Malignant Mesothelioma Presenting as Large Neck Mass

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Malignant mesothelioma is the most common primary malignant tumor involving pleura, but its diagnosis is difficult to determine by pathology in addition to the fact that it is rare. We present an unusual case of malignant mesothelioma, which initially presented as large neck mass contrary to the more common presentation of a rind like growth along the pleura demonstrated on imaging and by pathologic findings.

Key Words: Mesothelioma, Neck, Pleura

Introduction

Malignant mesothelioma is a rare and aggressive malignant tumor that arises from the pleura or, rarely, peritoneal and pericardial cavities, and the tunica vaginalis testis. Its incidence is increasing due to long latency period from asbestos exposure and widespread use of asbestos before strong relationship of malignant mesothelioma with asbestos was documented. Immunohistochemical staining technique becomes important to confirm diagnosis1-3. We present unusual malignant mesothelioma presenting as large neck mass.

Case Report

A 61-year-old male admitted for the evaluation of right shoulder pain for several months and acute swelling of right lower neck for last 1 week. He had diabetes mellitus and no history of asbestos exposure. On physical exam, there was diffuse swelling in right lower neck as compared with left side and decreased breathing sound of right lung field. Patient did not complain chest pain or dyspnea. Laboratory findings were within normal range except elevated glucose level.

Chest roentgenogram (Figure 1A) showed large soft tissue mass in right lower neck and apical portion of right hemithorax with minimal pleural reaction at costophrenic sulcus. Non-ECG gated 16-row multi-detector computed tomography (MDCT, Light Speed 16; GE, Milwaukee, WI, USA) of chest was performed with 1.25 mm slice thickness. MDCT showed large muscle density mass in right lower neck, connected with apical portion of right hemithorax beyond rib cage, and this mass showed poor contrast enhancement (< HU 10) (Figure 1B). Magnetic resonance imaging (MRI, SIGNA HDx; GE) was performed, and this mass showed homogenous iso-signal intensity (SI) on T1 weighted image and homogenous high SI on T2 weighted image with well defined border compared to the signal of adjacent muscles. This mass showed homogenous enhancement after contrast infusion (Figure 1C-E). There was no definite evidence of necrosis or hemorrhage within the mass on CT and MR images. Integrated computed tomography-positron emission tomography (CT-PET, Biograph LSO; Siemens, Knoxville, TN, USA) was done and intense uptakes of this large mass and scattered small pleural plaques were revealed (Figure 1F). There was
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Figure 1. Images of malignant mesothelioma. Chest radiography (A) shows large soft tissue mass (☆) in right lower neck and right apex and small pleural effusion (arrow) in right costophrenic sulcus. There is no definite displacement or destruction of rib and contracture of right hemithorax. Contrast enhanced coronal CT image (B) reveals large muscle density malignant mesothelioma (☆) in right lower neck and right apex through rib cages, Mild pleural thickening (arrow head) and small pleural effusion (arrow) are also seen. The mass (☆) shows iso-signal intensity on T1 weighted axial MR image (C) and high signal intensity on T2 weighted image (D) with good contrast enhancement on coronal T1 weighted image (E). Soft tissue contrast between tumor and adjacent soft tissue is superior in MR than CT. Coronal PET image (F) reveals the intense uptake (SUV=9.6) of large malignant mass (☆) and small pleural nodules (arrow heads). SUV: standardized uptake valve.

no displacement or destruction of ribs.

Closed pleural biopsy and thoracentesis were done, and poorly differentiated adenocarcinoma was revealed. Open surgical biopsy of neck mass was performed, and poorly differentiated adenocarcinoma was revealed on hematoxylin-eosin (H&E) stain (Figure 2A). Special immunohistochemical stains using calretinin and cytokeratin 5/6 confirmed the epithelial type of malignant mesothelioma (Figure 2B, C).

Patient has received chemotherapy using pemetrexed-cisplatin (9 cycles) and gemcitabine-carboplatin (2 cycles) and been tolerable state for 17 months after diagnosis, and he is still alive with relatively good general status and regression of neck mass (Figure 3).

Discussion

Malignant mesothelioma is originated from the mesothelial tissues of the pleura, peritoneum, pericardium, and tunica vaginalis testis. Most tumors are originated from pleura (>90%) and followed by the peritoneum (6~10%). About 2,000~3,000 new cases are reported in the United States every year. Approximately 80% of malignant mesothelioma is associated with prior as-
bestos exposure. Mean age at diagnosis is 60 years because of long latent period between occupational asbestos exposure and tumor development and the ratio of male to female is 5 : 1. A review about mesothelioma revealed that 99% had a latent period of more than 15 years and 96% had at least 20 years latent period. Median latent period was 32 years.

Frequent symptoms are dyspnea, chest pain, cough, fatigue, and weight loss. Common physical findings are dullness to percussion and decreased breathing sound. Marked unilateral contraction of affected side of hemithorax may be possible in advanced case. Laboratory findings are usually nonspecific. Patient in this case does not complain dyspnea and chest pain and complaint neck mass and shoulder pain. Breathing sound is decreased in right hemithorax.

Common abnormal findings of chest radiograph are
pleural effusion, pleural plaque, and pleural thickening. Diffuse pleural thickening is most common finding5, CT is the primary modality for tumor staging. Common CT findings are pleural thickening, interlobular fissure thickening, pleural effusion, pleural calcification, and chest wall invasion5,6. Pleural thickening is also most common feature on CT, Tumor growth leads to rindlike encasement of the lung, and contraction of the affected hemithorax is also common. In chest wall involvement, frequent manifestations are obliteration of extrapleural fat planes, invasion of intercostal muscles, rib displacement, and bone destruction5. In our case, malignant mesothelioma showed unusual growth into neck beyond rib cage and did not show rindlike growth within the hemithorax or rib destruction, MRI permits determination of tumor extent for staging and preoperative evaluation because of its excellent soft tissue contrast resolution. Compared with the SI of adjacent muscle, malignant mesothelioma reveals iso- or slightly high SI on T1-weighted images and moderately high SI on T2-weighted images. With gadolinium-based contrast, this tumor is well enhanced5. Our case showed same nature with good enhancement, and tumor was well delineated on MR, MR imaging is problem solving method to evaluate questionable area of local tumor extension5,7. PET in the evaluation of malignant mesothelioma can be useful in the detection of occult distant metastasis and nodal involvement, but the evaluation of local tumor extension is limited due to its poor spatial resolution. Integrated CT-PET imaging provides anatomic and functional information and improves the accuracy of staging. PET standardized uptake value (SUV) is also used in treatment prognosis. Low SUV and epithelial histology mean the best survival and high SUV and nonepithelial histology mean the worst survival7. This case shows high SUV and epithelial histology, and has been tolerable and still alive for 19 months.

Malignant mesothelioma is divided into three histologic categories: epithelial, sarcomatous/fibrous, and biphasic or mixed. About 50% of pleural and 75% of peritoneal mesotheliomas is epithelial type, Mixed type is 30% and sarcomatous type is 15~20%5. Sarcomatous type has poorer prognosis than epithelial or mixed type6, Adequate tissue sampling is important to permit accurate diagnosis. Open pleural biopsy, thoracoscopy, and core needle biopsy are recommended to confirm diagnosis but thoracentesis or closed pleural biopsy are not diagnostic in about two thirds of malignant mesothelioma5,55. In our case, open biopsy was performed. Immunohistochemical stains, especially calretinin and cytokeratin 5/6, are useful to differentiate malignant mesothelioma from adenocarcinoma5.

Radiation therapy or chemotherapy alone showed limited response. Combined chemotherapy using pemetrexed with cisplatin is effective in response rate and median survival time9. Our case received above mentioned combined chemotherapy and is well responded. Surgery alone showed poor survival time less than 1 year, and multimodality therapy composed of surgery, chemotherapy, and radiation therapy showed longer survival time (18.1~19 months)5. So accurate staging is important to select surgical candidate, and CT, MR imaging, and PET are crucial in staging.

We report an unusual case of malignant mesothelioma appearing as large neck mass with its radiological and pathologic findings contrary to usual rindlike growth along the pleura.

References

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