Management of Primary Chest Wall Tumors: 14 Years' Clinical Experience

Po-Kuei Hsu[1], Han-Shui Hsu[1]*, Hui-Chen Lee[2], Chih-Cheng Hsieh[1], Yu-Chung Wu[1], Liang-Shun Wang[1], Biing-Shiun Huang[1], Wen-Hu Hsu[1], Min-Hsiung Huang[1]

Divisions of [1]Thoracic Surgery and [2]Experimental Surgery, Department of Surgery, Taipei Veterans General Hospital and National Yang-Ming University School of Medicine, Taipei, Taiwan, R.O.C.

Background: Primary chest wall tumor is rare but it encompasses tumors of various origins. We analyzed our experience with primary chest wall tumors with emphasis on its demographic presentation and management.

Methods: From 1991 to 2004, 62 patients with the diagnosis of primary chest wall tumors were enrolled. Lipoma, chest wall metastasis, direct invasion from nearby malignancy, infection, and inflammation of chest wall were excluded. The clinical features, management, and the outcome of these patients were retrospectively reviewed.

Results: There were 37 males and 25 females. Malignant and benign tumors were equally distributed. Chondrosarcoma and lymphoma were the 2 most common types of malignant chest wall tumors. The most common clinical symptoms were palpable mass (54.8%) and pain (40.3%). Nine of 31 patients (29.0%) with benign chest wall tumors were free of symptoms whereas patients with malignant chest wall tumors were all symptomatic ($p = 0.002$). A definite diagnosis was obtained in 21 of 26 patients (80.7%) who received nonexcision biopsy. All patients with primary chest wall tumors, except 6 who had medical treatment only, underwent surgical resection. Patients with malignant chest wall tumors were older than those with benign tumors ($p < 0.001$). The mean largest diameter of tumors was also larger in malignant tumors than in benign tumors ($p = 0.04$).

Conclusion: Patients with primary malignant chest wall neoplasm were older than those with benign tumors. The mean size of malignant tumors was larger than that of benign tumors. Adequate surgical resection remains the treatment of choice for patients with primary chest wall tumors. Nonexcision biopsy should be reserved for patients with a past history of malignancy, suspicion of hematologic disease, and with high operative risk. For patients with isolated chest wall lymphoma, surgical resection followed by chemotherapy can be considered to obtain a better outcome. [*J Chin Med Assoc* 2006;69(8):377–382]

Key Words: chest wall tumor, metastatic chest wall tumor

Introduction

Primary chest wall tumor is rare and represents about 5% of all thoracic neoplasms.[1–3] It encompasses tumors of various origins, including bone and cartilage, soft tissue such as muscle, vessel, nerve, and even some hematologic diseases.[1,4,5] Only 8% of primary bone tumors occur in the chest wall.[6] The clinical presentation of primary chest wall tumor is nonspecific. It is sometimes difficult to make an accurate diagnosis before histologic examination. Although the technique of chest wall reconstruction has evolved and aggressive resection is now safe and reliable,[4,5,7–9] wide resection is not applicable to all kinds of primary chest wall tumors. The dilemma met in precise preoperative diagnosis complicates the treatment planning. Here, we retrospectively reviewed our experience in managing 62 patients with primary chest wall tumors from 1991 to 2004. This study puts emphasis on the demographic presentation and the approaches in the management of primary chest wall tumors.

*Correspondence to: Dr Han-Shui Hsu, Division of Thoracic Surgery, Department of Surgery, Taipei Veterans General Hospital, 201, Section 2, Shih-Pai Road, Taipei 112, Taiwan, R.O.C.
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Methods

From 1991 to 2004, 62 patients with the diagnosis of primary chest wall tumors were enrolled in this study. The definition of primary chest wall tumors is neoplasms arising from structures that support the thorax, including bone, cartilage, and associated soft tissue. Patients with chest wall lipoma, metastatic chest wall lesions, chest wall invasion from nearby malignancy (breast cancer, lung cancer, mesothelioma), chest wall infection (e.g. cold abscess by *Mycobacterium tuberculosis*), and chest wall inflammation (e.g. Tietze's syndrome) were excluded. The medical charts of 62 patients were retrospectively reviewed. Clinical data including history, gender, age, clinical symptoms, imaging findings (chest X-ray [CXR], computed tomography [CT], bone scan), tumor size, location, hospital stay, operation methods, pathology reports, and outcome were collected. Pathologic diagnosis was made by nonexcision biopsy (fine needle aspiration and/or incision biopsy) or histologic examination from surgical specimens. Origins of neoplasms were classified into osseous and cartilaginous, soft tissue (e.g. muscle, vessel, peripheral nerve, fibrous tissue), and hematologic (e.g. lymphoma, plasmacytoma). The follow-up period was determined from the date of admission to the latest date of medical record or telephone contact. Outcome was recorded as with or without recurrence or metastasis. Two patients were lost to follow-up and were not included in the outcome analysis. The mean follow-up period was 42 months (range, 1–152 months) among the remaining 60 patients. The follow-up rate in this study was 96.8%.

Table 1. Demographic data of 62 patients with primary chest wall tumor

	n (%)
Benign/malignant	31/31
Sex (male/female)	37/25
Age (yr)	51.18 ± 19.97
Tumor size (cm)	7.84 ± 5.35
Symptoms	
None	9 (14.5)
Pain	25 (40.3)
Mass	34 (54.8)
Cough	3 (4.8)
Dyspnea	1 (1.6)
Neurologic	3 (4.8)
Treatment	
Medical treatment	6 (9.7)
Surgical excision	30 (90.3)

Continuous variables were expressed as mean ± SD. Statistical analysis was performed using Fisher's exact test and χ^2 test for categorical variables, and *t* and ANOVA tests for continuous variable comparison. A value of $p < 0.05$ was considered statistically significant.

Results

Tables 1 and 2 show the demographic data and pathologic diagnosis of 62 patients with primary chest wall tumors. Their mean age was 51.18 ± 19.97 years (range, 11–84 years). There were 37 males and 25 females. Thirty-one patients had benign tumors

Table 2. Pathologic diagnosis of 62 patients with primary chest wall tumor

	n
Benign	
Bone and cartilaginous	
Chondroma	3
Chondroblastoma	1
Chondromatous hamartoma	1
Chondromyxoid fibroma	1
Fibrous dysplasia	2
Giant cell tumor	1
Oseochondroma	3
Soft tissue	
Angiolipoma	1
Cavernous lymphangioma	1
Fibrolipoma	1
Fibrous tumor	4
Ganglioneuroma	2
Hemangioma	1
Leiomyoma	2
Schwannoma	5
Neurofibroma	2
Malignant	
Bone and cartilaginous	
Chondrosarcoma	7
Osteosarcoma	2
Soft tissue	
Dermatofibrosarcoma protuberance	3
Epithelioid angiosarcoma	1
Hemangiopericytoma	1
Leiomyosarcoma	1
Liposarcoma	1
Malignant fibrous histiocytoma	2
Neuroendocrine tumor	1
Sarcomatoid carcinoma	1
Hematologic disease	
Lymphoma	9
Plasmacytoma	2

and 31 had malignant tumors. The mean tumor diameter at the greatest dimension was 7.84 ± 5.35 cm. The most common clinical symptoms were palpable mass (54.8%) and pain (40.3%). In benign tumors, 12 neoplasms were of bone and cartilaginous origin and 19 were of soft tissue origin. In primary malignant tumors, 9 neoplasms were of bone and cartilaginous origin, 11 of soft tissue origin, and 11 of hematologic disease origin. Twenty-six patients received nonexcision biopsy for the initial presentation. Definite diagnosis was obtained in 21 of these 26 patients (80.7%). Five patients with negative pathologic result subsequently underwent surgical excision. Thirty-six patients received surgical intervention initially for the removal of their chest wall tumors. Among 56 patients who received surgical treatment, the chest wall defect was too large to be repaired by muscle flap in 12 patients. Mesh repair was performed in 12 patients. Surgical margin was free of tumor in all patients who underwent excision. The management algorithm of our patients in this study is presented in Figure 1.

Table 3 shows the data comparison between patients with benign and malignant primary chest wall tumors. Patients with malignant chest wall tumors were significantly older than those with benign tumors (59.94 ± 17.05 years *vs.* 42.42 ± 19.01 years, $p < 0.001$). Mean tumor diameter was also significantly greater in malignant tumors compared to benign tumors (9.17 ± 5.36 cm *vs.* 6.52 ± 5.06 cm, $p = 0.04$). Nine of 31 patients (29.0%) with benign chest wall tumor were free of symptoms whereas patients with malignant chest wall tumors were all symptomatic ($p = 0.002$). Table 4 shows the data comparison among patients with different origins of chest wall tumor. Patients with hematologic origin of chest wall tumor were more male predominant than those with tumors of bone or soft tissue origins ($p = 0.03$). Patients with tumors of hematologic disease origin (66.36 ± 14.79 years) were also older than those with tumors of bone and cartilage origin (50.05 ± 21.50 years), and soft tissue origin (46.40 ± 18.23 years, $p = 0.01$). Presence of symptoms and mean tumor size were not different among the 3 tumor origins. Tumor recurrence or metastasis was found in 10 patients, including 5 chondrosarcomas, 1 lymphoma, 1 plasmacytoma, 1 leiomyosarcoma, 1 osteosarcoma, and 1 sarcomatoid carcinoma. All recurrences or metastases developed in

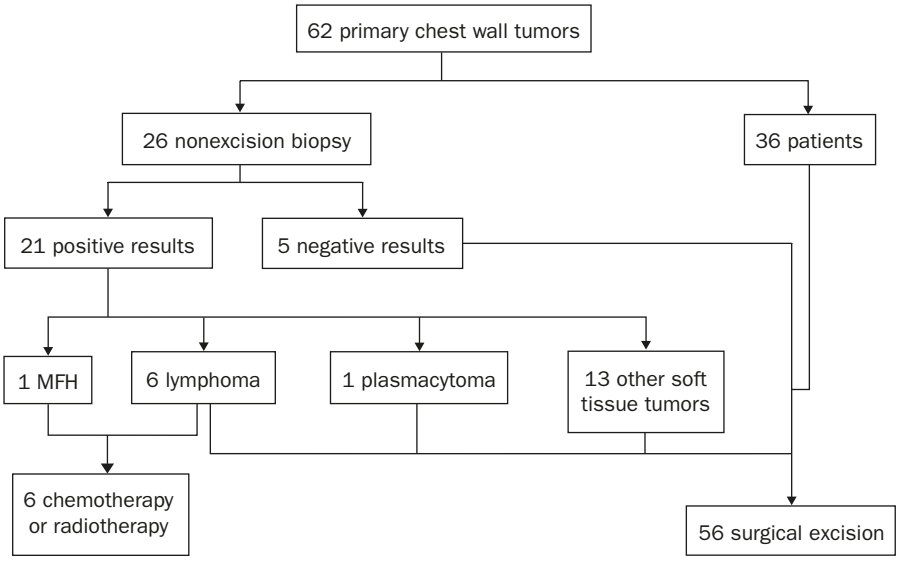

Figure 1. Management algorithm of 62 patients with primary chest wall tumors. MFH = malignant fibrous histiocytoma.

Table 3. Comparison between patients with benign and malignant chest wall tumors

	Benign ($n = 31$)	Malignant ($n = 31$)	p
Sex (male/female)	16/15	21/10	0.3
Age (yr)	42.42 ± 19.01	59.94 ± 17.05	< 0.001*
Presence of symptoms	22	31	0.002*
Presence of pain	10	15	0.3
Tumor size (cm)	6.52 ± 5.06	9.17 ± 5.36	0.04*

*$p < 0.05$. Fisher's exact test for sex, presence of symptoms, and presence of pain comparison. Student's t test for age and size comparison.

Table 4. Comparison among patients with different origins of chest wall tumors

	Bone and cartilage ($n=21$)	Soft tissue ($n=30$)	Hematologic disease ($n=11$)	p
Sex (male/female)	13/8	14/16	10/1	0.03
Age (yr)	50.05 ± 21.50	46.40 ± 18.23	66.36 ± 14.79	0.01*
Benign/malignant	12/9	20/10	0/11	0.000*
Tumor size (cm)	7.17 ± 4.95	7.82 ± 5.79	9.16 ± 5.05	0.61
Presence of symptoms	19	23	11	0.12
Presence of pain	6	14	5	0.42

*$p < 0.05$. χ^2 exact test for sex, benign or malignant, presence of symptoms, and presence of pain comparison. ANOVA test for age and size comparison. Patients in hematologic origin group are significantly older than in bone/cartilage origin group ($p = 0.024$) and soft tissue origin group ($p = 0.004$) by post hoc test.

the first 2 years after treatment and 70% were within the first postoperative year. Of 10 patients, 3 died of disease-related complications (at 3, 9, and 25 months after diagnosis), 3 died of unrelated conditions, and 4 were still alive with follow-up periods of 2, 14, 70, and 159 months, respectively.

Chondrosarcoma and lymphoma were the 2 most common types of malignant chest wall tumors in our series. All patients with chest wall chondrosarcoma received surgical excision. Recurrence developed in 5 of 7 patients at 3, 5, 6, 7, and 20 months after operation. Four of 5 patients received repeated surgical excision and 3 had radiotherapy. The other patient with superior vena cava (SVC) syndrome died due to respiratory failure. This was the only disease-related mortality in the chondrosarcoma group in our series. All cases of chest wall lymphoma were of diffuse large B cell type in our study. Five of 9 patients with chest wall lymphoma received chemotherapy. The regimens included cyclophosphamide, doxorubicin, vincristine, and prednisolone (CHOP), cyclophosphamide, vincristine, and prednisolone (COP), and cyclophosphamide, epirubicin, vincristine, and prednisolone (CEOP). Two of 5 patients had other organ involvement. Regression of chest wall tumors after chemotherapy was noted in 4 of them. Two of 4 patients died at 7 and 10 months after diagnosis due to pneumonia and sepsis, respectively. Another patient who had recurrence of lymphoma received further chemotherapy and radiotherapy for multiple bone metastasis subsequently. He died of pneumonia at 49 months after diagnosis. The other patient with isolated chest wall lymphoma improved after chemotherapy and is currently free of disease. Only 1 patient with chest wall lymphoma was refractory to chemotherapy. This patient died at 7 months after diagnosis because of chemotherapy-related complications. The other 4 patients with isolated chest wall lymphoma received surgical excision followed by adjuvant chemotherapy, and they were without recurrence at 36, 38, 52, and 111 months after treatment.

Discussion

Primary tumors of the chest wall are uncommon; therefore, the experience in managing this kind of tumor is limited in each medical unit. In clinical practice, it is difficult to diagnose from clinical features, radiologic examination, or even aspiration biopsy. Most patients with primary chest wall tumor received excision biopsy or radical resection for the chest wall tumor before the diagnosis was obtained.[1] Of the 157 patients who presented with chest wall mass, only 62 patients had primary chest wall tumor and were included in our study. It has been reported that primary chest wall tumors occur mostly in the 5th and 6th decades of life with equal gender distribution and possibility of benign or malignancy.[1,2,5,7,10–14] The mean age was 51.18 years in our study and patients with malignant chest wall tumors were significantly older than those with benign tumors. The male to female ratio was about 1:1, although slight male predominance was found in the malignant tumor group. The ratio of benign to malignant primary chest wall tumors was 1:1 in our series. The most frequent symptoms in patients with primary chest wall tumors were palpable mass and pain.[8] Neurologic symptoms such as muscle weakness and brachial plexus neuropathy have also been reported.[12,13] In our study, 29.0% of patients with benign primary chest wall tumors were asymptomatic, whereas all patients with malignant neoplasm were symptomatic. Among the patients with primary chest wall tumors of bone and cartilaginous origin, rib was the most common source.[1,5,10] In our series, 5 primary chest wall tumors arose from the sternum. Four of them (80%) were malignant. This result is similar to previous reports in the literature that neoplasms from the sternum, though rare, are usually malignant.[6,8]

Imaging studies, including CXR and CT, are useful in determining the extent of tumor invasion, surgical treatment planning, and follow-up evaluation of patients with chest wall tumors. However, imaging

features of chest wall tumors are nonspecific.[9,15] Soft tissue mass, bone destruction, calcification, and bone deformity are frequently described, but are not diagnostic of malignancy. Although some typical features can be linked to certain types of neoplasms, such as the presence of phleboliths and vascular enhancement in hemangioma, target-like appearance on magnetic resonance images (MRI) in neurofibroma, well-defined cortical and medullary continuity in osteochondroma, fusiform expansion and ground glass matrix in fibrous dysplasia, expansile osteolytic lesion with fluid–fluid levels in giant cell tumors, presence of chondroid or osteoid matrix in osteosarcoma, and diffuse osteolytic change with a soft tissue rib mass in multiple myeloma,[16,17] it is still difficult to make the diagnosis of chest wall tumors or to differentiate benign from malignant chest wall tumors from imaging findings.

Osteochondroma, chondroma, and fibrous dysplasia constitute 60–70% of all benign chest wall tumors, whereas chondrosarcoma is the most common primary malignant chest wall tumor.[3,12] In our study, chondroma, osteochondroma, and schwannoma were the most common benign chest wall tumors. All these patients received surgical excision and recovered well. Chondrosarcoma and chest wall lymphoma were the most frequent malignant chest wall tumors in our series. All 7 patients with chondrosarcoma received surgical excision. The recurrence rate after excision was 62.5%. One patient with recurrence developed pleural effusion and SVC syndrome and died of disease. Briccoli et al[15] reported 16 cases with chondrosarcoma and found recurrence in 6 of 16 patients after wide excision and that it may occur as late as 68 months after operation. In our study, 1 patient experienced tumor recurrence at 5, 17, 23, 47, and 71 months after the first excision. Repeated excisions and radiotherapy were carried out. This patient is still alive and under regular follow-up. Consistent to other reports, our experience indicates that patients with chest wall chondrosarcoma are at high risk of tumor recurrence.[10,15] For chondrosarcoma, 2 peak periods of prevalence have been identified—at less than 20 years of age and at more than 50 years of age. Most frequently, they are found along the upper 5 ribs. Bone destruction, irregular contours, and intratumoral mineralization are characteristics on CXR. CT scan is more sensitive for the delineation of chondroid matrix calcifications. MRI usually shows heterogeneous contrast enhancement.[17] Wide excision and careful postoperative evaluation are mandatory for these patients.

The role of fine needle aspiration or nonexcision biopsy for the diagnosis of primary chest wall tumor is controversial.[5,9,12,13] Caution is warranted when using fine needle biopsy as a diagnostic modality since diagnosis by cytologic specimen is sometimes unsatisfactory.[18] Wrong diagnosis due to inadequate tissue specimen may even lead to less than optimum treatment and worse prognosis.[19] Meanwhile, the risk of tumor implantation makes fine needle biopsy an unpopular tool in the diagnosis of primary chest wall tumors.[4,7] Some authors suggest that all patients with primary chest wall tumors should receive at least excision biopsy, whereas those with high suspicion of malignancy should receive wide radical resection or subsequent resection for safe margin.[1,3–5,7,9,12–14] Nonexcision biopsy should be reserved for those with suspicion of metastatic or hematologic disease, in whom aggressive surgical resection is less beneficial.[3,5,7–9,11,20,21] In our studies, 26 patients received nonexcision biopsy for initial diagnosis. Definite diagnosis was obtained in 21 of these 26 patients.

Whether or not patients with primary chest wall lymphoma should receive surgical resection is controversial. Lymphoma with chest wall involvement is not uncommon. However, isolated primary chest wall lymphoma is not frequently seen.[22,23] In Ryan et al's series,[7] 2 patients with chest wall lymphoma were found during a 5-year period. One of them received surgical resection and was free of disease after 49 months. In our study, the clinical outcome was poor in 5 of 9 patients with chest wall lymphoma who received chemotherapy after diagnosis, which might be related to multiple organ involvement and chemotherapy toxicity. The other 4 patients with solitary chest wall lymphoma received surgical excision followed by adjuvant chemotherapy. These patients remained free of disease after a mean follow-up period of 84 months. Although the primary treatment of choice for lymphoma with or without chest wall involvement is chemotherapy, surgical intervention can be considered in some selected patients, especially those with solitary chest wall lymphoma without other organ involvement.

In summary, our limited experience in the management of primary chest wall tumors showed that patients with malignant neoplasms were older than those with benign tumors. The mean size of malignant tumors was larger than that of benign tumors. Absence of symptoms was more frequently seen in those with benign than malignant primary chest wall tumors. Wide excision and careful postoperative evaluation are mandatory for patients with chest wall chondrosarcoma who are at high risk of tumor recurrence after surgical resection. Adequate surgical resection remains the treatment of choice for patients with primary chest wall tumors. Nonexcision biopsy should be

reserved for patients with a past history of malignancy, suspicion of hematologic disease, and with high operative risk. For patients with isolated chest wall lymphoma, surgical resection followed by chemotherapy can be considered to obtain a better outcome.

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