INFILTRATING MALIGN MESENCHYMOMA ARISING FROM MEDIASTINUM, A CASE REPORT

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SUMMARY : Mediastinum is a rare location for malign mesenchymoma of the nerve sheat (malign schwannoma) though it can be seen everywhere nerve tissue is found.

For this reason, a malign mesenchymoma case which is located in the anterior mediastinum is reported here.

***Key Words** : Malign Mesenchymoma, Mediastinum.*

INTRODUCTION

Malign mesenchymoma of the nerve sheat was first defined by Stout in 1948 (21). It is frequently seen in the retroperitoneal region and thigh and less frequently in the head, neck and kidney mesentery (9, 17). It is a kind of soft tissue tumor, consisting of two or more histological structures of mesenchymal origin. Histologically, it contains rhabdomyosarcoma, osteosarcoma and condrosarcoma structures (6, 10, 19). Later, it was shown by Harkins and Reed that it can also originate from nerve sheat (8). It may have various prognosis according to its dominant histological component (6).

A case of malign mesenchymoma of the lung detected by bronchoscopy has been reported. The origin of malign mesenchymomas located in the lung tissue is not known definitely, whereas the origin of mediastinal cases is the schwannien cells (10).

CASE REPORT

O.D.A 59- year old male patient with the protocol number : 5328/92 admitted to the hospital with the complaints of hoarseness, anorexia and weight loss, had a mediastinal mass lesion detected by a chest X-ray study. Patient and family history were of no diagnostic value and he was a non - smoker.

Physical examination revealed normal blood pressure and normal pulse rate (100/80 mm Hg and 70 beats / minute regular, respectively) and no pathologic physical findings. Routine laboratory analysis were normal, too.

Laboratory analysis : Sedimantion rate : 100 mm/hr, RBC count : 3.2.80x106, WBC count : 9200. Urine examination; microscopically 7-8 leukocyte, hyaline cylinders was detected. When he was admitted to the hospital, his fasting blood sugar was 114 mg/dl, later on in the follow up period in the hospital it decreased to a level of 44 mg/dl associated with a hypoglycemic crisis. Other routine biochemical and haemalogical laboratory analysis had

normal results.

In his P-A Chest X-ray, left hemidiaphragma was elevated in diameter and in the apex of the left lung, a mass of about 2 cm with regular boders superimposed on clavicula was detected (Fig 1). On CT examination of the chest, in the left upper mediastinum a solid mass with irregular border extending from superior apertura of the chest to arcus aorta was detected. The apicoposterior and anterior segments of the superior lobe of the left lung were infiltrated (Fig 2-3).

Fig - 1 : In P-A X Ray, left diafragma was seen higher than normal position. In the left lung apex : there was about 2 cm diameter smooth limited, round lesion that superposed on clavicula.

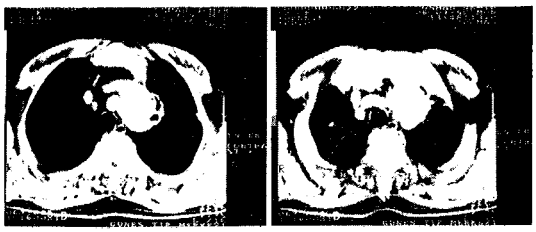

Fig - 2 and 3 : On thorax CT, at superior mediasten, there was unlimited solid tumoral invasion into arteria carotis and arteria subclavia in the left side that extented from apertura thoracica superior to the arcus aorta. In the same time this tumoral mass had invasion into apicoposterior and anterior segments of superior lob in the left lung.

The bronchoscopic examination revealed no endobronchial lesion. At the thoracotomy surgeons found a mediastinal, infiltrating, seemingly malignant lesion and removed it as much as possible. The study of biopsy specimen resulted in malignant mesenchymoma (Specimen no : 2584/92).

Pathology report : During histological analysis of the material, a tumoral structure of malignant appearance located in some areas of lung tissue (Fig 4) and fat tissue of chest wall (Fig 5) was detected. Besides diffuse infiltration of the tumor into the adjacent structures, it was seen to cause tumoral trombi in the vessels (Fig 4, 6). Tumoral mass is mostly composed of spindle cells with wavy contours un-

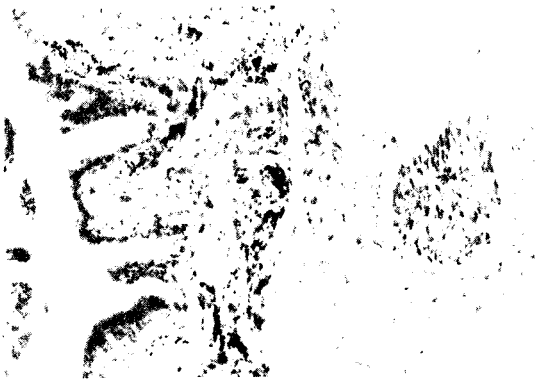

Fig - 4 : In the lung, under the bronchus an arter which is partially obstructed by a trombus of tumor has been demonstrating.

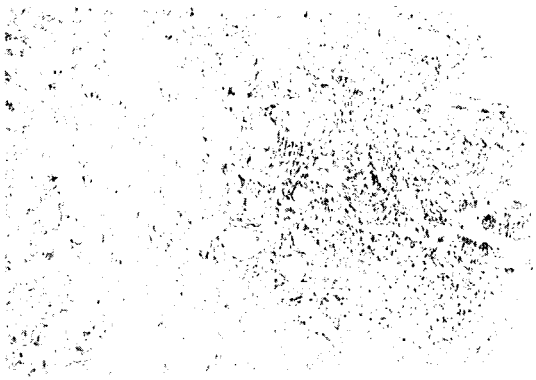

Fig - 5 : Tumor mass is composed of spindle cells showing invasion mediastinal and thoracal fat tissue. In figure, honey comb appearence of tumor is penetrating into fat tissue.

Fig - 6 : There is tumoral trombus in the vein.

der going atypical and frequent mitosis, it is also characterized by large areas of necrosis and showed high grade malignant mesenchymal tumor histology. Spindle cells, in some areas, form palisade formation around necrotic area, as the characteristical finding in malignant schwannoma (Fig 7). In tumoral mass, besides lung and bone tissue, there was small chondroid tissue islands, having the chondrosarcomatous histology with atypical nuclei and mitoses.

Fig - 7 : Lung tissue, central with pallisading tumor cells

After about 1.5 months of operation, the patient was given 3 courses of chemotherapy consisting of endoxan, vincristine and actinomycin with intervals of one month. After the 3 rd-course of chemotherapy, the patient was reviewed by P-A Chest X-ray which indicated progression of the lesion (Fig 8) and the patient's performance status was worse. It was thought that he would not be able to tolerate chemotherapy and therefore supportive measures were taken into consideration.

Fig - 8 : After treatment in the P-A X ray, there is a progression of the lesion.

DISCUSSION

Malign mesenchymoma is mostly seen in the fifth decade. It is rarely seen in children and adults (13, 14). But the in childhood, it is more often in the boys (sex ratio is 2/1). In adulthood there is no significant sex difference (8). Environmental, genetic factors and trauma are thought to play some roles in the etiology of malignant mesenchymoma (6). It can also arise from radiation exposure of burned tissue (7, 21). Histologically, it consists of simple fibrosarcomatous osteogenic and rhabdomyosarcomatous elements which orginate from soft tissues (1, 11). Clinically, especially the patients with mesenchymoma containing nerve cells, have prepedance to hypoglycemia. Hypoglycemia is thought to be due to elevated insulin levels which in turn decreases after surgical intervention (15).

In our case, the patient is 59 years old and male. In the preoperative period, he had undergone frequent hypoglycemic attacks, in spite of negative history for diabetes mellitus and after operation hypoglycemic attacks subsided.

The tumor has tendency to invade surrounding tissues and it frequently relapses and metastasizes (9, 12, 21).

Two cases of malignant mesenchymoma, which can arise from every location where nerve tissue is present, were reported that were located in the lung and bronchi of adults (10, 20) and in the lung and mediastinum of children (2). In most cases of mediastinal involvement the origin of the tumor cannot to be found. But it probably arises from soft tissue of mediastinum and invades lung tissue (2).

In soft tissue tumors, in order to diagnose a malignant schwannoma. There should to be two groups of criteria :

1. Criteria suggesting malignancy : In soft tissue tumors malignancy criteria changes according to histological type of tissue, but generally accepted measures are large areas of necrosis, frequent and atypical mitoses. They are also measures of high grade besides malignancy. Clinical findings and surgical evaluation of our patient, large necrosis, frequent typical and atypical mitosis, proves malignancy.

2. Criteria suggesting that origin of tumor is nerve sheat (schwannoma) : Some outhors suggesting few strict criteria about that subject. According to this group of researchers, to show whether a tumor is a schwannoma or a malign schwannoma :

a. Presence of neurofibromatozis

b. Presence of plexiform neurofibroma which is previously extracted or with tumor tissue

c. Demonstration of electron microscopic or immunohistochemical signs of schwannian origin.

d. Demonstration of tumor which is originated in a major nerve.

The ones who have wider criteria pointed out that some signs on the light microscopic level are characteristic such as in our case palizating around necrosis is an important feature (6, 8). In our case clinically there is no neurofibromatozis or neurofibroma, but pathological findings support diagnosis of malign mesenchymoma.

It is very well known that other than fibrosarcomatous forms of malign schwannoms, there is heterogen forms which contain areas similar to soft tissue tumors. Among these, emergence of rhabdomyoblastic component in tumor tissue is often seen (malign triton tumor) (1). But osteosarcomatous, chondrosarcomatous and other malign mesenchymal components can be seen in tumor tissue. Also glandular epithelium is found very rarely. In our case, we think that due to presence of two different components chondrosarcomatous and fibrosarcomatous, malign mesenchymoma name is suitable.

There is no lymphatic metastases of malignant mesenchymoma practically. Whereas, as pointed in our case, direct invasion to great vessels or to lung paranchyma can be observed (Fig 4-6). This finding is generally accepted as a criterion for a high - grade malignancy and metastasis are expected (6, 8).

Surgery is the treatment of choice in malign mesenchymoma (11, 18). Generally it is accepted as a soft tissue tumor and after surgery, chemotherapy and radiotherapy are given. With this combined therapy, cases with successful results are reported (14, 15). But in practice the success of surgical therapy varies according to the location, size and the dissemination of the tumor. But despite these, more agressive surgery is also advised (22).

A malignant mesenchymoma found on the chest wall was given vincristin sulphate, cyclophosphamid, dactinomisin as chemotherapy after surgical intervention and in the follow up period of 44 months no relapse was detected (14).

A malignant mesenchymoma case found on the right thigh was given RT before surgical procedure and vincristin, actinomycin-D, cyclophosphamid and doksorubicin was used alternatively on three weeks intervals. After this treatment, a disease - free period of 17 months was gained (18).

Chemotherapy periods depends on the extend of surgical excision. After complete excision, treatment has to last for 6 weeks whereas in partial excisions it has to last for 1 year. After operation if there is a tumoral residue, firstly RT has to be given (14).

As malignant mesenchymoma is a rare tumor which doesn't respond to vast chemotherapeutic treatment. But recently mitomisin, doksorubucin (Adriamisin), cisplatin (MAP), mesna, ifosfamid, doksorubucin (Adriamisin), DTIC (MAID), ifosfamid, etoposide, mesna like combined chemotherapy is advised as in the advanced cases (3, 4, 5). According to some authors, new and more effective chemotherapeutics can be used before surgical treatment (18).

Lung is the most common site of metastasis of malignant mesenchymoma. In our case, there was no metastases except the invasion of the lung.

Though prognosis depends largely on the histological type, it is commonly bad. Prognosis is better in the liposarcomatous type rather than the rhabdomyosarcomatous type (1, 14, 17). In the literature, a case containing rhabdomysarcoma and chondrosarcoma components were found at the necropsy and the tumor was found to invade the thorax wall totally. A similar case consisting of the same histological types and location was reported to survive 16 months (16).

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