SCLEROSING HEMANGIOMA OF THE LUNG AND CONCURRENT SARCOIDOSIS OF THE HILAR LYMPH NODES: REPORT OF A CASE

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ABSTRACT

Objective: To present chest radiographs, CT and pathological findings of pulmonary sclerosing hemangioma with concurrent sarcoidosis. To review the literatures regarding lung cancer and associated sarcoidosis and/or sclerosing hemangioma.

Materials and Methods: A 50 year-old female patient was screened for health at human dock at Preventive Medicine Research Center of our institution, and a rounded nodule, measuring about 15 mm in diameter, was found in the left lower lung field medially. Non-enhanced CT showed a round nodule at the left cardiac border. The patient was sent to a clinic of our institution.

Results: The patient was further studied by contrast enhanced CT (CECT), which showed marked increase of CT value to 91.8 HU from 50.1 HU on non-enhanced CT. It was difficult to differentiate a benign lesion from malignant lesion. CECT also showed enlarged hilar and mediastinal lymph nodes. Cytology at the time of bronchoscopy revealed Class 3 at the left B5b bronchus. Partial resection of the lingual was carried out by video-assisted thoracoscopic surgery. Biopsy of the left hilar nodes (#10, #11) at the same time revealed sarcoidosis.

Conclusion: A rare case of pulmonary sclerosing hemangioma and concurrent sarcoidosis was reported with imaging results and pathological proof.

Sclerosing Hemangioma Paru dan Sarkoidosis Konkuren pada Limfonodi Hilus: Laporan Kasus

ABSTRAK

Tujuan: Melaporkan hasil pemeriksaan foto toraks, CT dan patologi pada kasus pulmonary sclerosing hemangioma yang disertai dengan sarcoidosis. Melaporkan juga penelusuran literatur berkaitan dengan kanker pulmo yang disertai dengan sarcoidosis dan/atau sclerosing hemangioma.

Bahan dan Cara: Seorang pasien perempuan usia 50 tahun dilakukan pemeriksaan skrining kesehatannya di Preventive Medicine Research Center di rumah sakit kami, dan didapatkan satu nodul bentuk bulat, ukuran diameter sekitar 15 mm, terletak di medial lapangan pulmo kiri bawah. Pada CT scan tanpa kontras menunjukkan satu nodul bentuk bulat pada batas kiri jantung. Kemudian pasien dikirim ke klinik di rumah sakit kami.

Kesimpulan: Dilaporkan satu kasus yang jarang, yaitu pulmonary sclerosing hemangioma dan disertai dengan sarcoidosis berdasarkan hasil pemeriksaan imejing dan pembuktian dengan pemeriksaan patologi.

Kata Kunci: pulmonary sclerosing hemangioma, pneumocytoma, sarcoidosis, lung cancer

INTRODUCTION

It seems to be rather rare to see sclerosing hemangioma of the lung and concurrent existence of sarcoidosis in the hilar lymph nodes. It is the purpose of this communication to report such a case with radiological and pathological findings. It seems to be rather rare to see sclerosing hemangioma of the lung and concurrent existence of sarcoidosis in the hilar lymph nodes. It is the purpose of this communication to report such a case with radiological and pathological findings.

Case History

A 55-year-old woman received health screening in the human dock at Preventive Medicine Research Center in our institution. Abnormal chest x-ray findings with a round mass in the left lower lung field on the plain chest radiograph were reported (Figure 1).

Non-enhanced CT scan of the chest at the same time showed a round mass in the lingula of the left lung (Figure 2).

Enhanced CT of the chest on the later day at the clinic showed enhancement of the mass to 91.8 HU from 50.1 HU on the plain CT (Figure 3).
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Figure 4. Mediastinal lymph nodes. CE-CT: Mediastinal window. (A - F). There are multiple swollen lymph nodes (arrows) in the mediastinum and both hili. Two lymph nodes (#10, #11) were resected. Pathology of the lymph nodes revealed sarcoidosis

Past History

Nothing was contributory. At the same time diabetes mellitus was suspected.

Laboratory Findings

Laboratory findings: The blood count was within normal limits. Elevated blood sugar level was noted.

Clinical Course and Treatment

The patient was placed on dietetic treatment for her diabetes mellitus at first. The condition became better, revealing improvement of the serum glucose (95 mg/dl) and HbA1C (6.2).

She was studied by bronchoscopy, which revealed a lesion in the left bronchus, B5b, and cytology revealed class 3. The patient developed sputa and fever after bronchoscopy. She was seen in the emergency room and elevated WBC (10,400), CRP (6.9), and a small infiltrate in the left lower lung field on the chest radiograph were found. She was placed on chemotherapy with CFPM, and the disease (pneumonia) was cured. WBC returned to the normal range, and CRP dropped to 2.19. Then thoracoscopic segmental resection of the lingula with a round mass was carried out. Hilar lymph node was also resected for pathological examination.

Pathology

Rapid pathological examination of the frozen section of the lung tumor revealed sclerosing hemangioma (Figure 5). Histologic preparation of the hilar lymph nodes (#10, #11) revealed sarcoidosis (Figure 6).

Figure 5. Photomicrograph of the pulmonary nodule (Hematoxylin-eosin stain). Sheet-like growth of cells with rounded to elliptic shapes and light eosinophilic cytoplasm were noted. Vascular luminal structures are observed. The inner layer (intima) of the luminal structure showed Type II pneumocytes. Hemorrhage, hemosiderosis and fibrosis were seen in the tumor tissue. Numerous foreign body giant cells appeared around the tumor. The findings are consistent with pulmonary sclerosing hemangioma.

Figure 6. Photomicrograph of lymph nodes. Granulomas with Langerhans giant cells are noted in the lymph nodes. Asteroid body (arrow) is noted in the giant cells. The findings are consistent with sarcoidosis. H & E.

DISCUSSION

Pathology of the pulmonary sclerosing hemangioma has been controversial. According to
One primary lesion. Chan and Chan reported one out of 10 cases of multicentric in origin or intralobar metastases from that it was not certain whether the lesions were sclerosing hemangioma of the lung. They stated Satoh's findings in stating that pulmonary sclerosing hemangioma is an epithelial tumor with differentiation towards type II pneumocytes by immunohistochemical and immunoelectron microscopic studies. They stated that this tumor has to be considered a low-grade malignant tumor.

Nagata et al. studied sclerosing hemangioma using immunohistochemistry with antibody against surfactant apoprotein, and concluded that the proliferating cells in the stroma might be epithelial in origin and would differentiate to type II pneumocytes. Satoh et al. support Kennedy's findings in stating that pulmonary sclerosing hemangioma is an epithelial tumor with differentiation towards type II pneumocytes by immunohistochemical and immunoelectron microscopic studies. They stated that this tumor has to be considered a low-grade malignant tumor.

Noguchi et al. reported a case of multiple sclerosing hemangioma of the lung. They stated that it was not certain whether the lesions were multicentric in origin or intralobar metastases from one primary lesion. Chan and Chan reported one out of 16 cases of pulmonary sclerosing hemangioma, which metastasized to regional lymph nodes. They also studied the cases with Thyroid Transcription Factor-1 (TTF-1), and suggested the tumor was an epithelial neoplasm derived from primitive respiratory epithelium or incompletely differentiated type II pneumocyte or Clara cell. They also found metastases in the lymph nodes in their cases, and commented that identification of metastasis in rare cases of sclerosing hemangioma did not necessarily put this group of tumors in the potentially malignant category, because other benign tumors had also been reported to metastasize, for example, metastasizing pleomorphic adenoma, giant cell tumor of bone and chondroblastoma. Most cases of pulmonary sclerosing hemangioma were found incidentally by chest roentgenography.

Yi et al. studied one hundred and thirty-one solitary pulmonary nodules with unenhanced CT and dynamic CT. Among the lesions they found 5 cases of sclerosing hemangioma. They stated that sclerosing hemangiomas and active granulomas are highly enhancing benign nodules, although dynamic study of pulmonary nodules is highly sensitive (99%) in differential diagnosis between malignant and benign nodules, when they set the cut-off value at 30 HU. We also found marked enhancement of the nodule by contrast-enhanced CT, and CT value increase by 41.7 HU.

Tanaka et al. reported a case of pneumocytoma (so-called sclerosing hemangioma) with lymph node metastasis. The lesion was a well-defined round lesion in the right lung, which increased in size slightly during a 2-year follow-up period. The bronchial arteriography of the nodule showed irregular hypervascularity in the tumor.

The tumor and hilar lymph nodes were resected. The nodule and lymph nodes were examined by light microscopy, electron microscopy and immunohistochemistry. The tumor showed various histological features characteristic of sclerosing hemangioma. There were a few small metastatic foci in the hilar lymph nodes. They support the neoplastic theory, indicating that sclerosing hemangioma is a true neoplasm derived from alveolar epithelial cells showing differentiation toward type II pneumocytes and having the capability to metastasize, although metastasis is extremely rare.

Im et al. reported CT findings of pulmonary sclerosing hemangioma in eight patients and also CT-pathology correlation. Seven out of eight patients were female, and one was male. The lesion was found as an incidental lung mass on chest radiographs. The tumor enhanced up to 96 to 157 HU in density. The markedly enhanced area corresponded with angiomatous area, and isodensity area to solid and sclerotic areas. Three patients had calcification. They concluded that a well defined juxtapleural mass with marked contrast enhancement coupled in select cases with foci of sharply margined areas of low attenuation and calcification are characteristic CT findings of sclerosing hemangioma and should suggest this diagnosis especially in women with these findings. Nakanishi et al. also reported strong enhancement on CECT and CE-MRI. In case of CT, the CT value increased to 139 HU from 57 HU. In case of dynamic MRI, the marked increase in intensity took place at 2 minutes after the injection of Gd-DTPA.

There are several reports on primary lung cancer associated with sarcoidosis or vice versa. Yamauchi reported the incidence of associated cancer of the lung in cases of sarcoidosis. She found 3 cancer of the lung (0.2%) among 1,487 cases of sarcoidosis, which were registered in Japan. She introduced the results of follow-up study of sarcoidosis and development of cancer from the registered cases of sarcoidosis in Denmark, reported by Brincker and Wilbek. They found 48 cases...
(1.9%) of malignant lesions out of followed 2544 cases of sarcoidosis.

Kawakami et al.\textsuperscript{13} reported a case of primary lung cancer associated with sarcoidosis. Cancer was situated in the left upper lobe and sarcoidosis is located in the left lower lobe, which was difficult to differentiate from metastasis of cancer. They postulated that sarcoidosis had close relationship with development of cancer in this case, although they suggested further studies.

Hirasawa et al.\textsuperscript{14} followed 688 patients with sarcoidosis in 14 years, and found 43 malignant tumors (6.25%). They found various cancers in the past history in 29 cases and new cancer was noted in 24 patients. The origins of new cancers are uterus, breast, lung, skin, stomach, kidney, colon, thyroid, lymphoma, caecum, ovary, esophagus, and brain. Ogata et al.\textsuperscript{15} noted that six (0.8%) of 753 cases with lung cancer had concurrent sarcoidosis.

Co-existence of pulmonary sclerosing hemangioma and sarcoidosis seems to be very rare. We found only one literature on such lesions. Maruyama et al.\textsuperscript{16} reported a case of pulmonary sarcoidosis with lung cancer and sclerosing hemangioma. The female patient had been followed up for abnormal shadow in the hilar region of the right lung. She was admitted for the treatment of glaucoma caused by ocular sarcoidosis. Chest CT revealed a mass in the right S5 and a nodule in the left S3, and multiple mediastinal lymph node swelling. Operation was performed. The lesion in the left upper lobe was adenocarcinoma, but the right mass was sclerosing hemangioma. Frozen sections of some of diffusely scattered 2 to 3 mm nodules on the pleura revealed sarcoidosis. This case showed triple lesions.

Our case is of double lesions of pulmonary sclerosing hemangioma and incidentally found sarcoidosis. In case of a hilar lesion associated with a pulmonary lesion one must consider and differentiate sarcoid reaction. There is practically no appearance of Langerhans giant cells in sarcoid reaction. We confirmed Langerhans giant cells in the lymph nodes in our case, and concluded that the lymph nodes were consistent with sarcoidosis.

**CONCLUSION**

A case of pulmonary sclerosing hemangioma and concurrent sarcoidosis was reported. Plain chest radiographs, non-enhanced and enhanced CT and microscopic findings in this case were presented. This combination of such lesions seems to be very rare. The literatures regarding imaging and pathology of pulmonary sclerosing hemangioma and concurrent occurrence of lung cancer and sarcoidosis were reviewed.

The term of pneumocytoma was refered from the pathological standpoint. The term pneumocytoma, will be used with sclerosing hemangioma interchangeably or mainly in future.

**REFERENCES**

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