Extensively Calcified Hemangioma of the Diaphragm with Increased $^{99m}$Tc-Hydroxymethylene Diphosphonate Uptake

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Abstract

A 31-year-old woman visited an outpatient clinic, because of low-grade fever and general fatigue. She was referred to our hospital and admitted for examination of an abnormal shadow which had been found on the chest radiograph. She had experienced faint right lateral chest pain several times on the deep inspirations. Chest radiography showed a mass shadow with calcification in the right lower lung field on the mediastinal side. Chest radiographic computed tomography showed a 6×6 cm tumor in the right lung field. There were low-density areas with septae inside the tumor. Bone scintigraphy showed extremely high uptake of $^{99m}$Tc-HMDP in the tumor. After surgical resection and pathological examination, we concluded that the tumor was an extensively calcified benign hemangioma of the diaphragm.

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Key words: tumor, scintigraphy, calcification

Introduction

Primary neoplasms of the diaphragm are extremely uncommon. The most common benign diaphragm tumor is lipoma followed by bronchial cyst, teratoid cyst, fibroma, angiofibroma, neurofibroma, hamartoma and neurilemoma. Primary malignant diaphragm tumors include fibrosarcoma, rhabdomyosarcoma, hemangioendothelioma, fibromyxosarcoma, myosarcoma and neurofibrosarcoma (1). However, tissue types not normally found in the diaphragm may also produce benign diaphragm tumors, which include adrenal cortical adenoma, liver cell adenoma, chondroma, hamartoma and mesothelioma (2). Benign and malignant fibrous tumors arise from the parietal pleura of the diaphragm (3).

About half of primary diaphragm tumors are benign (4) and patients with a benign diaphragm tumor usually have no complaints. These benign tumors are found on a chest radiograph or at the post-mortem examination. The diaphragm can be a target of direct and distant metastases from internal malignancies. These malignancies directly invade the diaphragm, although blood borne metastatic implants are rare (2). The symptoms of malignant diaphragm tumors are not specific, and even in patients with malignant diaphragm tumors, pre-mortem diagnosis of the diaphragm invasion is infrequent.

In the present report, we describe a case with extensively calcified hemangioma in the diaphragm showing increased uptake in the bone scintigraphy.

Case Report

A 31-year-old woman visited an outpatient clinic, because of low-grade fever and general fatigue. Although the doctor of the clinic considered the symptoms not specific, he found an abnormal shadow in the right lung on the chest radiogram. She experienced faint right lateral chest pain several times on the deep inspirations. She was referred and admitted to Asahikawa Medical College Hospital in June 1994. On admission, her respiratory rate was 14/minute, body temperature was 36.5°C, blood pressure was 108/68 mmHg and heart rate was 70/minute. Heart sounds and breath sounds were normal. No tumors were found on abdominal palpation. Blood examinations showed no abnormalities, except the white blood cell count was 8,650/μl. Serum tumor maker levels including carcinoembryonic antigen, CA 19-9, α-fetoprotein, human chorionic gonadotropin and PIVKA-II were normal. Serum C terminus and mid portion of parathyroid hormone (PTH) levels were 0.32 ng/ml and 0.42 ng/ml, respectively. Serum total calcium level was 9.0 mg/ml and phosphate level was 3.1 mg/ml. The surface of her body, however, was entirely lacking in hair. She had no exceptional family history or personal history except this hair loss.

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Chest radiography showed a mass shadow in the right lower lung field on the mediastinal side. The tumor was isodense, and calcification within the tumor was seen on the chest radiograph (Fig. 1). Chest radiographic computed tomography (CT) showed a 6 x 6 cm tumor in the right lung field. The tumor was touching the right cardiac atrium and the border between the right cardiac atrium and the tumor was not clear. The outer part of the tumor was calcified. There were low-density areas with septae inside the tumor. The tumor was slightly enhanced by an administration of contrast medium (Fig. 2). Magnetic resonance imaging (MRI) of the chest revealed that almost the entire tumor was enhanced by gadolinium enhancement (figure not shown). Bone scintigraphy showed extremely high uptake of 99mTc-hydroxymethylene diphosphonate (99mTc-HMDP) in the tumor suggesting high metabolism of minerals (Fig. 3). Ga-citrate scintigraphy and 123I-metaiodobenzylguanidine (123I-MIBG) scintigraphy revealed no abnormal uptake by the tumor. We considered that neuroblastoma and paraganglioma were less likely because 123I-MIBG uptake into the tumor was not increased. The chest imagings and high uptake of 99mTc-HMDP suggested that the tumor in the right hemithorax is a benign calcified diaphragm tumor.

The patient underwent surgical resection. The tumor was located within the right hemi-diaphragm. Its surface was smooth and there was no adhesion to the lung, although the tumor was weakly adhered to the pericardium. The size of the tumor was 9x7 cm while inside it was cavernous and filled with blood. Pathological examination revealed that the lesion was composed of multiple vascular spaces, varying in size from cavernous to capillary-size. Focal areas were present in which spindled endothelial cells proliferated with a solid architecture in a pattern somewhat reminiscent of a spindle cell hemangioendothelioma. Calcifications were present throughout the lesion, and appeared to be dystrophic in nature (Fig. 4).
The lesion was composed of multiple vascular spaces. Focal areas were present in which spindled endothelial cells were seen. Calcification was present throughout the lesion. The diagnosis was extensively calcified benign hemangioma.

was no finding which suggested the lesion to be malignant. The tumor did not contain any epidermal component. From these findings, we concluded that the tumor was extensively calcified benign hemangioma of the diaphragm.

Discussion

Only 84 recognized cases were found in a literature search over 100-year period (from 1868 to 1968) (4). A variety of histological types are seen in diaphragm tumor (5). In the present case, the pathological diagnosis was hemangioma, because the lesion was composed of multiple vascular spaces, spindled endothelial cells were present and the tumor did not contain an epidermal component. According to a review by Olafsson et al, hemangioma of the diaphragm was less than 5% (4).

Usually, patients with benign diaphragm tumor complain of no symptoms; in contrast, malignant diaphragm tumors cause epigastric or lower chest pain, cough, dyspnea and gastrointestinal discomfort. In the present case, the patient had experienced faint right lateral chest pain several times on the deep inspirations. This lateral chest pain probably was correlated with the diaphragm tumor.

The present case had systemic alopecia. Skin biopsy was done to confirm the cause of the hair loss. Her hair loss was diagnosed as alopecia areata, because normal hair follicular cells were found in the skin. We believe that her alopecia areata had no correlation with the diaphragm tumor.

Radiologically, most diaphragm tumors appear as smooth or lobulated soft tissue masses protruding into the inferior portion of the lung. A target-like appearance has been reported to be a characteristic finding of primary diaphragmatic schwannoma (6). Our case showed a mass shadow with calcification in the inferior portion of the right lung. The enhanced CT and MRI showed septae inside the tumor. These findings suggested that the tumor was benign. By the rapid introduction of CT and MRI into clinics, the incidence of diaphragm tumor may increase.

In the present case, extensive calcification and high $^{99m}$Tc-HMDP uptake were characteristic. Bone scintigraphy was performed because we first considered that the tumor was calcified teratoma. However, high $^{99m}$Tc-HMDP uptake did not support this possibility. Mechanisms of $^{99m}$Tc-HMDP uptake into bones are not well understood. Phosphate compounds are thought to chemisorb at kink and dislocation sites on the surface of the hydroxyapatite, with release of tin and $^{99m}$Tc, which are hydrolyzed and bound to bone either separately or together as hydrated tin oxide and technetium dioxide (7). Bone-scanning radiopharmaceuticals concentrate in many soft-tissue lesions (8). The mechanism for localization of bone scanning agents in noncalcified soft tissue may be related to movement of calcium from plasma into damaged muscle cells through abnormally permeable sarcolemma (9). In the present case, bone marrow component, osteogenic cells or osteolytic cells were not found in the tumor. Therefore, we speculated that the reason for the high $^{99m}$Tc-HMDP uptake into the tumor was due to calcium metabolism of the vascular endothelial cells of the hemangioma, which caused the extensive calcification.

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