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Mediastinal Myxoid Liposarcoma with Intrapericardial Involvement and Large Pericardial Effusion

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ABSTRACT

Liposarcoma is the name given to a group of soft tissue sarcomas (STSs) with adipocytic differentiation. As a group, liposarcomas are the second most common STSs in adults. In 1951 Kozonis et al published that in the English language only four cases of liposarcomas originating in the mediastinum had been described. Primary mediastinal liposarcoma is an uncommon neoplasm of intrathoracic origin. We present the case of a 47-year-old woman diagnosed with a large mediastinal mass with intrapericardial invasion and massive pericardial effusion; biopsies showed a mediastinal liposarcoma.

CASE REPORT

A 47-year-old female presented to our hospital with a 2-month history of retrosternal-epigastric pain and a ten-pound weight loss in a 12-month period. Physical exam was remarkable for tachycardia, jugular venous distention, muffled heart sounds, and lower extremity edema. An EKG was done revealing low voltage and sinus tachycardia (Figure 1, A), and an echocardiogram showed a large pericardial effusion with free floating fibrin (Figure 1, B) (Video). With these results, a contrast CT of the chest and abdomen was done showing a large anterior mediastinal mass (11 × 8 cm), important pericardial effusion, and bilateral small pleural effusions (Figure 2A-B). The abdomen was normal. The patient was taken to the operating room and a small antero-lateral thoracotomy was performed, and a pericardial window drained 1000 mL of sanguineous fluid and a large amount of fat-gelatinous nodules (Figure 2C) (Video). Through this incision and with video assistance of a thoracoscope, multiple biopsies of the mediastinal mass were taken (Video). A chest tube was left in place, and the patient was taken to the recovery room and later to the surgical ward. Pathology results from the mediastinal mass and free floating pericardial fat-gelatinous nodules were positive for a myxoid liposarcoma (Figure 2C). The patient was taken back to the operating room and through a median sternotomy, surgical debulking of a friable mass with partial pericardiectomy was performed. We were unable to fully resect the tumor due to intrapericardial invasion with aortic and left pulmonary artery involvement (Video). Her postoperative course was uneventful; she was referred to oncology and radiation therapy (RT) for adjuvant treatments.

DISCUSSION

Primary mediastinal liposarcoma is an uncommon neoplasm of intrathoracic origin [Barbetakis 2007]. In 1951 Kozonis et al published that in the English language only four cases of liposarcomas originating in the mediastinum had been described [Kozonis 1951]. Liposarcoma is the name given to a group of STSs with adipocytic differentiation. As a group, liposarcomas are the second most common STSs in adults [Katz 2012].

The predominant finding of mediastinal liposarcoma on conventional chest radiography is usually a widened mediastinum. On CT, the appearance of mediastinal liposarcomas varies from a predominantly fat-containing mass to a solid mass [Barbetakis 2007; Eisenstat 2000].

Surgical removal is the optimal treatment for a mediastinal liposarcoma, as in other sites. If the entire tumor cannot be resected, surgical debulking often results in symptomatic relief. Radiation therapy (RT) and chemotherapy may be added as adjuncts to surgical excision but liposarcomas seem to have low sensitivity [McLean 1989]. In our case complete removal of the mass was not accomplished. For this reason it

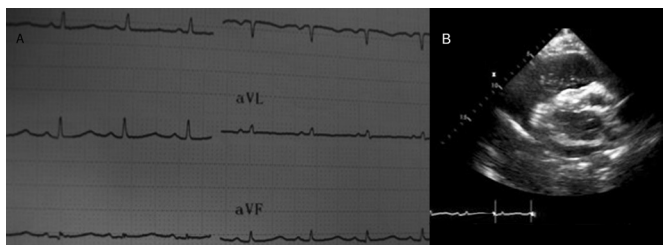

Figure 1 A: The EKG shows low voltage with sinus tachycardia. B: Echocardiogram shows a large pericardial effusion with free floating fibrin.

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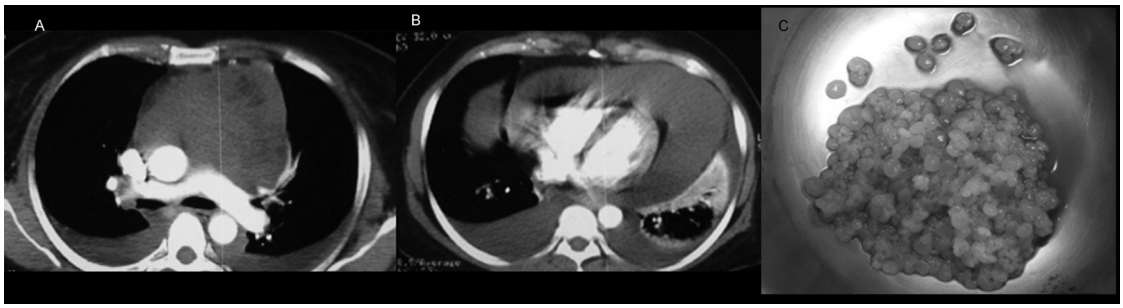

Figure 2 A: The CT scan of the chest reveals a large mediastinal mass. B: CT scan image shows an important pericardial effusion and small bilateral pleural effusions. C: Fat gelatinous nodules drained during surgery (pericardial window).

was recommended to the patient to continue with the adjuvant treatments.

Guadagnolo et al have shown that RT and conservation surgery for localized myxoid liposarcoma provide excellent local control, and in the same study the threshold for the use of chemotherapy was typically a tumor size >10 cm [Guadagnolo 2008]; it is the mainstay treatment of metastatic disease [Katz 2012]. They also demonstrated that in patients with positive margins after conservation surgery, none experienced local disease relapse, suggesting that RT is effective in controlling this tumor [Guadagnolo 2008].

Myxoid liposarcomas are very chemotherapy-responsive using doxorubicin and ifosfamide [Katz 2012] and also quite RT sensitive [Guadagnolo 2008]. Because of the low incidence of mediastinal sarcomas, treatment strategies are extrapolated from sarcomas of other sites of origin [McLean 1989].

The free-floating intrapericardial fat-gelatinous nodules were also liposarcoma tumors; this particular finding was not suspected.

The patient received chemotherapy with doxorubicin and ifosfamide and also radiation therapy. She died 12 months after her initial surgery.

REFERENCES

Barbetakis N, Samanidis G, Samanidou E, et al. 2007. Primary mediastinal liposarcoma: a case report. J Med Case Reports 1:161.

Eisenstat R, Bruce D, Williams LE, Katz DS. 2000. Primary liposarcoma of the mediastinum with coexistent mediastinal lipomatosis. Am J Roentgenol 174:572-3.

Guadagnolo BA, Zagars GK, Ballo MT, et al. 2008. Excellent local control rates and distinctive patterns of failure in myxoid liposarcoma treated with conservation surgery and radiotherapy. Int J Radiat Oncol Biol Phys 70:760-5.

Katz D, Boonsirikamchai P, Choi H, et al. 2012. Efficacy of first-line doxorubicin and ifosfamide in myxoid liposarcoma. Clin Sarcoma Res 2:2.

Kozonis MC, Wiggers RF, Golden HM. 1951. Primary liposarcoma of the mediastinum. Ann Intern Med 35:703-10.

McLean TR, Almassi GH, Hackbarth DA, Janjan NA, Potish RA. 1989. Mediastinal involvement by myxoid liposarcoma. Ann Thorac Surg 47:920-1.