Plexiform Neurofibromatosis of the Mediastinum: CT Findings

Chul Joong Kim, M.D., Min Jin Lee, M.D., Yu Whan Oh, M.D., Won Hyuck Suh, M.D., Yung Suk Lee, M.D.

Purpose: To evaluate the findings and the role of CT in plexiform neurofibromatosis of the mediastinum.

Materials and Methods: We retrospectively reviewed the CT scans of five patients with plexiform neurofibromatosis of the mediastinum. The CT scans were reviewed with attention to the distribution of the lesions, appearance and attenuation of mediastinal lesions, enhancement pattern after intravenous contrast infusion and associated findings such as intercostal neurofibromas.

Results: In all five patients CT scans demonstrated fusiform low attenuated masses which were oriented longitudinally and extended over multiple contiguous scans along the distribution of major mediastinal nerves. In four patients, mediastinal lesions appeared infiltrative, obliterating adjacent mediastinal fat plane. One patient had well defined fusiform masses along the major mediastinal nerves. Postcontrast enhanced CT scans revealed slight central enhancement in two patient and no contrast enhancement in three patients. Associated findings such as neurofibromas of intercostal nerves and sympathetic trunk, or subcutaneous neurofibromas were detected on CT scans in all five patients.

Conclusion: Characteristic CT findings of low attenuation masses along the major mediastinal nerves are helpful to differentiate plexiform neurofibromatosis from mediastinal lymphadenopathy and to prevent from misreading as a malignant disease.

Index Words: Neurofibromatosis, Mediastinum, CT

INTRODUCTION

Plexiform neurofibromas usually occur in the neck, pelvis and extremities, but they may seen virtually at any location, including the thorax(1–5). In the thorax, the sympathetic chains are most commonly involved, but plexiform neurofibromas of the vagus and phrenic nerves are extremely rare(6). Due to its anatomical position it is important to differentiate these lesion from mediastinal lymphadenopathy to prevent a mistake of falsely diagnosing as a malignant disease.

There are several previous descriptions of the CT appearance of plexiform neurofibromas involving the major mediastinal nerves(1, 6). We want to describe about the findings and the roles of CT for plexiform neurofibromatosis of the mediastinum.

MATERIALS and METHODS

We retrospectively reviewed the clinical results and thoracic CT scans in five patients with pathologically proved plexiform neurofibromatosis of the mediastinum. In four patients with cafe-au-lait spots in skin, neurofibromatosis was diagnosed by biopsy of one of the subcutaneous nodules. One patient had neurofibromas in neck and superior mediastinum that were confirmed by surgery. Five patients included four men and one woman and the patients age ranged from 10 to 49 years (mean age, 25 years). None of the patients had a positive family history of von Recklinghausen neurofibromatosis. Two complained episodic chest pain and one was asymptomatic, and their tumors had been found incidentally upon a routine radiographic examin-
ation. Remaining two patients each had palpable neck mass. Four had multiple subcutaneous neurofibromas and café-au-lait spots in skin. The plain chest radiograph showed superior mediastinal widening in all patients.

All five patients underwent both precontrast and postcontrast CT examinations with a Somatom 2 (Siemens, Erlangen, Germany) and Somatom Plus-S CT scanner (Siemens, Erlangen, Germany). CT study consisted of scans of 10 mm collimation at 10 mm intervals extending from the lung apices to the bases. In contrast studies, 100 ml of nonionic contrast medium (lopamiro 300; Bracco, Milano, Italy) was administered by hand injection. The images were photographed at conventional mediastinal window setting (level = 40 HU; width = 350 HU).

CT scans were reviewed with attention to the following findings: (1) distribution and anatomical location of the mediastinal lesions, demonstrating the involved major mediastinal nerves; (2) appearance and definition of the mediastinal lesions (obliteration of fat plane surrounding mediastinal lesions), (3) attenuation of the mediastinal lesions with respect to chest-wall muscle and subcutaneous fat, (4) enhancement pattern after IV contrast infusion, (5) associated intrathoracic findings such as neurofibromas of the intercostal nerve, subcutaneous area, and of sympathetic trunk.

RESULTS

CT scan revealed that the lesions involved predominantly both vagus and/or phrenic nerves in the mediastinum in all five patients (Table 1). These lesions longitudinally oriented and extended over multiple

<table>
<thead>
<tr>
<th>Case No/ Age/ Sex</th>
<th>Plexiform Neurofibromatosis of Mediastinum</th>
<th>Associated Neurofibromas</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 / 25 / M</td>
<td>both phrenic and vagus n,</td>
<td>multiple intercostal n,</td>
</tr>
<tr>
<td></td>
<td>left recurrent laryngeal n,</td>
<td>both brachial plexuses,</td>
</tr>
<tr>
<td></td>
<td>both sympathetic trunk</td>
<td>n in left back muscle,</td>
</tr>
<tr>
<td>2 / 49 / M</td>
<td>both phrenic n</td>
<td>multiple intercostal n</td>
</tr>
<tr>
<td>3 / 10 / F</td>
<td>both vagus n</td>
<td>n in right neck muscle</td>
</tr>
<tr>
<td>4 / 16 / M</td>
<td>both vagus n</td>
<td>multiple intercostal n</td>
</tr>
<tr>
<td>5 / 20 / M</td>
<td>both vagus and phrenic n,</td>
<td>subcutaneous neurofibromas</td>
</tr>
<tr>
<td></td>
<td>both sympathetic trunk</td>
<td>n in left back muscle</td>
</tr>
</tbody>
</table>

(n = nerve)

Fig. 1. a. Postcontrast CT scan in 25-year-old man with plexiform neurofibromatosis shows round low attenuation masses in both vagus nerves and nodular thickening of left recurrent laryngeal nerve (white arrow). Fusiform enlargement of left brachial plexus (black arrow) and peripheral nerve in left rhomboideus muscle (open arrow) are also seen.

b. Contiguous next CT scan demonstrates that the lesions extend along their distribution of involved nerves, such as the left recurrent laryngeal nerve (white arrow = left recurrent laryngeal nerve, arrow = left brachial plexus, open arrow = nerve in left rhomboideus muscle). Nodular thickening of intercostal nerve (small white arrows) is seen.
contiguous scans along the distribution of the major mediastinal nerves. The lesions involved the right vagus nerve were located in right paratracheal area, displacing superior vena cava and right brachiocephalic vein anteriorly. The lesions of the left vagus nerve were laterally located to the left common carotid artery & left subclavian artery and the lesion of the left recurrent laryngeal nerve was located between the left common carotid artery and esophagus(Fig. 1a, Fig. 1b, Fig. 2). The lesions involved phrenic nerve appeared nodular thickening along the mid-paracardiac area(Fig. 3). Two patients also had nodular thickening along the thoracic and/or lumbar sympathetic trunks. The thorax CT scan revealed that there was no lesions positioned in subcarinal area.

In four of five patients, the lesions showed round and nodular in appearance. Outer margin of the lesions was not well-defined and obliterating adjacent intervening mediastinal fat plane(Fig. 2) The first patient had well-defined fusiform mass, not obliterating adjacent mediastinal fat plane(Fig. 1). In all five cases, precontrast CT scan showed that all lesions had lower attenuation values than chest-wall muscle and higher attenuation values than subcutaneous fat. CT values of the lesions ranged from 15 to 20 HU. The lesions were homogeneous and had no necrotic or cystic areas within them. Postcontrast CT scans demonstrated slight enhancement at the central portions of the lesions in two patients and there was no contrast enhancement in the other patients.

The four of five patients had multiple nodular areas of soft-tissue density adjacent to inner aspect of ribs (Fig. 1). Although no histological confirmation was obtained in these patients, we had CT evidence of thickening and nodularity of the intercostal nerve. Thus these were thought to be fusiform neurofibromatous enlargement of intercostal nerves. In addition, CT scans demonstrated the neurofibromas as associated findings of plexiform neurofibromatosis of mediastinum in variable sites such as back and neck muscles, brachial plexus, or subcutaneous area(Table 1).

**DISCUSSION**

von Recklinghausen neurofibromatosis is a hereditary, autosomal dominant, hamartomatous disorder that can involve all three germ cell layers, but it primarily involves the ectoderm and mesoderm. Its frequency

---

**Fig. 2.** Postcontrast CT scan of a 20 year-old man shows nodular low attenuation masses in both vagus nerves, obliterating adjacent mediastinal fat plane. Slight contrast enhancement(white arrow) is seen in the lesion of the right vagus nerve. CT scan also demonstrates fusiform thickening of the left sympathetic trunk and intercostal nerve(black arrow) and multiple subcutaneous nodules(short arrows).

**Fig. 3.** a. Postcontrast CT scan at infrapulmonary level in 49-year-old man with plexiform neurofibromatosis shows nodular masses in both lateral sides of the aorta and pulmonary trunk(black arrows). Nodular thickening of the intercostal nerve(white arrow) is seen.

b. CT scan at midcardiac level demonstrates nodular thickening along the both phrenic nerves(arrows).
is approximately 1/2,000 to 1/5,000, and it occurs in both sexes (2,3,7). This disease is characterized clinically by a skin syndrome with cutaneous pigmentation (cafe-au-lait spots) and tumors (fibroma molluscum and plexiform neurofibroma). Tumors of the central nervous system and bony abnormalities also are common features. Thoracic manifestations in von Recklinghausen’s disease represent neurofibromas in sympathetic trunk and intercostal nerve. Involvement of plexiform neurofibromatosis of major mediastinal nerves is rare (1,6,8). Other findings include interstitial pulmonary fibrosis, lateral meningoceles, ducal ectasia, and some skeletal deformities such as kyphoscoliosis (2,9).

Plexiform neurofibroma is a diffuse and abnormal growth of Schwann cells, which results in nodular enlargement of affected nerve. Plexiform neurofibroma grossly appears as a generalized thickening or fusiform enlargement of the involved nerves. The tumors form an intricately intercommunicating network, which often traps contiguous soft tissue such as adipose tissue, presence of lipid-rich Schwann cells, presence of adipocytes (transformed fibroblasts), cystic degeneration, and myxoid matrix which has a high water content (1,2,12,13). It is important to recognize that plexiform neurofibromatosis can be shown as diffusely and uniformly nonenhanced, low attenuation values than did chest-wall muscle. The low attenuation of these lesions is histologically related to the entrapment and incorporation of adipose tissue, presence of lipid-rich Schwann cells, presence of adipocytes (transformed fibroblasts), cystic degeneration, and myxoid matrix which has a high water content (1,2,12,13). It is important to recognize that plexiform neurofibromatosis can be shown as diffusely and uniformly nonenhanced, low attenuation pattern as shown in our cases. Bourgouin et al. (1) reported the CT features of four cases of plexiform neurofibromatosis of mediastinum from which they arise (11). The resemblance of these lesions to lymphadenopathy is evident. It is important to differentiate mediastinal neurofibromatosis from mediastinal lymphadenopathy because mediastinal lymphadenopathy is frequently indicative of malignant disease. Since associated subcarinal lymphadenopathy would reasonably be expected if such extensive mediastinal masses were indeed due to enlarged lymph nodes from malignant disease. The absence of any lesion in subcarinal position was meaningful.

In conclusion, characteristic CT findings of low attenuation masses along the major mediastinal nerves are helpful to differentiate plexiform neurofibromatosis from mediastinal lymphadenopathy and to prevent from misreading as a malignant disease.

REFERENCES

12. Mirich DR, Gray RR, Grosman H. Abdominal plexiform neurofibromatosis simulating pseudomyxoma peritonei on computed
종격동의 총상 신경섬유용중: CT 소견

목적: 종격동에 발생한 총상 신경섬유종증의 CT 소견과 CT의 역할을 알아보고자 하였다.

대상 및 방법: 저자들은 종격동에 발생한 총상 신경종증이 있는 5명을 대상으로 종격동 병변의 분포, 모양, 감약정도, 조영 증강 양상 그리고 녹간신경 신경섬유종과 같은 동반소견 등을 후향적으로 조사하였다.

결과: 5명의 환자 모두에서 낮은 감약정도를 보이는 총상형의 종양이 종격동의 주 신경을 따라 분포하여 CT상 연속된에서 관찰되었다. 4명의 환자에서 종격동 병변 경계가 불분명하여 주위 지방층 경계를 소실시켰고, 한명의 환자에서 종격동 병변 경계는 분명하였다.

조영제 주입후 시행한 CT 상 2명의 환자에서 미미한 조영증강이 병변의 중심부에서 관찰되었고 3명의 환자에서는 조영증강이 없었다. 5명의 환자에서 녹간신경과 교감신경간 신경섬유종 피하결절 등이 동반되었다.

결론: 종격동의 주요 신경을 따라 자음염 종괴를 보이는 총상 신경섬유종증의 CT 소견은 악성종양으로 인한 종격동 인피절 종괴와 종격동에 발생한 총상 신경섬유종증을 감별하는 데에 도움이 될것으로 생각된다.
국제 학술대회 일정표[Ⅰ]

1994/12/13－15  26th Annual SC. Meeting British Medical Ultrasound Society
venue: Spa Center Scarborough, United Kingdom.
contact: General Secretary, Bmus,
36 Portland Place, London WIN 3DG, United Kingdom.
(tel: 44－71－6363714; fax: 44－71－3232175)

1995/01/07－13  Annual Meeting Society of Uroradiology
venue: Ritz-Carlton Palm Beach, FL, USA.
contact: David S. Hartman, M.D., University Hospital,
P.O. Box 850, Hershey, Penns. 17033, USA.
(tel: 1－717－5318044; fax: 1－717－5315596)

1995/01/25－29  6th International Symposium on Magnetic Resonance － MR '95
venue: Congress Center Garmisch-Partenkirchen, Germany.
contact: PD Dr. M. Seiderer, Radiologische Klinik,
Marchioninistrasse 15, D－81366 Muenchen, Germany.
(tel: 49－89－70952750; fax: 49－89－70958838)

1995/02/05－09  Annual Meeting Society of Thoracic Radiology
venue: Ritz Carlton San Francisco, CA, USA.
contact: STR, Univ. of CA Med. C.,
Box 0628, San Francisco, CA 94143, USA.
(tel: 1－415－4765926; fax: )

1995/03/05－10  9th European Congress of Radiology
venue: Austria Center Vienna, Austria.
contact: MR. P. Bairol, ECR-Office,
Neutorgasse 9/2A, A-1010 Vienna, Austria.
(tel: 43－1－5334064; fax: 43－1－5334064-9)

1995/03/12－16  Int. London Courses in Cimouted Tomography and Magnetic Resonance Imaging
venue: The Gleneagles Hotel Perthshire, Scotland, United Kingdom.
contact: Mrs. T. Seear, The London Clinic,
20 Devonshire Place, London WIN 2DH, Unites Kingdom.
(tel: 44－71－2240164; fax: 44－71－9352430)

1995/03/25－29  Annual Meeting Society of Magnetic Resonance Imaging
venue: Washington Hilton & Tower Washington, DC, USA.
contact: SMRM,
1918 University Ave., Suite 3C Berkeley, CA94704, USA.
(tel: 1－510－8411899; fax: 1－510－8412340)

1995/03/26－29  Annual Meeting American Institute of Ultrasound in Medicine
venue: San Francisco, USA.
contact: Convention Department, AIUM,
11200 Rockville Pike, MA 20852-3139 Rockville, USA.
(tel: 1－301－8812486; fax: 1－301－8817303)