are located in extremities, trunk, retroperitoneum, bone, and breast. Primary sarcomas of the chest wall such as fibrosarcoma, liposarcoma, neurofibrosarcoma, dermatofibrosarcoma, malignant fibrous histiocytoma, and leiomyosarcoma are the most common tumor types. Yet there have been very few cases reported of LGMS arising in the chest wall that recurred locally.

The diagnosis of LGMS is usually made on clinical and pathological grounds, and the latter consist of morphology, immunohistochemistry and/or ultrastructure\cite{2,3}. Clinically, the tumors can recur locally and infrequently metastasize\cite{2}. Local recurrence in this case was documented after 34 months from the diagnosis of the primary lesion, which is consistent with the biological behaviour of LGMS. Morphologically, the tumors are composed mostly of spindle-shaped cells with plump or ovoid nuclei and eosinophilic cytoplasm. Mitotic activity is variable, but abnormal mitotic figures are typically absent. Hemorrhage and necrosis are rare\cite{2}. The clinical and morphological features of our case served to exclude the diagnosis of a benign myofibroblastic disease.

Immunohistochemically, LGMS stains positively for at least one myoid marker. Frequently, this tumor is positive for a combination of several markers\cite{2,3}. As shown in our case, the tumor was positive for SMA, actin and desmin. Immunohistochemistry is especially important to differentiate LGMS from leiomyosarcoma, liposarcoma and fibrosarcoma. It has been suggested that the best way to diagnose LGMS is to demonstrate the ultrastructural features of spindle cells by electron microscopy\cite{3}, but this examination is not always available for general diagnostic practice. Based on histopathology and immunohistochemistry, LGMS diagnosis can be made\cite{3}. According to the literature, nearly half of the reported cases (48.9%) were not examined by electron microscopy. Even in those cases in which electron microscopy was applied, not all had ultrastructural confirmation, and some had inconclusive ultrastructure\cite{2}.

The primary lesion was interpreted as low-grade fibroblastic/myofibroblastic sarcoma, and in comparison with the histopathological findings, between the primary and the recurrent lesion, remarkable histopathological overlap did exist. Although there are controversial issues surrounding the concept of a myofibroblastic sarcoma\cite{2-4}, myofibroblasts do resemble fibroblasts, and it remains difficult to verify myofibroblastic differentiation without electron microscopy. There has been disagreement on the ultrastructure definition of the myofibroblast: a sarcoma fulfills criteria for a myofibroblastic sarcoma when it is composed primarily of myofibroblasts, but the relative proportion of myofibroblasts to fibroblasts has not been defined. All these features show the close