

Post Radiation Inflammatory Malignant Fibrous Histiocytoma Arising from the Chest Wall

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A 59-year-old man who underwent radiation therapy (41 Gy) to the mediastinum through the anterior chest for Hodgkin's disease presented with a painful anterior chest wall tumor 18 years later. The tumor originated from the left parasternal region and was excised with the sternum. Chest wall reconstruction was performed. The tumor measured 45×45 mm and invaded the sternum. The pathologic diagnosis was malignant fibrous histiocytoma. Early and complete excision of the tumor is indicated. (Ann Thorac Cardiovasc Surg 2001; 7: 371-4)

Key words: malignant fibrous histiocytoma, sternum, chest wall tumor, chest wall reconstruction, soft tissue tumor

Introduction

Malignant fibrous histiocytoma (MFH) is one of the most common soft tissue sarcomas of adulthood, accounting for about 10% of all sarcomas.¹⁾ MFH has been reported in various organs,¹⁾ although the usual primary site is the deep musculature of the upper extremity, the lower extremity, or the retroperitoneum. The chest wall is an extremely uncommon primary site.^{2,3)} The purpose of this report is to improve the understanding of the clinical behavior and predictors of biological aggressiveness of MFH of the chest wall. A case of surgically treated post radiation inflammatory MFH arising in the anterior chest wall is reported.

Case Report

A 59-year-old man presented with a painful anterior chest wall tumor. Radiation therapy was performed for Hodgkin's disease to the cervix (54 Gy) at age 41. A gastrectomy was also performed at age 41 for gastric

cancer. Additional radiation therapy through the anterior chest wall to the mediastinum (41 Gy) and to the abdominal paraaortic region through the anterior abdominal wall (40 Gy) was performed for recurrence of Hodgkin's disease in the same year. A small intestinal perforation and panperitonitis requiring a laparotomy occurred one year later. Steroid therapy had been instituted because of a pericardial effusion due to radiation-induced pericarditis, which began at age 54. A pacemaker was implanted via the left subclavian vein because of complete atrioventricular block at age 57. At that time, he began outpatient psychiatric treatment for neurosis and insomnia. Severe dysphagia due to pseudobulbar paralysis was noted. The focus for the paralysis was not identified by the psychologists, neurologists, neurosurgeons, and otolaryngologists. Chest computed tomography (CT) revealed destruction of the sternum due to radiation-induced osteitis.

He was referred to our department and admitted for the diagnosis and treatment of chest wall and abdominal wall abscesses (Fig. 1). The anterior chest wall tumor originated from the left parasternal region. The chest wall tumor was at the area of the skin burn that was due to radiation therapy (Fig. 1B). Chest CT showed an inhomogeneous abscess with destruction of the sternum (Fig. 2). Magnetic resonance imaging (MRI) was not performed because of the presence of the pacemaker. Needle aspiration biopsy of the tumor revealed bloody pus.

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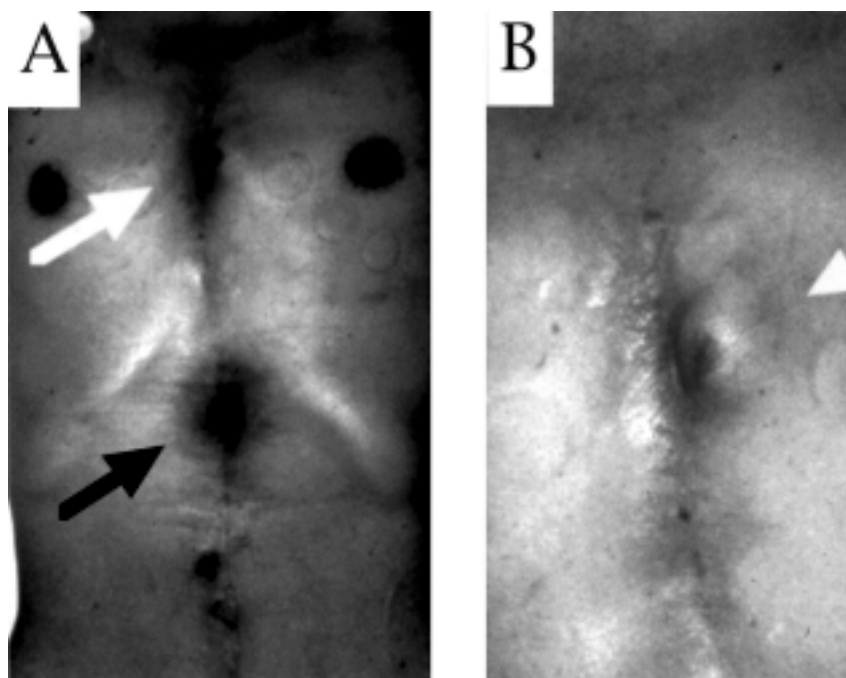


Fig. 1. Photograph showing an anterior chest wall abscess in an area previously treated with radiation (white arrow, A). An anterior abdominal wall abscess is also observed in the radiated area (black arrow, A). Magnified chest wall abscess was shown in B.

Pathologic examination of specimens from multiple needle biopsies showed no malignant features. Chest wall abscess associated with destruction of the sternum due to radiation therapy and steroid use was suspected. After anti-inflammatory therapy, the size of the abdominal wall abscess decreased, but the size of the chest wall abscess increased. Examination of the chest wall abscess with bone scintigraphy (Tc-99m-HMDP) was negative, and gallium scintigraphy (Ga-67 citrate) was positive. The tumor was so painful that the patient required morphine.

Surgical excision of the chest wall tumor was planned

because of the rapid growth and because malignancy had not been excluded. Under general anesthesia, a skin incision was made around the tumor (Figs. 3A, B). The tumor was excised in continuity with part of the sternum and the third to seventh ribs because it invaded the sternum and ribs (Fig. 3C). Three centimeters of the tumor margin was taken. Intraoperative microscopic examination of a frozen section revealed negative surgical margins. Chest wall reconstruction was performed with wire, marlex mesh sheets, and a right pectoralis major myocutaneous flap though his muscle was poor (Fig. 3D). A free skin graft was also used. Extubation was successful immediately after surgery. Spontaneous respiration was possible.

The tumor was elastic and firm, with dimensions of 45 × 45 mm. The cut surface of the tumor was solid and whitish yellow (Fig. 4A). Microscopic examination showed spindle-shaped cells, anaplasia, pleomorphism, and mitoses. Storiform (Fig. 4B) and antler-like patterns were seen. The background included inflammatory cells, which were mainly lymphocytes. The tumor cells were stained by vimentin, but not α -SMA, desmin, S100, LCA, EMA, or keratin. The pathologic diagnosis was inflammatory malignant fibrous histiocytoma. The tumor invaded the sternum. The surgical margins were negative.

The postoperative course was uneventful. However, the severe dysphagia persisted as in the preoperative period. The patient died 64 days after surgery because of severe pneumonitis associated with pseudobulbar paralysis.

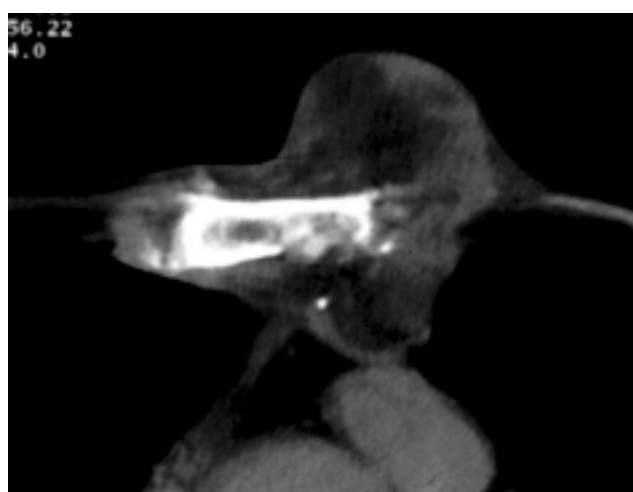


Fig. 2. Chest computed tomography showing an inhomogeneous tumor with necrotic nonenhancing regions. Destruction of the sternum is also noted.

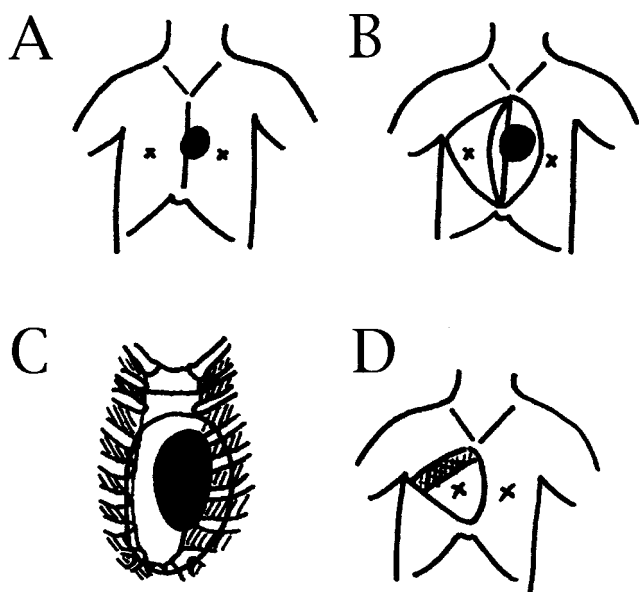


Fig. 3. Diagram of the surgical procedure showing the tumor (A). B: The skin incision surrounding the entire tumor. C: The tumor and part of the sternum were excised en bloc. D: A right pectoralis major myocutaneous flap was used for chest wall reconstruction.

Discussion

MFH is a primitive sarcoma originating in the deep soft tissues.¹⁾ It occurs most frequently in the deep fascia and skeletal muscle of the extremities and trunk.¹⁾ Although MFH is not an uncommon tumor, it rarely arises from

the chest wall.^{2,3)}

Carcinomas and deep-seated sarcomas such as MFH have been reported to occur with increased frequency in patients who were treated with radiotherapy for malignant disease.^{4,5)} Clear association has been found between the dose of radiation and the incidence of sarcoma.⁴⁾ Our patient had received radiation therapy to the mediastinum through the anterior chest wall for Hodgkin's disease 18 years prior to presentation. The relationship between the radiation therapy and the MFH was unclear pathologically but the localization was the same in this case. Cahan et al. have suggested the following criteria for a sarcoma to be considered radiation induced: 1) the sarcoma should arise in the area subjected to the radiation, 2) a latency period (in years) must exist between the time of radiation and the development of the sarcoma, and 3) the sarcoma must be diagnosed histologically.^{6,7)} The radiation induced solid tumor is developed for at least 10 years.⁵⁾ Our case fulfilled these criteria.

Although MFH is a solid tumor, it sometimes exhibits cystic or necrotic findings on CT.⁸⁾ In our case, chest CT showed abscess formation and destruction of the sternum. An MRI would have been helpful, but was not possible because of the existing pacemaker.

Since the morphology of MFH is highly variable, even within an individual tumor, diagnosis is difficult with small biopsy specimens.³⁾ A preoperative pathologic diagnosis was not achieved by needle biopsy in our case. The inflammatory form of MFH is particularly difficult

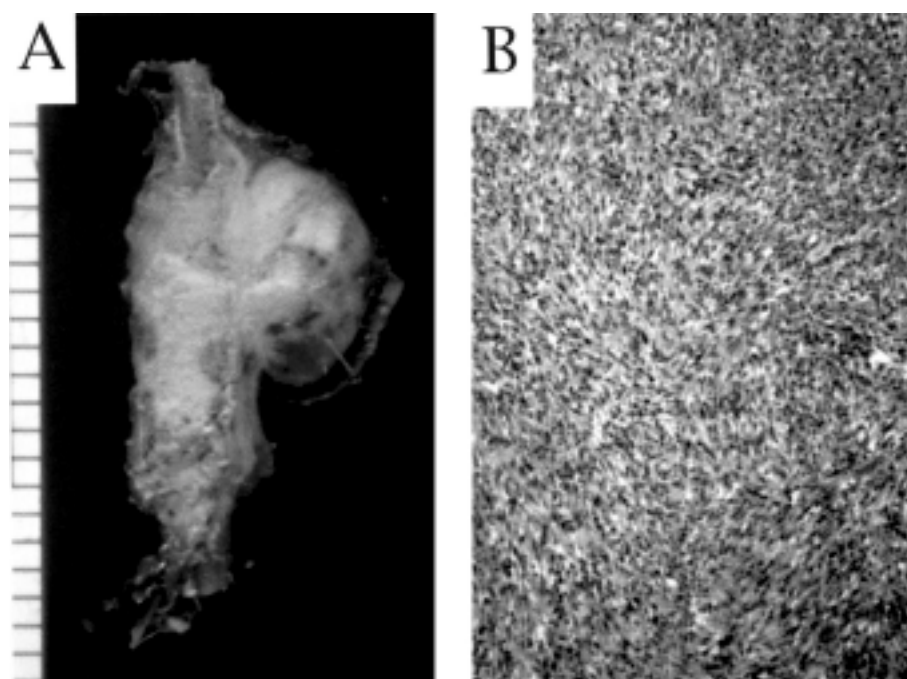


Fig. 4. Macroscopic findings of the tumor and sternum (A) and microscopic findings (hematoxylin and eosin) of the tumor (B). Note the storiform (B).

to diagnose. It is often confused with a benign inflammatory process.⁹⁾ Because of the presence of the destruction of the sternum, we selected complete surgical resection for the diagnosis and treatment.

Radical en bloc resection is required because MFH is aggressive, with a high propensity for local and distant spread.³⁾ The feasibility of en bloc resection, however, is dependent on the anatomic location of the tumor and the nature of the extent of invasion.³⁾ Many reconstructive methods have been reported after full-thickness anterior chest wall resection.¹⁰⁻¹²⁾ In our case, the sternum and ribs had to be removed with the tumor. The selection of a muscle flap was difficult. The rectus abdominis muscle was not selected because of the previous laparotomies and because the tumor invaded the intrathoracic artery. The left pectoralis major muscle was not used because of the pacemaker implantation. The right pectoralis major muscle was selected. The wire, mesh sheets, and muscle flap were tightly fixed to avoid postoperative flail of the chest wall.

MFH is characterized by a bimorphic population of fibrocytic and histiocytic cells usually arranged in a storiform pattern.¹⁾ There are many diseases which must be differentiated from MFH, including spindle carcinoma, carcinosarcoma, and inflammatory pseudotumor. Though the characteristic patterns, such as a storiform pattern and antler-like pattern, are useful, immunohistochemical staining is helpful for the diagnosis as our case.

MFH is a soft tissue tumor. However, some show intralesional calcifications,¹³⁾ and some arise in the bone.¹⁴⁾ In our case, the sternum was destroyed first, and then, the tumor arose. This was confirmed by negative bone scintigraphy. It was not histopathologically clear whether the tumor arose from the sternum and invaded the surrounding soft tissue, or arose in the soft tissue and invaded the sternum. However, it is suggested that the tumor originated from the soft tissue because of the clinical course and because there were no bony islands in the tumor.

MFH has a high propensity for local recurrence (44%) and distant metastasis (42%).¹⁵⁾ Mean survival rate of the surgically treated chest wall MFH is 23 months.²⁾ The inflammatory type is aggressive and is associated with a worse prognosis.⁹⁾ Radiation therapy for post radiation MFH is effective in some cases and is not in other cases.¹⁶⁾ The effects of chemotherapy are also controversial. Due to the high recurrence and metastasis, early detection and complete excision of the tumor is indicated.

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