Angiofollicular Lymph Node Hyperplasia (=Castleman's Disease)

—Report of A Case—

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Angiofollicular lymph node hyperplasia (AFLNH) with well marginated lymphoid masses, is a rare benign disease of unknown etiology. The majority of the disease develop intrathoracically. Histologically this disease can be divided into the hyaline-vascular and the plasma cell types with the hyaline-vascular type prevailing. The plasma cell variant has been associated with nephrotic syndrome, anemia, growth failure, fever, hyperglobulinemia, peripheral neuropathy, and hypoalbuminemia. Surgical resection is known to be treatment of choice in most cases, and radiotherapy is reserved for advanced, unresectable lesions. We report a complete remission of AFLNH in a case treated by sugical excision followed by irradiation.

Key Words: Angiofollicular lymph node hyperplasia, Hyaline-vascular type, Plasma cell type, radiotherapy

INTRODUCTION

Angiofollicular lymph node hyperplasia (AFLNH) was initially described in 1953 by Castleman and his co-workers.[1] This disease has been described with "large, asymptomatic, benign hyperplastic" mediastinal lymph nodes.[13] The majority of patients are asymptomatic, with the tumor presenting as an incidental finding on chest X-ray films or as a long-standing painless mass.[3,4] The diagnosis of angiofollicular lymph node hyperplasia is usually made by direct biopsy of the enlarged node. Therapy for angiofollicular lymph node hyperplasia has included surgical resection, radiotherapy, and corticosteroids.[4,13]

Our patient presented with chest discomfort and bilateral multiple palpable neck masses of one month's duration, and the antero-superior mediastinal masses. He underwent an open thoracotomy with subtotal excision of tumors which were pathologically diagnosed as angiofollicular lymph node hyperplasia. Post-operatively, he was treated with good clinical response with radiation therapy (RT). After RT, neck masses and the mediastinal residual mass disappeared completely. A state of complete remission was maintained for 12 months.

*This paper was supported by 1987 CUMC Clinical-Research-Fund.

CASE REPORT

A-61-year old man was seen first at St. Vincent's Hospital, Suwon, Catholic University Medical College, in June 1986 for the evaluation of the chest discomfort and indigestion of one month's duration and a large lobulated mass at the antero-superior mediastinum was detected by chest X-ray (Fig. 1). The patient was slightly obese with multiple non-tender palpable neck masses bilaterally along the jugular chains. The abdomen was soft without organomegaly or tenderness. There was no neurologic deficits. The past history and the family history were nonspecific. A computed tomography (CT) scan of the chest including neck showed multiple nodular soft tissue masses with moderate contrast enhancement (Fig. 2).[14] Bone marrow biopsy revealed normal morphology. A lymph node biopsy of neck mass showed the pattern of suspicious malignancy without discernible primary site. And the patient was transferred to Kangnam St. Mary's Hospital, Catholic University Medical College for further evaluation. An open thor-

acotomy was carried out with subtotal excision of the mediastinal tumors. The histological diagnosis was AFLNH (Fig. 3). The patient was treated with external irradiation with 6 MV X-ray from post-operative 20th day on the entire neck and thoracic area (Fig. 4). A cumulative dose of 2,550 cGy/15fx/21 days was delivered for the control of inoperable

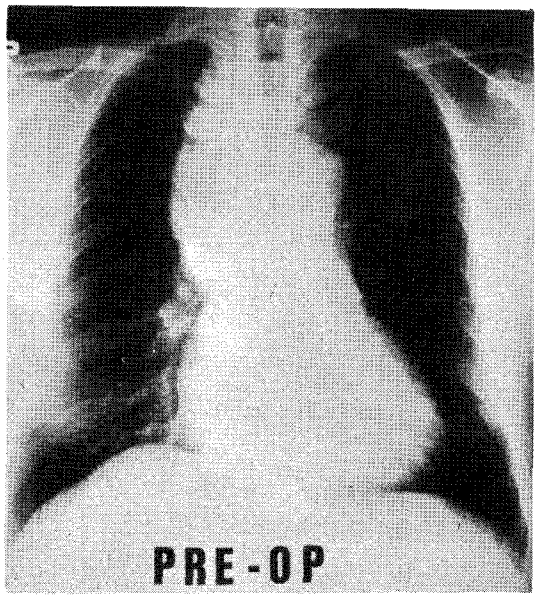

Fig. 1. Chest PA showing a large lobulated mass at supero-anterior mediastinum.

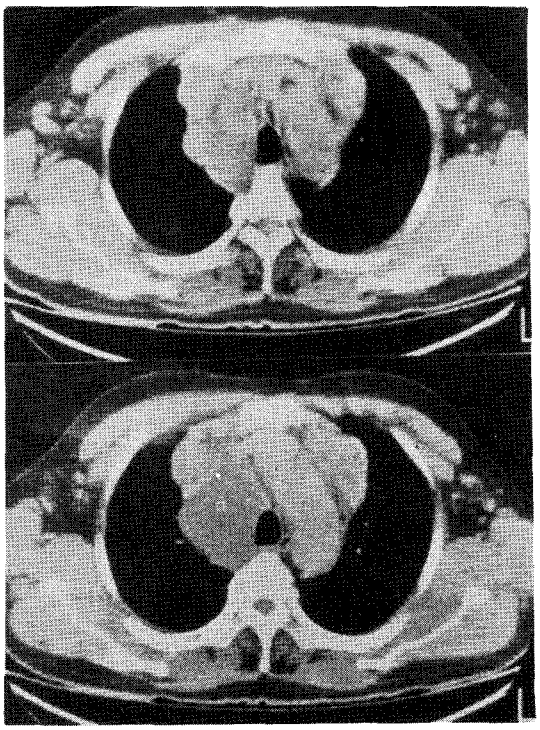

Fig. 2. Upper : Pre-contrast CT showing that multiple nodular soft tissue masses are aggregated and well demarcated from adjacent aorta and trachea. Bottom : Post-contrast CT shows that these masses are moderately (50–70HU) enhanced.

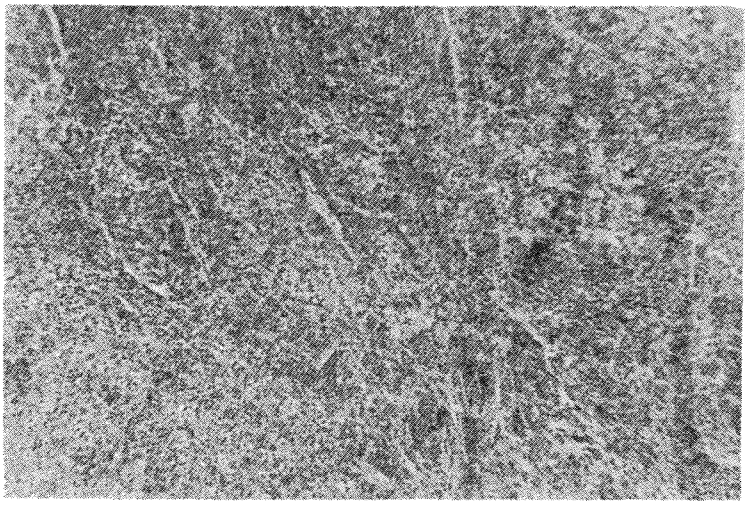

Fig. 3. Lymphoid follicles are greatly increased in numbers. Concentric cuffs of lymphocytes surround the germinal centers. (HE stain, x100).

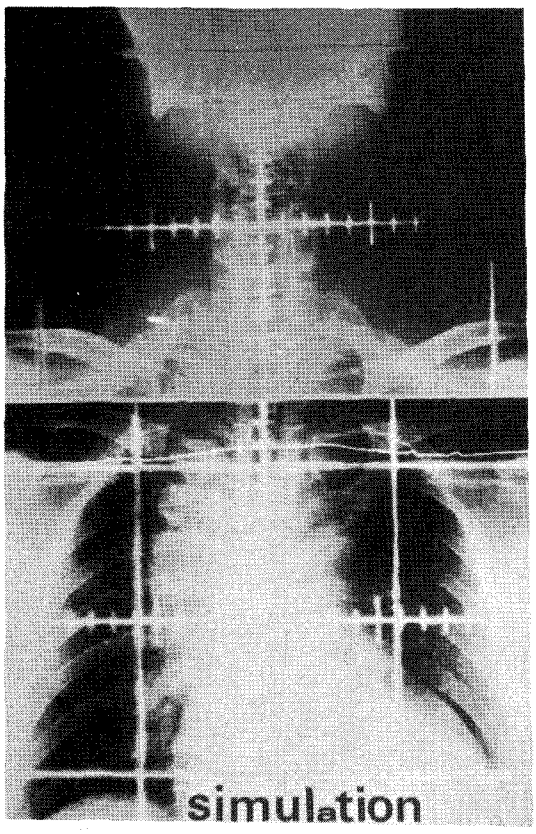

Fig. 4. Post-operative radiation therapy on the residual mediastinal mass (metallic clips) encompassing entire neck likewise as modified mini-mantle field, daily 170 rad, 5 fractions a week, up to 2,550 rad/15fx/3 weeks.

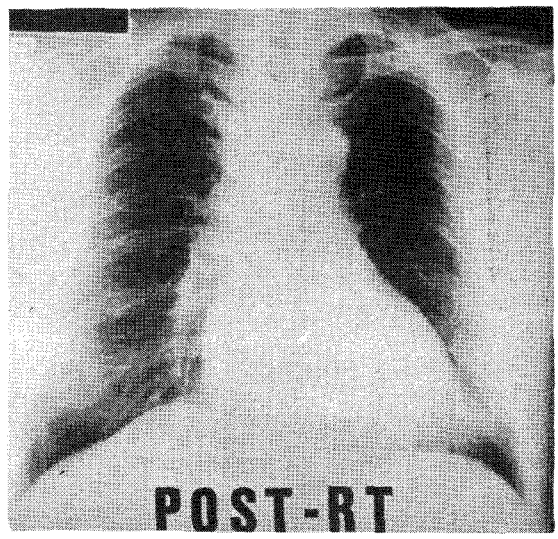

Fig. 5. The mediastinal masses were almost regressed.

residual masses, in which surgeon left surgical clips (Fig. 5). Laboratory data were hemoglobin 13.6 g/dl, leukocyte count 4,500/mm^3, and platelet count 7,000/mm^3. The bleeding time was 4 min 30 sec, and the ESR was 75 mm/hr. The patient was given platelet-rich plasma and steroids, and then the platelet count returned to normal range. After completion of therapy, the bulging neck masses were not palpable clinically and the mediastinal mass was not discernible by follow-up X-ray study. He is well after 12 months without evidence of recurrence. All tumor markers (beta-HCG, alpha-FP, CEA) were also within normal limits.

DISCUSSION

AFLNH is considered to be benign reactive lymphoid proliferation. It has been described by a variety of names including angiomatous lymphoid harmatoma, giant lymph node hyperplasia, lymphoid harmatoma, and benign giant lymphoma.[1~7] Keller and colleges[2] described two histological types, the hyaline-vascular and the plasma cell. The hyaline-vascular type is characterized by a prominent vascular proliferation in the interfollicular areas and hyalinization of follicular vessels. The follicles show a tight onion-skin layering of small lymphocytes. This subtype comprises 90% of the cases, is virtually always localized, and is infrequently associated with systemic symptoms. The plasma cell type is characterized as a diffuse plasma cell proliferation in the interfollicular areas and inconspicuous hyaline-vascular changes in the follicles. This subtype comprises about 10% of cases and may present a systemic manifestations such as generalized lymphadenopathy, macrocytic normochromic anemia, thrombotic throm-bocytopenic purpura, hypergammaglobulinemia, and other associated systemic symptoms which include fever, elevated erythrocytes sedimentation rate, growth retardation and the nephrotic syndrome.[2,4] Although the lesion is usually solitary, multicentric disease can occur. Multi-centric giant lymph node hyperplasia may terminate in malignant lymphoma.[5] In most cases, the surgical resection of the lesion is the curative treatment, and after complete excision of the lesion, these symptoms are resolved. Radiotherapy is resevered for advanced, unresectable lesions. Keller et al[2] repor-

ted four patients with the hyaline-vascular varient who were treated with radiation of total dose of 1, 800 to 4,300 cGy. None of these experienced a response to radiation therapy, and all tumors subsequently required resection. Weisenburger et al[8] and Caba and colleges[12] reported two patients with one plasma cell type and the other hyaline-vascular type, respectively. They were treated with the total dose of 2,700 and 4,500 cGy. One patient developed regrowth 10 months after therapy, and the other showed no response to radiotherapy. Nordstrom et al[9] and Emson[10] and Fitzpatrick and Brown[11] reported three patients who were treated successfully with the total dose of 2,700 to 4,500 cGy. All patients remained well without evidence of disease from 8 months to 5 years after radiotherapy. Our case also showed complete remission after subtotal excision of lesions followed by postoperative adjuvant radiotherapy with a moderate dose for 12 months after initiation of radiotherapy.

REFERENCES

1. **Castleman B, Ivers L, Menendez V:** Localized mediastinal lymph node hyperplasia resembling thymoma. Cancer 9:822-830, 1956
2. **Keller AR, Hochholzer L, Castleman B:** Hyalinevascular and plasma-cell types of giant lymph node hyperplasia of the mediastinum and locations. Cancer 29:670-683, 1972
3. **Hineman VL, Phyliky RL, Banks PM:** Angiofollicular lymph node hyperplasia and peripheral neuropathy association with monoclonal gammopathy. Mayo Clin Proc 1982; 57: pp379-382
4. **Stokes SH, Griffith RC, Thomas PRM:** Angiofollicular lymph node hyperplasia (Castleman's disease) associated with vertebral destruction. Cancer 56:876-879, 1985
5. **Kessler E:** Multicentric giant lymph node hyperplasia. Cancer 56:2446-2451, 1985
6. **Rywlin AM:** Hemopoietic system. In Kissane M, Anderson WAD, (ed.) Anderson's pathology. C.V. Mosby Company, Vol. II. 1985; pp1299-1300
7. **Olscamp G, Weisbrod G, Sanders D, Delarue N, Mustard R:** Castleman's disease: Unusual manifestations of an unusual disorder. Radiology 135:43-48, 1980
8. **Weisenburger D, DeGowin R, Gibson D, Armitage J:** Remission of giant lymph node hyperplasia with anemia after radiotherapy. Cancer 44:457-462, 1979
9. **Nordstrom D, Tewfik H, Latourette H:** Plasma cell giant lymph node hyperplasia responding to radiation therapy. Am J Roentgenology 130:169-171, 1978
10. **Emson H:** Extrathoracic angiofollicular lymphoid hyperplasia with coincidental myasthenia gravis. Cancer 31:241-245, 1973
11. **Fitzpatrick P, Brown T:** Angiofollicular lymph node hyperplasia. Can Med Assoc J 99:1259-1262, 1968
12. **Caba A, Stein R, Sweet D, Variakojis D:** Multicentric giant lymph node hyperplasia. Am J Path 69:86-90, 1978
13. **Passero MA, Calabresi P:** Neoplasms of the mediastinum and mesothelioma. In Calabresi P, Schein PS, Rosenberg SA, (ed.) Medical oncology 1985; pp774
14. **Han H, Kim KK, Im JG, Han MC:** CT findings of Castleman disease (giant mediastinal lymph node hyperplasia). Journal of Korean Radiology Society, 22:738-742, 1986

국문초록 =

혈관여포양 임파선 증식증

—1예 보고—

가톨릭의과대학 강남성모병원 방사선치료실

길학준 · 오윤경 · 윤세철
신 경 섭 · 박 용 휘

혈관여포양 임파선증식증(angiofollicular lymph node hyperplasia)은 원인 미상의 드문 양성 질환으로서 대부분 흉곽내에 발생한다. 진단은 임상증상, 이학적 검사 그리고 X-선 촬영을 비롯한 각종 영상술에 의하며 여러 다른 질환과 감별진단이 어렵다. 대개 수술적 적출에 의한 조직학적 검색으로 확진이 된다. 조직학적으로는 두 가지로 분류되며 하나는 초자양혈관형으로서 여포내에 혈관이 증식되고 초자양화(hyalinization)가 일어나는 것이 특징이다. 90%가 이에 속하고 다른 하나는 형질세포형으로서 10%를 차지한다. 후자는 여포간 조직내에 형질세포가 미만성으로 증식되고 신증후군, 발열, 빈혈, 적혈구침강속도 증가, 고감마글로블린혈증, 저알부민혈증 등의 변화가 있는 점이 전자와 다른 점이다. 치료는 주로 수술적 제거술에 의하며 수술로서 절제되지 못하는 것은 방사선 치료에 의하게 된다.

저자들은 경도의 흉부불쾌감과 경부 임파절 종대를 보인 61세 남자에서 전상종격동 종괴를 수술적 부분제거후 외부 방사선 치료를 실시하여 완치된 1예를 경험하였기에 보고하는 바이다.