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Primary Leiomyosarcoma of the Lung

Leiomiosarcoma pulmonar primario

To the Editor:

Primary sarcomas of the lung are uncommon, representing less the 0.5% of all malignant lung tumours. To confirm the diagnosis, it is necessary to rule out the existence, at the present time or in the past, of sarcomas in other organs.[1] Among the different types, leiomyosarcomas are the most frequent.[2]

Here we describe a 60 year old patient with no history of smoking. She visited doctors at the Hospital de Clinicas Dr. Manuel Quintela with month-long symptoms of cough and general health implications such as weight loss of 5 kg, with the examination confirming a poor general condition, and BMI of 18. She presented with polypnea of 24 breaths/min and, at pleuropulmonary level, dullness to percussion and decreased breath sounds in the middle third of the left hemithorax. The gynaecological examination was normal.

The chest x-ray revealed a homogeneous polylobulated mass in the hilar and parahilar regions of the left lung. Computerized tomography revealed a solid-looking voluminous tumour with a poorly-defined, irregular outline; a region compatible with central necrosis; it contacted with mediastinal vascular structures; multiple ipsilateral nodules which looked secondary. A transparietal puncture biopsy was performed for an anatomical pathology study, and, using conventional techniques, the optical microscope showed malignant neoplasia whose morphology was compatible with a highly necrotic sarcoma. Immunohistochemistry showed an immunophenotype concordant with a mesenchymal tumour with smooth muscle cell lines (fig. 1). The diagnosis was primary leiomyosarcoma of the lung.

Palliative treatment was begun based on analgesics, corticoids, polychemotherapy and anticoagulants due to unresectability because of the size of the tumour and the invasion of mediastinal structures. The patient progressed unfavourably and the progress of the tumour was fast, and she died.

Primary sarcomas of the lung are mesenchymal-type tumours which originate in bronchial wall, vascular or interstitial cells. There are various types, amongst which leiomyosarcomas are the most frequent, followed by fibrosarcomas and hemangiopericytomas.[4]

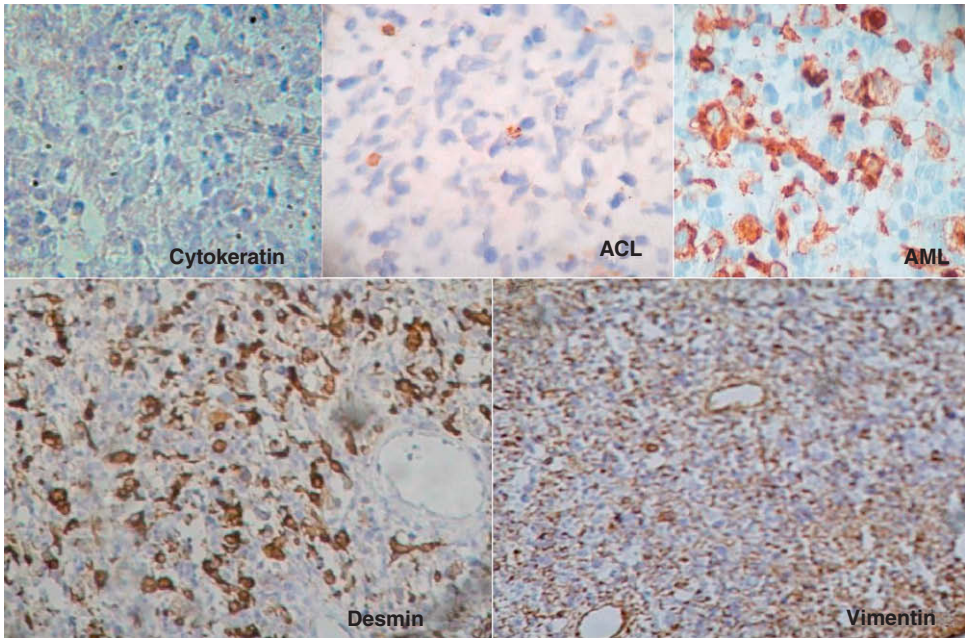

Figure 1. Immunophenotype concordant with a mesenchymal tumour with smooth muscle cell lines (negative cytokeratin, negative common leukocyte antigen in the proliferating cell component and positive in lymphocytes, smooth muscle actin positive in proliferating cell component displaying focal distribution, highly desmin and vimentin positive in the proliferating cell component).

Other less common ones are fibrohistiocytomas, sarcomatoid mesotheliomas, synovial sarcomas, carcinoid tumours, benign pleural fibromas and neurogenic sarcomas. Since Davidsohn reported the first case of leiomyosarcoma in 1907, just over 100 more cases have been published.[3,2] The series published, which in general are formed of 12-14 patients in 30-year follow ups, show how rare this type of tumours are, not reaching 0.5% of all malignant tumours of the lung.[1] Generally, they affect people over 60 years of age, although some cases have occasionally been described in children.[7]

Patients have little clinical expression. The most common symptoms are cough, chest pain, dyspnoea, haemoptysis, asthenia and general implications.[5] Since these tumours tend to grow quickly and invade other tissues, they are diagnosed at advanced stages, even after metastases have spread hematogenously to the brain, lungs and/or bones.[1]

The treatment providing the best results is surgery. The options are a lobectomy or pneumonectomy, which on occasions can require resection of sectors of the chest wall, diaphragm or vascular structures.[6] Adjuvant radio- or chemotherapy treatment, although not improving survival, is indicated in cases of incomplete resection, technically unresectable tumours and those with a high degree of histological malignancy.[8]

Factors for a poor prognosis have been described: tumour size (over 4-5 cm), the degree of histological malignancy, endobronchial invasion and the impossibility of performing complete resection.[1,6]

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Palliative Thoracocentesis in Low Income Countries

Toracocentesis paliativa en países de bajos recursos

To the Editor:

I read with great interest the paper published recently by Cases et al[1] regarding their experience using a commercial tunnelled catheter in patients with malignant pleural effusion. This paper has shown that once enough experience has been gained using it and if the necessary facilities are available for patients to be handled as outpatients, it is an effective option for controlling recurrent malignant pleural effusion. However, in underdeveloped countries, like ours, it is rather unfeasible to apply this technique given that this pleural drainage device is not readily available and there is limited experience using it. Furthermore, costs associated with maintenance and purchasing specific vacuum bottles are high. These factors mean that chest-tube pleural drainage, repeated thoracocentesis and small-diameter catheters are the main palliative methods for treating patients with symptomatic malignant pleural effusions. In order to reduce patient discomfort and costs derived from hospitalisation and, bearing in mind the high burden of institutional work in our country, we have used a personalised thoracocentesis technique aided by wall suction. Our method follows the same principles used in thoracocentesis, and is preferably guided by ultrasound to locate the worst-affected area. It involves free pleural fluid aspiration using a 3 ml syringe as an adaptor between a fenestrated angiocath and a 6 mm diameter tube (Argyle connecting tube; Kendall-LTP; Chicopee, MA), connected to the vacuum collection system on the wall (fig. 1). The wall suction regulator reduces the high negative pressure to more physiological levels. The suction pressure must be gentle and similar to the flow generated by a syringe during standard thoracocentesis. Optionally, a 3-way stopcock can be placed between the syringe and the catheter so as to provide manual control or to monitor intrapleural pressure with one of the methods described previously. The procedure should be interrupted if the patient suffers oppressive chest pain, dyspnoea or other significant symptoms, even if a substantial amount of pleural effusion remains. One advantage of continuous aspiration is that it minimises the risk of iatrogenic pneumothorax, which in theory could occur, principally during intermittent handling of a syringe. When the liquid has been drained, a guide wire is inserted through the angiocath and using the Seledinger technique, a 20 cm-long triple-lumen central venous catheter (Arrow International, Reading, Pennsylvania, USA) is inserted into the pleural cavity. The catheter can be used to drain pleural fluid on a daily basis in recurrent cases and to perform chemical pleurodesis in the outpatient clinic. The effectiveness of using small-diameter catheters has been shown in previous studies.[2,3] The inconvenience of the increased likelihood of obstruction is outweighed by the fact that it is easy to handle and exchange when necessary. Furthermore, the insertion technique is less painful and thus less local anaesthetic is needed. However, its use can be limited by the presence of a malignant bronchopleural fistula, in which case a large-diameter catheter or chest tube may allow greater flow during the fluid and/or air aspiration. In this case, connecting the chest tube to a urine bag can be useful as it has a valve which stops its contents from backflowing and can be emptied easily.[4] With the techniques described above, our main aim is to propose ideas for improving the quality of life of patients with malignant pleural effusion in centres that have limited resources and whose objective is to offer safe treatment to patients through simple, economical and easily implemented actions, guided by a sense of ethics, humanity and quality.