Myxoid liposarcoma of the anterior mediastinum. A case report and bibliography review

ABSTRACT

Background: Soft tissue sarcomas have an incidence rate of 1.8 to 5 cases per year. Of those cases, 50% are located in the extremities and 40% are located in the trunk and retroperitoneum. Primary mediastinal liposarcomas represent < 1% of mediastinal tumors.

Clinical case: A 53-year-old female, native and resident of Tabasco, with a history of anterior mediastinal tumor was treated with resection at the National Institute of Cancerology approximately 16 years ago with histopathological diagnosis of pleomorphic liposarcoma. Her condition began with chest pain, cough and hyaline expectoration, managed as pneumonia in her unit. Other symptoms occurred such as moderate exertion dyspnea and edema of lower limbs. Chest computed tomography documented mediastinal tumor of 9 × 9 cm. The patient was sent to our unit where she was managed with resection.

Conclusion: Liposarcomas represent < 1% of mediastinal tumors. They require long-term monitoring due to the high rate of recurrence after a long disease-free period.

Key words: Myxoid liposarcoma, mediastinal neoplasms.
BACKGROUND

Soft tissue sarcomas are malignant tumors that appear in the mesodermal tissues of the limbs, trunk and retroperitoneum; its incidence is from 1.8 to 5/100,000 cases per year. In the U.S. in the year 2012 there were 11,280 new cases estimated and 3900 deaths due to soft tissue tumors. In Mexico, in 2006, the histopathology registry reported 1595 cases of tumors of the connective and soft tissues, which represented 1.5% of malignant neoplasms. Primary liposarcomas of the mediastinum (considered among the group of the soft tissue sarcomas) represent < 1% of the mediastinal tumors with < 150 cases reported in the literature, of which the majority appear in the posterior mediastinum. Only a few cases are reported to be found on the anterior mediastinum, indicating that these are rare neoplasms. Of the primary mediastinal sarcomas, 9% correspond to liposarcomas with an average age of presentation of 43 years. The majority of the cases manifest with dyspnea, chest pain, cough, constitutional symptoms and, to a lesser frequency, signs of superior vena cava obstruction; 15% are asymptomatic. The recurrences of these types of tumors almost always appear in the first 6 months but can also present even after 20 years of the initial surgery.

CLINICAL CASE

We present the case of a 53-year-old female patient originally from and resident of the state of Tabasco. Her past history reports grade III pleomorphic liposarcoma diagnosed in the National Institute of Cancer at the age of 37 during the course of her third pregnancy at 24 weeks of gestation. During the puerperium she underwent a left posterolateral thoracotomy and teletherapy with a total dose of 56 Gy for invasion to < 1 cm of one of the surgical margins. She had annual follow-up during the last 16 years and her last consult was in 2012 in the National Institute of Cancer.

The current condition began a year earlier with chest pain, cough and hyaline expectoration diagnosed and treated as pneumonia at her assigned hospital (Villahermosa Regional Hospital of Petroleos Mexicanos). Four months prior to admission to our hospital the following complaints were added: dyspnea on moderate effort and lower extremity edema. Imaging studies documented a mediastinal tumor, which was the reason for the patient being sent to our hospital (Central Sur de Alta Especialidad de PEMEX) for scheduled resection of the mediastinal tumor. On admission the patient was found to be in good general condition with ECOG scale of 0, weight 86 kg, and height 1.42 m. Thoraco-abdominal computed tomography showed a rounded, well-differentiated, heterogeneous tumor in the left anteroinferior mediastinum of 91 × 90 × 90 mm in diameter that displaced the heart in a cephalad direction, the stomach in a posterior direction and ipsilateral diaphragm in an inferior direction. The tumor was discretely enhanced after the administration of contrast media with densities between 7 and 23 Hounsfield Units (HU) (Figures 1 and 2). Magnetic resonance imaging reported a left inferior mediastinal tumor (99 × 92 mm) with multiple internal septa and weak enhancement after the administration of intravenous contrast that extrinsically compressed the left ventricle (Figure 3).

Positron emission tomography (PET) was done to rule out distant tumor activity. This was because at the time of admission there was no histopathological report of the initial tumor resected at the National Cancer Institute. The official report indicated intrathoracic tumor activity with probable pericardial and pleural nodes of the esophagogastric junction and retrocrural space without data of distant tumor activity (Figure 4). Laboratory studies done on May 29, 2012 report a blood count with leukocytes 10.37 mil/μl, hemoglobin 15.44 g/dl, hematocrit 48.56%, platelets 240,000 mm³, glucose 93 mg/dl, BUN 22, urea 47.1 mg/dl, creatinine 0.74 mg/dl,
sodium 133 mEq/l, potassium 4.3 mEq/l, chlorine 93 mEq/l, glutamic oxaloacetic transaminase 68 IU/l, glutamic pyruvic transaminase 71 IU/l, and lactate dehydrogenase 541 IU/l. With the imaging results the patient was scheduled for surgery and a median sternotomy was carried out revealing a tumor of 10 × 9 cm (Figures 5 and 6), which was resected and sent to pathology for definitive diagnosis. The histopathological report was low-grade mixoid liposarcoma of 10 cm in diameter with three lymph nodes with mixed hyperplasia. The patient was admitted to the intensive care unit for postoperative monitoring with adequate progress and improvement. She was discharged and referred to the Oncology Department for follow-up.

**DISCUSSION**

Primary mediastinal tumors and anterior mediastinal liposarcomas are rare neoplasms according...
Fig. 4. Positron emission tomography, image in coronal cut. Intrathoracic tumor activity is seen with likely infiltration to the pericardium, pleura, lymph nodes of the esophagogastric junction and retrocrural space, without data of distant tumor activity.

Fig. 5. Median sternotomy where left ventricle is shown (black arrow) to be displaced by a heterogeneous tumor of firm consistency.

Fig. 6. Surgical specimen of firm, ovoid tumor of irregular lobulated surface of 10 × 9 cm at its largest diameters.

to Klimstra et al., ⁶ and are more common at the average age of 43 years. Because the symptoms are nonspecific, the diagnosis is difficult to make. Some manifestations are dyspnea, cough and chest pain.⁷ The predominant findings of these
tumors on conventional chest x-ray are widening of the mediastinum and deviation of the trachea and vessels. Tomography and MRI identify heterogeneous fatty tumors that vary in appearance, depending on their content of fatty tissue, fibrous bands and solid components, with the latter decreasing after contrast administration. Values between 50 and 150 HU on CT support the existence of tissue composed of fat, whereas on MRI the signal intensity is increased in T1 and decreased on T2. Among the differential diagnoses are lipoma, teratoma, thymolipoma, lymphoma and even herniated preperitoneal fat.

The optimal treatment for these neoplasms consists of surgical removal with negative margins. Radiotherapy and chemotherapy can be given as adjuvant therapy; however, it has been reported that these tumors have low sensitivity.

The case presented is of interest because of the patient’s past medical history and the possibility of comparing on the different imaging studies the characteristics of this disease, which is not found in any of the literature consulted.

In conclusion, we present the case of a patient with a history of mediastinal tumor resection with histopathological report of grade III pleomorphic liposarcoma. The tumor appeared 16 years after initial treatment of an anterior mediastinal tumor with histopathology report of low-grade mixoid liposarcoma. The symptoms, radiological findings and tumor behavior of our patient coincide with what is reported in the literature. It is of interest for the medical community to stress the need for continuous follow-up of these types of tumors in which the recurrence is high. Although the majority of these recurrences occur during the first 6 months, recurrence could be delayed 5 to 10 years or even according to what was reported by Matsubara et al., after 20 years from the initial surgery, which was implicated due to an incomplete resection.

REFERENCES