Tzu Chi Medical Journal 28 (2016) 121–123

Contents lists available at ScienceDirect

Tzu Chi Medical Journal

journal homepage: www.tzuchimedjnl.com

Case Report

Insidious malignant triton tumor of the chest wall with late flare-up

Jeng-Yuan Wu [a, b, *], Lai-Fa Sheu [b, c], Chao-Yuan Yao [d]

[a] Department of Thoracic Surgery, Taichung Tzu Chi Hospital, Buddhist Tzu Chi Medical Foundation, Taichung, Taiwan
[b] School of Medicine, Tzu Chi University, Hualien, Taiwan
[c] Department of Pathology, Taichung Tzu Chi Hospital, Buddhist Tzu Chi Medical Foundation, Taichung, Taiwan
[d] Department of Hematology–Oncology, Taichung Tzu Chi Hospital, Buddhist Tzu Chi Medical Foundation, Taichung, Taiwan

ARTICLE INFO

Article history:
Received 27 August 2014
Received in revised form
22 October 2014
Accepted 5 November 2014
Available online 19 December 2014

Keywords:
Chest wall tumor
Malignant peripheral nerve sheath tumor
Malignant triton tumor

ABSTRACT

Malignant triton tumor is a rare malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation. Most of these tumors are located in the head, neck, and extremities, and about half of cases are associated with neurofibromatosis type 1 featuring cafe-au-lait spots or cutaneous neurofibromas. We present a 76-year-old man with an insidious chest wall tumor with late progressive painful enlargement and pleural and pulmonary involvement. Complete resection of the affected thoracic wall as well as single separate lesions in the parietal pleura and left upper lung was carried out. The pathological examination confirmed that it was a malignant triton tumor. The patient received adjuvant chemoradiotherapy but eventually succumbed to disease relapse and distant metastases 6 months after the surgery.

1. Introduction

Malignant triton tumor (MTT) is rare and highly aggressive, and tends to develop early local recurrence and distant metastases. Immunohistochemistry (IHC) showing malignant schwannoma cells coexisting with rhabdomyoblasts confirms the diagnosis. Aggressive surgical management is of crucial importance in the management of MTT, with adjuvant chemotherapy and/or radiotherapy being effective in sporadic cases. Here we report an unusual presentation of an MTT in the chest wall of an elderly man who had no skin lesions or family history of neurofibromatosis. The case is unique in terms of its slow disease progression and late flare-up of the tumor, leading to an initial clinical impression that did not favor MTT. The patient died after *en bloc* resection followed by adjuvant chemoradiotherapy because of ongoing disease exacerbation with poor response to multimodality treatment.

2. Case Report

A 76-year-old otherwise healthy man presented with a large, firm mass on his left chest wall for more than 20 years. It had enlarged progressively over the past 2 years and chest pain developed 1 month prior to admission. The physical examination revealed an ovoid 7 cm × 4.5 cm bony tumor beneath the pectoralis major muscle. Chest computed tomography (CT) revealed a heterogeneous tumor occupying the chest wall including the fourth rib (Fig. 1A), a single small nodular lesion over the left upper lung (Fig. 1B) and right lower lung, and focal parietal pleural thickening in the left upper thorax. A bone scan showed Technetium-99m methylene diphosphonate (Tc-99m MDP)-avid lesions of an undetermined nature in the left fourth rib and right upper femur. He received CT-guided fine-needle biopsy of the chest wall tumor that pathology reported as fibromatosis. The patient underwent wide excision of the affected chest wall with synthetic mesh replacement as well as resection of the nodules in the pleura and left upper lobe. Histopathology displayed hypercellularity, cells with spindle-shaped nuclei arrayed in a fascicular pattern, nuclear pleomorphism, brisk mitotic activity, and areas of necrosis. There were many rhabdomyoblasts with eosinophilic cytoplasm (Fig. 2A). IHC demonstrated positivity of the tumor cells for anti-S100 (Fig. 2B) and antivimentin stains (Fig. 2C), but CK7, SMA, and TTF-1

Conflict of interest: none.

* Corresponding author. Department of Thoracic Surgery, Taichung Tzu Chi Hospital, Buddhist Tzu Chi Medical Foundation, 88, Section 1, Fongsing Road, Tanzih District, Taichung, Taiwan. Tel.: +886 4 36060666x3030; fax: +886 4 36065928.
E-mail address: jyuan.wu@tzuchi.com.tw (J.-Y. Wu).

http://dx.doi.org/10.1016/j.tcmj.2014.11.001

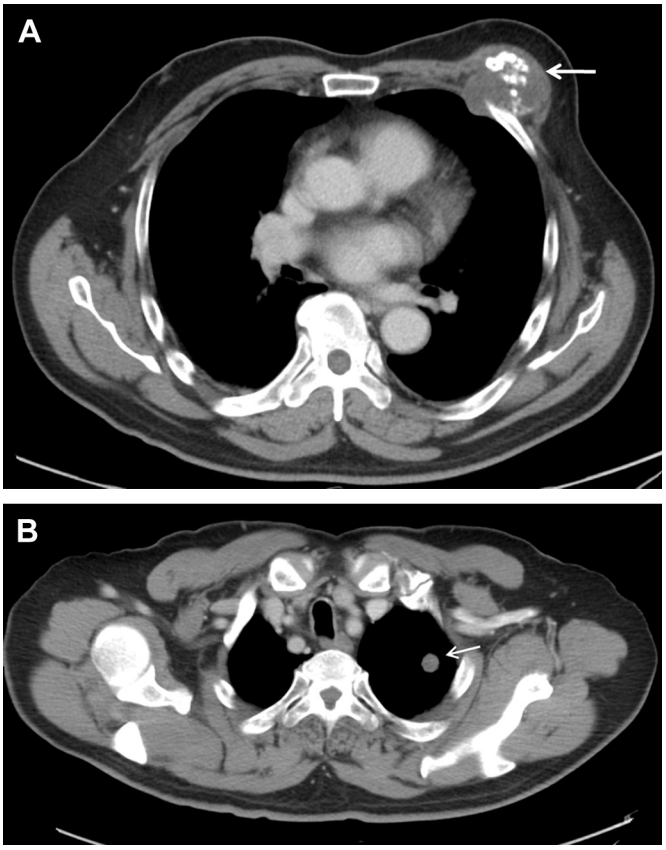

Fig. 1. Chest computed tomography (CT) scan demonstrating (A) a heterogeneous soft tissue mass with calcification (arrow) involving the anterior segment of the left fourth rib and (B) a nodular lesion about 2 cm in diameter (arrow) in the left upper lobe.

immunostains were negative. Both morphological findings and immunophenotypes were compatible with malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation, which confirmed the diagnosis of MTT. Postoperatively, the patient received three cycles of chemotherapy with cisplatin and etoposide and concurrent radiotherapy to the involved thoracic wall, at a total dose of 7000 cGy in 35 fractions. Nevertheless, local relapse and lung-to-lung, nodal, and spinal metastases occurred subsequently, and he died of pneumonia and respiratory failure in relation to adjuvant chemoradiotherapy 6 months after the operation.

3. Discussion

MTT is a very rare but highly aggressive subtype of malignant peripheral nerve sheath tumor with rhabdomyoblastic differentiation. About one-third of MTTs are located in the head and neck regions [1]. The mean age of patients is about 30 years with an equal sex distribution. MTT coexists with neurofibromatosis type 1 (NF-1) in 44–69% of cases, but can also occur sporadically or after radiotherapy. When associated with NF-1, MTT tends to occur at a young age in males. Clinical symptoms are nonspecific and are generally related to a rapidly enlarging mass that invades or imposes pressure on adjacent structures. Image studies usually delineate a picture of soft tissue sarcoma. Pathological examination and IHC are essential to establish the diagnosis. The outcome of patients is very poor with 5-year survival rates of 11–15% and high rates of metastases (48%) and local recurrence (43%). Complete surgical resection is the most effective treatment, and adjuvant therapy may be of value in individual patients [2–5]. The location of the tumor is also considered a key factor in the prognosis; those in the extremities, head, and neck have a better prognosis than those in the retroperitoneum, buttock, and trunk [3]. In our elderly patient, who presented without NF-1, the rare location and bizarre behavior of the chest wall MTT with its insidious course, and the

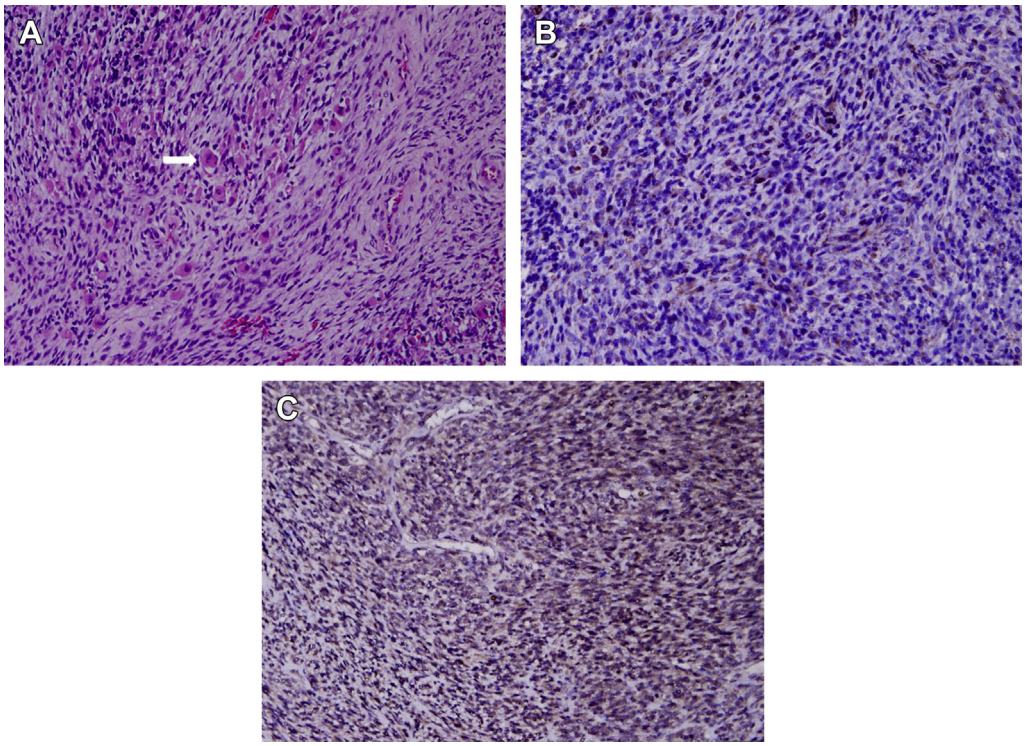

Fig. 2. Pathology showing (A) numerous pleomorphic rhabdomyoblasts containing eosinophilic cytoplasm (arrow) embedded in spindle cells with a fascicular growth pattern in a malignant triton tumor (hematoxylin and eosin, ×300). (B) Anti-S100 (×200) and (C) antivimentin (×200) immunostains of the tumor cells are positive.

late development of chest pain because of unexpectedly rapid growth and rib invasion, complicated by pleural and pulmonary involvement, are seldom seen with this tumor. We believe that the long-standing, asymptomatic chest wall lesion was possibly precancerous and the late flare-up accelerated its malignant clinical course.

The differential diagnosis of MTT consists of a large variety of neoplasms such as rhabdomyosarcoma, leiomyosarcoma, fibrosarcoma, malignant fibrous histiocytoma, monophasic synovial sarcoma, ectomesenchymoma, neurofibroma, benign triton tumor, and hemangiopericytoma [4]. Chest CT in our patient showed mainly a chest wall soft tissue tumor containing conglomerated, amorphous calcifications of various sizes coupled with eccentric calcification of lamellar distribution, a particular pattern not specified in those MTTs reported in the literature. Comparable features of scattered calcifications with sunburst or lamellated periosteal reactions are associated with osteosarcoma or chondrosarcoma, but infrequently with other sarcomatous lesions. Preoperative diagnostic procedures such as sonography and CT-guided fine-needle aspiration or biopsy are often used, as in this case. However, needle biopsy and incisional biopsy are not reliable enough to establish a conclusive result in such circumstances [4]. Microscopic examination along with IHC indicating rhabdomyoblasts among malignant Schwann cells in a tumor arising from a peripheral nerve verified by immunostaining with S-100 protein is essential for an accurate diagnosis of MTT [6]. The course of illness and image findings in our patient indicated a likely diagnosis of chest wall osteosarcoma or chondrosarcoma; however, characteristic histopathological findings as well as positivity of the resected specimen to anti-S100 and antivimentin stains confirmed the diagnosis.

Treatment strategies for MTT include surgical resection, neoadjuvant and adjuvant chemotherapy, and irradiation. Complete tumor resection appears to be associated with an improved chance of survival, decreased rates of local recurrence and metastasis, and a better response to adjuvant therapies than that in patients without resection [2]. Although the role of adjuvant therapy is not well defined and no treatment guidelines have been established because of the rarity of MTT, radical excision with adjuvant radiotherapy may achieve a satisfactory outcome [1,7]. In our patient, aggressive management was undertaken, including wide excision of the chest wall tumor, and lung and pleural nodules and postoperative chemoradiotherapy. Although ensuing distal metastases led to a fatal outcome, we consider the operation itself was still applicable to relieve symptoms primarily caused by the enlarging chest wall mass. In fact, MTT has a poor prognosis owing to its aggressive biological behavior. As in this case, most patients die within months even after multimodal therapy because of disease progression and treatment-related events.

MTT should be regarded as a distinct clinical entity associated with a high incidence of local recurrence and distant metastases. IHC showing nerve sheath differentiation with rhabdomyoblastic cells confirms the diagnosis. Early diagnosis and radical excision coupled with adjuvant chemoradiation seem to yield a better result than conservative management. However, the therapeutic plan should be individualized, taking into account the location and size of the primary tumor, curative or palliative intent, and the patient's performance status. Furthermore, rapid clinical deterioration and death often occur despite multidisciplinary management.

References

[1] Tian L, Shang HT, Bilal S, Li YP, Feng ZH, Lei DL, et al. Treatment of malignant triton tumor in zygomatic region. J Craniofac Surg 2012;23:e265–8.
[2] Li Z, Xiang J, Yan S, Gao F, Zheng S. Malignant triton tumor of the retroperitoneum: a case report and review of the literature. World J Surg Oncol 2012;10:96–101.
[3] Yakulis R, Manack L, Murphy Jr AI. Postradiation malignant triton tumor. A case report and review of the literature. Arch Pathol Lab Med 1996;120:541–8.
[4] Kamperis E, Barbetakis N, Asteriou C, Kleontas A, Christoforidou V. Malignant triton tumor of the chest wall invading the lung. A case report and literature review. Hippokratia 2013;17:277–80.
[5] McConnell YJ, Giacomantonio CA. Malignant triton tumors—complete surgical resection and adjuvant radiotherapy associated with improved survival. J Surg Oncol 2012;106:51–6.
[6] Kiryu H, Urabe H. Malignant Triton tumor. A case with protean histopathological patterns. Am J Dermatopathol 1992;14:255–62.
[7] Lang-Lazdunski L, Pons F, Jancovici R. Malignant “Triton” tumor of the posterior mediastinum: prolonged survival after staged resection. Ann Thorac Surg 2003;75:1645–8.