Malignant Small Cell Tumor of the Thoracopulmonary Region

— Report of One Case and a Review of the Literature —

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The malignant small round cell tumor of the thoracopulmonary region was described by Askin in 1979 and called as Askin Tumor.

The Askin tumor is a rare, arises from the soft tissues of the chest wall or peripheral lung which is predominantly in younger generation.

Clinical and pathologic appearance were very similar to Ewing's sarcoma and rhabdomyosarcoma but when examined it by electron microscopy, there was some different in morphology.

The tumor tended to recur locally and did not seem to disseminated widely but the median survival was only 8 months.

Key words: Askin tumor, Combination radiotherapy and chemotherapy.

INTRODUCTION

The malignant small cell tumor of the thoracopulmonary region was first described by Askin[1] in 1979 with 20 cases and in 1985, additional 10 cases of Askin tumors were reported by Fink.[2]

The Askin tumor is a rare, malignant small cell neuroepithelioma that arises from the soft tissues of the chest wall or peripheral lung and is seen predominantly in children and young adults.[1]

The tumor tended to recur locally and did not seem to disseminate widely but the median survival was only 8 months.[2]

Histopathologically, the tumor consists of:

1. compact sheets of cells
2. a nesting arrangement of cells with an intervening fibrovascular stroma
3. serpiginous bands of cells with necrosis.

Although its appearance by light microscopy is similar to Ewing's, Rhabdomyosarcoma, neuroblastoma and malignant lymphoma.[3-6]

The Askin tumor is a distinct entity.

One patient has been treated with combination radiotherapy and chemotherapy and 5 months later, multiple lung nodules developed.

CASE REPORT

A 24 year old man was referred to our department with relatively sudden onset(several days) of pain on the left side posterior chest wall just below the spine of the scapula area.

Physical examination, a soft tissue mass was palpated over the left subscapula area extending to the lateral chest wall which was mildly tender to palpation.

There was no palpable lymphadenopathy or organomegaly. Full blood count, liver function tests were normal.

Chest x-ray(Fig. 1) was obtained and showed a large soft tissue mass on the left upper-middle lung with 5th rib destruction.

Computerized chest CAT scan(Fig. 2) was taken and revealed soft tissue chest wall mass which had an associated pleural and extrapleural mass with 5th posterior rib destruction, but there was no

pleural effusion, lung nodules and/or mediastinal lymphadenopathy.

Needle aspiration was performed and initially read as lymphoma or Ewing's sarcoma. However, biopsy specimen revealed small, round cell tumor (Fig. 3).

Total body scan(Fig. 4) was done and shows increased uptake on the soft tissue mass area of the chest wall and no other uptake was seen.

Combination radiotherapy to the chest wall mass and chemotherapy was administered.

Radiotherapy was delivered to a total tumor dose of 8,000 cGy/8 weeks via Co-60 teletherapy unit with opposed oblique ports to the tumor bearing area of the chest wall.

At 5,000 cGy/5 weeks, follow up chest CAT scan (Fig. 5) was done and the field was reduced according to the finding of CAT scan.

During the radiotherapy, the patient tolerated his treatment extremely well and the skin over the treatment area appears dry and discolorated.

Chemotherapy was also administered with and after the radiotherapy and consists of VAC regimen (V: vincristine, A: adriamycin, C: cyclophosphamide).

After this, the patient was followed regularly and clinically appears to be local tumor control was very good but he developed small multiple lung nodules on the right lung, 2 months after completion of radiotherapy.

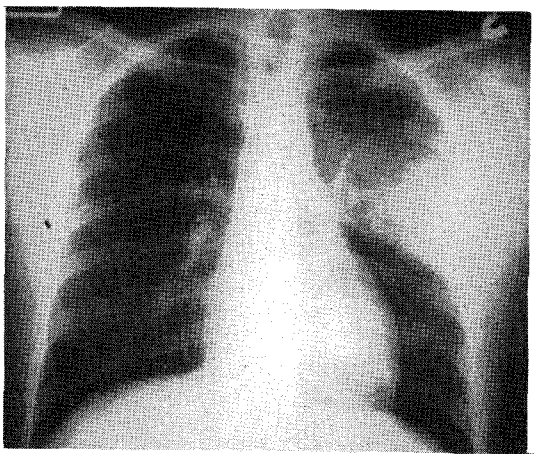

Fig. 1. Chest pA: Soft tissue mass of the left upper Mid lung with 5th Rib dectruction.

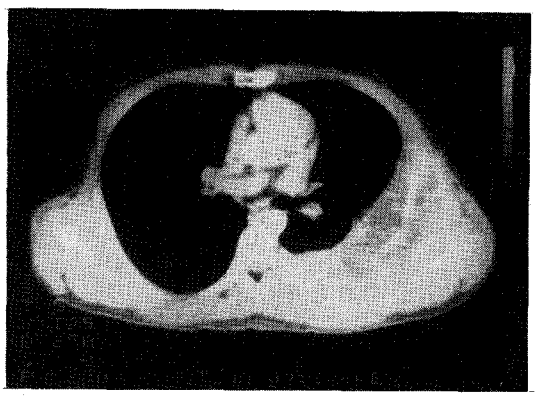

Fig. 2. CAT scan: Soft tissue chest wall mass.

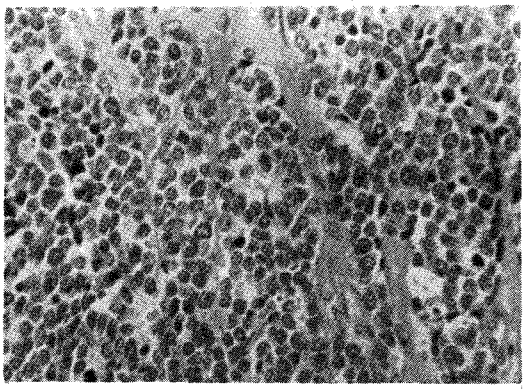

Fig. 3. Round small cell with pseudorossetes and fibrous septa.

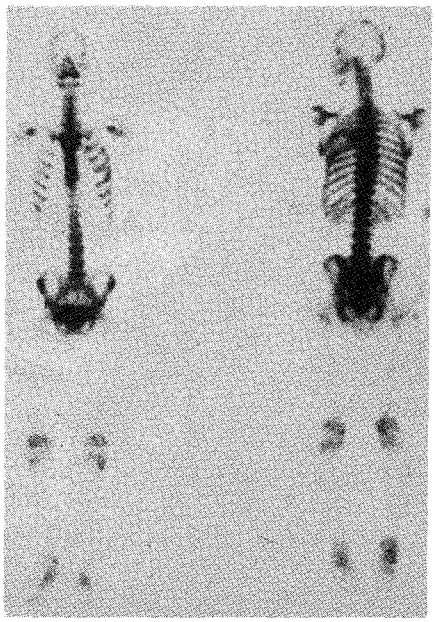

Fig. 4. Total body scan.

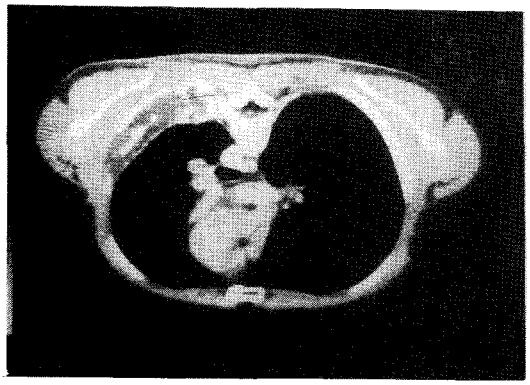

Fig. 5. Follow up chest CAT scan at 5.000 cGY

Therefore, further aggressive chemotherapy was given. However, the pulmonary nodules tend to increase in size inspite of intensive chemotherapy.

1500 cGy/2 weeks of palliative irradiation to the right lung was given.

Patient remains with disease 9 months from diagnosis.

DISCUSSION

Neoplasms of the anterior and posterior chest wall present diagnostic and therapeutic problems and are infrequent in young and have been reported to about 22(1.8%) out of 1,168 children with malignant solid tumors.[7]

The treatment plan for these chest wall tumors varied with the resectability of the tumor, but most combination of irradiation therapy to the primary site and chemotherapy was given.

The chemotherapy was given generally as combinations of drugs that most frequently included cyclophosphamide, vincristine and actinomycin D. Some times, methotrexate, 5-fu, adriamycin and dacarbazine were combined.

The Askin tumor is a distinct pathologic entity, However, histologically, these neoplasms usually makes difficult to diagnoses.

The radiographic appearance of the Askin tumor is also nonspecific and its pattern of metastases is similar to that of some other small cell neoplasms.

Usually, primary treatment failure is recurrent thoracic disease with direct extension into the pleura, lung or the development of pulmonary nodules.

In our patient, we found that the localization of the primary tumor to the thoracopulmonary region as well as the initial recurrences to the pulmonary nodules without widespread dissemination would seem to distinguish this lesion from some of the other malignant neoplasms in the younger age group.

An embryonal rhabdomyosarcoma, Ewing's sarcoma, neuroblastoma and reticulum cell sarcoma were the primary considerations in diagnosis, However, the Askin tumor were lacked the alternating areas of cellularity and myxoid changes when its were examined by electron microscopy.

Pathologically, our patient was most closely resembles of Ewing's sarcoma but the clinical course was quite different than the usual Ewing's sarcoma with its widespread involvement of other bones.

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=국문초록=

흉폐부위에 악성소세포종

—증례보고—

고려대학교 의과대학 치료방사선과, 진단방사선과*

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흉폐부위에서 발생하는 악성소세포종양은 1979년 Askin이 보고했고 이를 Askin종양이라고 부른다.

Askin종양은 상당히 희귀하며 흉벽에 연조직이나 폐가장자리에서 잘 생기며 특히 젊은 층에 호발한다.

임상적, 병리학적으로 Ewing's sarcoma와 Rhabdomyosarcoma와 상당히 흡사하나 정밀한 전자현미경 관찰시 병리학적으로 별개에 종양으로 간주되었다.

Askin종양은 국소재발이 많고 전신으로 잘 터지지 않는 경향이 있으나 생존율은 상당히 나쁜 것으로 되어 있다.

임상적으로나 병리학적으로 Askin종양에 해당되는 24세 환자를 치료결과와 더불어 보고한다.