Sclerosing Hemangioma of the Lung: An Analysis of 44 Cases

Background. Sclerosing hemangioma is a rare benign lung tumor. Preoperative diagnosis of this tumor is difficult, and some times even intraoperative frozen sections cannot differentiate it from malignant tumors. Herein, we present our experiences in investigating its characteristics.

Methods. The medical records and pathological examinations of patients diagnosed with sclerosing hemangioma from 1982 to 2001 at the Division of Thoracic Surgery in Taipei-VGH were retrospectively reviewed.

Results. The incidence of sclerosing hemangioma in benign lung tumors resected during that period was 32.8%. There were 44 patients (7 male and 37 female) aged from 16 to 72 years, with a peak at the fifth decade, and 72.7% of them were asymptomatic. Accurate preoperative diagnosis by chest CT could be achieved in only 20% (4/20) of patients, and malignancy could not be ruled out in 40% (8/20) of tumors. Histologically, this tumor exhibits four major patterns: solid, sclerotic, papillary and hemorrhagic. Five patients in this series had predominantly one pattern. At least two patterns existed in the remaining 39 patients, including eight patients who had tumors with all four patterns. There was no operative mortality or tumor recurrence despite that different operative methods were undertaken.

Conclusions. Although rare, sclerosing hemangioma still accounts for the second common benign lung tumor in the current series. Chest CT and bronchoscopic examinations could not make accurate diagnosis preoperatively, and thoracotomy is usually indicated for definitive diagnosis and treatment. Limited resection is warranted in view of uncommon tumor recurrence. In addition, multiple nodular lesions could not exclude the possibility of sclerosing hemangioma.

Original Article

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Key Words
benign lung tumor; pulmonary resection; sclerosing hemangioma

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examinations and follow-up data were reviewed. Student’s t test and Chi-square test were used for calculating statistical differences of clinical data.

RESULTS

Clinical data

The clinical data are summarized in Table 2. There were 7 male and 37 female patients with sclerosing hemangioma. The age at the time of operation ranged from 16 to 72 years (mean age, 46.5 ± 14.3 years), and appeared most commonly in the fifth decade of life (n = 12). The male (35.4 ± 19.8 years) patients were much younger than female (48.6 ± 12.3 years) patients (p = 0.023 by Student’s t test).

Of 44 patients, 32 (72.7%) patients were asymptomatic. These tumors were discovered either on routine chest roentgenograms or follow-up films for other diseases. Other presenting symptoms are also summarized in Table 2.

Radiographic manifestations

All but one patient had a solitary tumor on chest roentgenograms. The exceptional patient had one major tumor, 60 mm in diameter, located at right upper lobe and multiple smaller tumors scattering at right lower lobe. The distribution of all tumors is listed in Table 3. Of interest, one tumor crossed the left major fissure with the main tumor located at LUL.

Computed tomography (CT) scan of chest was performed in patients who under went surgically resection after 1990, but only 20 patients had data available. In four patients, sclerosing hemangioma was diagnosed correctly by radiologists. Nevertheless, malignancy was suspected, to various degrees, in 40% (8/20) of tumors for well-defined margin, large tumor size and older age of the patient. The remaining eight patients were diagnosed as having hamartomas, tuberculomas or other benign lesions. Meanwhile, calcification was observed on chest CT in only three patients and was considered a sign of benignity. Magnetic resonance imaging (MRI) of thorax was performed in an other two patients, and both tumors were considered benign with out a definite diagnosis of sclerosing hemangioma.

Bronchoscopy

Bronchoscopy was performed in 25 patients, and no gross tumor could be seen in all visible fields. Among them, seven patients received biopsy and four patients received brushing cytology. Chronic inflammation or fibrosis was the usual pathology diagnosis in biopsy, and no abnormal cell could be seen in brushing cytology.

Operations

The operative methods are also listed in Table 3.
Enucleation was the most common procedure performed, and accounted for 36.4% of the patients. A lobectomy was usually performed due to central location of tumor, and nearly 30% of the patients underwent this procedure. In our analysis (Table 4), major operations including pneumonectomy and lobectomy were associated with larger tumor size, especially those more than 30 mm (p = 0.001). Moreover, in eight patients with an RML tumor, lobectomy was the most common procedure (50%) carried out, for either central location of tumor or less normal lung tissue could be left after a wedge resection or an enucleation. One patient underwent a right side pneumonectomy. This patient was a case of multiple tumors in different lobes without exclusion of malignancy in frozen sections.

An intraoperative frozen section for pathological examination was performed in 31 patients. Among them, 18 patients were diagnosed as having sclerosing hemangioma. Ten patients were claimed to have a benign lesion without a definite diagnosis. Due to ambiguous morphology, however, malignancy had been suspected in three patients and thus led to a more extended resection. Lymph node sampling was performed in 11 patients including one male and 10 females, and no lymph node metastasis was found.

There were three patients with complications. One patient sustained chylothorax, one patient had wound infection and the other patient suffered from persistent air leak. There was no operative mortality.

During the follow-up ranging from three months to 19 years, none of these patients experienced tumor recurrence, despite different kinds of resections.

**Pathological findings**

The greatest diameter of the tumor ranged from 7 mm to 60 mm, and the average size was 29 × 24 × 19 mm. It was no more than 30 mm in 33 (75%) patients.

Microscopically, these tumors had four major patterns: solid, sclerotic, papillary and hemorrhagic (Fig. 1). The case numbers of each pat tern are listed in Table 5. We found that most tumors had a mixture of different patterns, and only five tumors had predominantly one pattern. Among these five tumors, papillary patterns accounted for three and hemorrhagic patterns accounted for two. On the other hand, eight tumors comprised all these patterns.

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four patterns, whereas 19 tumors had three patterns and 12 tumors had two patterns. The most common pattern in this series was sclerotic pattern, identified in 32 (72.7%) tumors. The least common pattern was solid pattern, found in 19 (43.2%) tumors.

DISCUSSION

In our 20 years’ experience, sclerosing hemangioma is the second common benign lung tumor. In the current series, it was less common than hamartoma but accounted for 32.8% of all cases. This incidence was much higher than the incidence of previous western reports, and seemed closer to a recent Japanese series that reported an incidence of 22% in 45 patients. The ethnic difference between Orientals and Caucasians needs further studies to verify.

This tumor was reported to have some clinical characteristics, such as female preponderance and occurrence mostly in the fifth decade. Our study showed that 84% of the patients were female and 27.3% of the patients aged from 40 to 49 years. Both were compatible with previous reports. Interestingly, the male patients was significantly younger than female patients. Nevertheless, its cause remains unclear.

Up to 72.7% of our patients were asymptomatic, and the common presenting symptoms were hemoptysis, cough and chest pain. In our experiences, the symptoms were not related to tumor size or distribution. These data were similar to previous reports.

It has been stated that the distribution of sclerosing hemangioma some what favors the lower lobes, if compared with the upper lobes. In Dail’s review, 47% of tumors were located at lower lobes and only 32% of tumors were located at upper lobes. In our series, however, lower lobes accounted for 43.2% and upper lobes accounted for 36.3%. The preponderance of lower lobe distribution seems not so distinct here. Moreover, we found that the tumor occurred in right lung almost twice more than in left lung. This interesting phenomenon could not be totally explained by volume of both lungs.
Multifocal sclerosing hemangiomas of the lung is a very rare condition. In the current series, there was a 32-year-old woman with a large (60 × 50 × 40 mm) main tumor over RUL and multiple smaller tumors over RLL. The main tumor lay adjacent to right pulmonary trunk and could not be dissected completely. Since the frozen section of the RUL tumor could not exclude a malignancy, the patient finally underwent a right pneumonectomy. The smaller tumors in RLL were later proved also to be sclerosing hemangiomas. Unlike the case reported by Noguchi et al. in 1986,8 which presented with a large tumor and multiple daughter tumors in RLL, this tumor appeared in distal lobes simultaneously. Mean while, the main tumor and the minute lesions in our case had the same pathologic elements of sclerotic, papillary and hemorrhagic components. This was somewhat different from Noguchi’s report, which included the minute lesions in our case had less structures than the main tumor and considered some structures as second ary changes. Despite these findings, it was still uncertain whether this was a con dition of transbronchial intrapulmonary spread or synchronous tumor growth. Nevertheless, the patient recovered well and no recurrence or metastasis was found during the follow-up.

Although sclerosing hemangioma is a well-documented benign tumor, it is sometimes misdiagnosed as a malignant lesion, especially in intraoperative frozen sections because of its variable histologic elements. For example, adenocarcinoma may be sus pected in areas with papillary fronds and angiosarcoma may be considered when hemorrhagic component is present. In the current series, three tumors were diagnosed as malignancy in intraoperative frozen sections. Two of them were later proved to have a papillary component and one had a hemorrhagic component. Features were ful ful to exclude a diagnosis of malignancy and metastasis, es pecially in larger tumors. In our series, lobectomy was performed in 29.5% of patients and this ratio may be higher than expected. We attributed this to larger tumor size, central location of tumor and an RML tumor itself. Recently, video-assisted thoracoscopic surgery (VATS) has become more popular for treatment of benign lung diseases. All the procedures listed above, including lobectomy, could be carried out safely by skilled surgeons.

Recently, there were some reports on lymph node metastasis, especially in larger tumors. In our series, 11 patients underwent lymph node sampling, with the number of the re moved lymph nodes ranged from 2 to 29. No lymph node metastasis was found, despite that two tumors were larger than 50 mm.

The histogenesis of sclerosing hemangioma remains obscure even decades after its first description by Liebow and Hubbell. At first, they suggested this tumor as a proliferation of endothelial cells, which was supported by some investigators.12 Different opinions appeared later, and Katzenstein et al. used histochemical technique and glycosaminoglycan electrophoresis in addition to electron microscopy to suggest this tumor as a form of benign epithelial mesothelioma.13 Nagata et al. and Satoh et al. utilized anti-lung surfactant apoprotein antibody to demonstrate that this tumor was constituted of epithelial cells with differentiation to type II pneumocytes.14,15 Yousem et al. further proved this view by applying a panel of various antibodies including cytokeratin, EMA, carcinoembryonic antigen and Clara cell antigen on eight cases of sclerosing hemangioma.16 Interestingly, Leong et al. demonstrated that this tumor expressed progesterone and estrogen receptor proteins, and thus it may be correlated with the predilection of this tumor for women.17

From previous reports as well as our study, it is obvious that sclerosing hemangioma seldom recurs after complete removal of tumor. Although a lobectomy is sometimes indicated for central location of tumor, a wedge resection or enucleation is usually sufficient. In our series, lobectomy was performed in 29.5% of patients and this ratio may be higher than expected. We attributed it to larger tumor size, central location of tumor and an RML tumor itself. Recently, video-assisted thoracoscopic surgery (VATS) has become more popular for treatment of benign lung diseases. All the procedures listed above, including lobectomy, could be carried out safely by skilled surgeons.18

Our study indicated a much higher incidence of sclerosing hemangioma in benign lung tumors in Chinese than in western populations. However, the clinical features are not different. Once a mid-life-aged Chinese woman presented with a well-defined coin lesion on the
chest film, sclerosing hemangioma should be considered first. Chest CT and bronchoscopic examinations of fersufficient in formation for accurate diagnosis and operative -

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fully. Lymph node metastasis does exist but is relatively rare, and lymph node sampling should be suggested in larger tumors. Multiple lesions, although usually considered as a sign of metastasis, should not exclude the possibility of sclerosing hemangioma.

REFERENCES