

Leiomyosarcoma Of The Diaphragm: Two Case Reports

A Louzi, R Benelkhaiat, B Finech, B Belaabidia, M Hakkou, A Elidrissi

Citation

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Abstract

Leiomyosarcoma of the diaphragm is an extremely rare entity. The diagnosis is more difficult in early stages with a very poor prognosis. Surgery represents the only radical treatment of this tumour. We report two cases of leiomyosarcoma of the diaphragm that presented clinically in the form of an epigastric mass. Radiography and computed tomography contributed to the diagnosis. Pathological examination after surgical resection confirmed the diagnosis. The first case died after six months due to tumour recurrence and the second case is still alive with twelve months freedom of relapse and other metastasis. The authors discuss the diagnosis and the therapeutic modalities raised by this entity.

INTRODUCTION

Primary tumours of the diaphragm are rare lesions. Leiomyosarcoma of the diaphragm is exceptional. The first case has been reported by Kirschbaum in 1935 [1]; and so far only ten cases have been reported in the literature to the best of our knowledge. The diagnosis is more difficult in the early stages and the prognosis is very poor. Radical surgical resection remains the treatment of choice; however, the prognosis is very poor.

We present two cases of leiomyosarcoma of the diaphragm that were clinically interpreted as abdominal masses. The correct diagnosis was only achieved during surgical excision and then confirmed by histological findings.

CASE 1

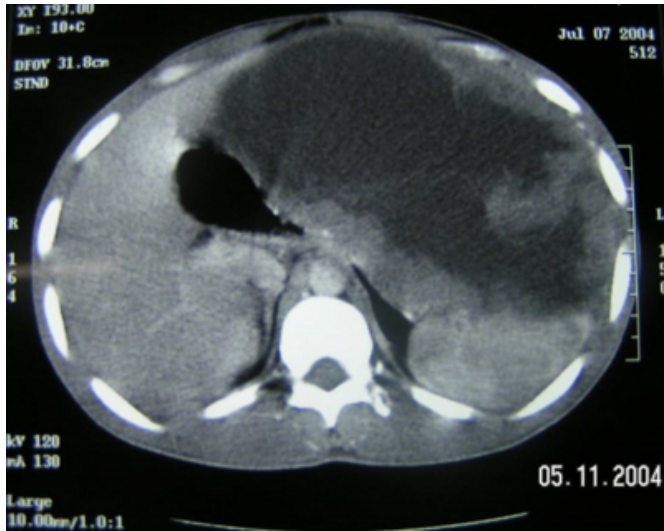
The first case was a young 18-year-old male who was admitted in our department in July 2004 complaining of an abdominal mass for 20 days without any relevant history. On examination, we found an epigastric mass, firm in consistency, deep to the muscle layer and not tender. Abdominal sonography revealed a cystic mass measuring 157 x 148 mm. Abdominal CT-scan showed a huge necrotic, supra-mesocolic tumour displacing the stomach to the right with peripheral irregular thickening (fig. 1). A weak uptake of the contrast material was observed. An upper GI endoscopy was performed which excluded gastric origin of the tumour. Concerning blood tests, we observed marked elevation of serum LDH: 3908 IU/L (normal value 200-400 IU/L).

Surgical exploration for resection by laparotomy revealed a cystic mass of pure diaphragmatic origin not related to the liver or to the stomach. Neither the regional lymph nodes nor the peritoneum had been infiltrated. The tumour mass was resected with a safety margin of about 2 cm separating it from the left diaphragmatic cupula. The diaphragmatic defect was repaired surgically by edge-to-edge sutures with no need of patch closure. Finally, surgical closure was achieved after placing an intercostal thoracic drain.

Macroscopic examination of the specimen revealed a cystic multicentric tumour, composed of whitish nodules with areas of haemorrhage and necrosis. Microscopic examination revealed a malignant fusiform structure formed out of malignant cells with eosinophilic cytoplasm and hyperdense atypical nuclei. This procedure explained the presence of anti-muscle antibodies and anti-HHF35 antibodies. Desmine had not been detected thus eliminating rhabdomyosarcoma. The final histopathological diagnosis was diaphragmatic leiomyosarcoma of grade 3 according to FNCLCC. The postoperative course was uneventful. The outcome was marked by tumour recurrence three months later and the patient died after six months.

Figure 1

Figure 1: CT scan: expansive inhomogeneous tumor mass in the left upper abdomen



CASE 2

A 54-year-old woman was admitted in our department in June 2005 complaining of epigastric painful mass of progressive course, dyspnea and loss of appetite for three months. Past and family history were irrelevant. The patient appeared in good condition. Clinical examination revealed an epigastric, slightly tender mass, firm in consistency, deep to the muscle layer and not freely mobile. Chest examination revealed no abnormal findings. Blood tests were normal and chest X-ray (fig. 2) revealed elevation of the left diaphragmatic copula.

Abdominal ultrasound and CT scan demonstrated an epigastric mass measuring 25cm x 15cm x 13 cm compressing the left lobe of the liver and the stomach. After injection of contrast material, the mass showed a non-homogenous enhancement (fig. 3).

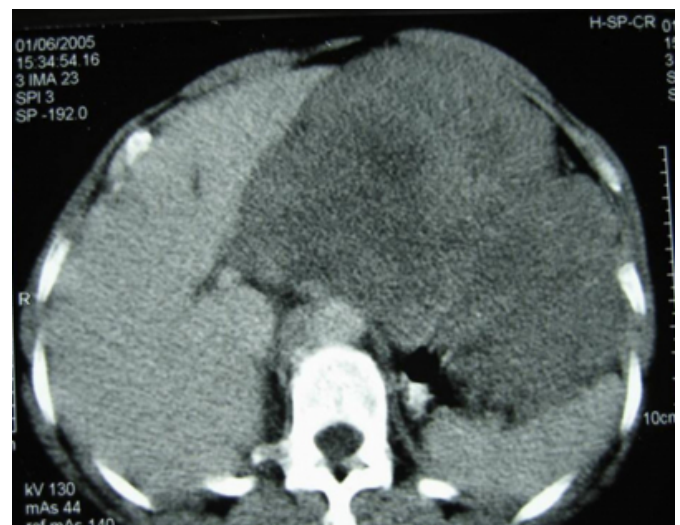
Figure 2

Figure 2: chest radiography showing a mass in the left side of the diaphragm



Figure 3

Figure 3: CT scan: expansive inhomogeneous mass in contact with the liver



A laparotomy was planned. Surgical exploration revealed a firm tumour mass of pure diaphragmatic origin invading the left liver but not related to the stomach or to the spleen. Neither the regional lymph nodes nor the peritoneum were infiltrated. The tumour mass and the left hepatic lobe were resected with a safety margin of about 2 cm separating it from the left diaphragmatic copula (fig. 4). The diaphragmatic defect was repaired surgically by edge-to-edge sutures after placing an intercostal thoracic drain.

Macroscopic examination of the specimen revealed a multilobular tumour, with areas of haemorrhage and necrosis. Microscopic examination revealed a malignant

fusiform structure formed out of malignant cells with eosinophilic cytoplasm and hyperdense atypical nuclei. These findings explained the presence of anti-muscle antibodies and anti-HHF35 antibodies. Desmine had not been detected thus eliminating rhabdomyosarcoma. The final histopathological diagnosis was diaphragmatic leiomyosarcoma.

The postoperative course was uneventful, and the patient was discharged from the hospital on the 11th day. She is free of recurrence or metastasis for more than 12 months.

Figure 4

Figure 4: Appearance of the resected tumour containing part of the left hepatic lobe



DISCUSSION

Primary neoplasms of the diaphragm are rare lesions that are often difficult to assess. Leiomyosarcoma of the diaphragm is an exceptional entity. Leiomyosarcomas of the diaphragm have been previously described in patients with a mean age of 55.4 years [2]. The role of pulmonary diseases in the aetiology of primary tumours of the diaphragm is not clear, but three cases suffered from tuberculosis [3], asbestosis [4] and anthracosilicosis [5], respectively.

Clinical presentations are not specific and depend on localization and volume of the tumours. Diagnosis is more difficult especially in the early stages when a differential diagnosis of intrathoracic and abdominal tumefaction can not easily be made. The most common symptoms when the tumour expands into the thorax are dry cough, when the pleura is irritated, and low chest pain [6].

However, in our cases, clinical manifestation was by abdominal mass, persisting discomfort in the upper abdomen

or gastric complaints resulting from the tumour's expansion into the abdomen [7]. Clinical examination is negative in the absence of either expansion of the tumour to the costal margin or the involvement of a large portion of the thoracic cavity or the abdomen [6].

Abdominal ultrasound and CT scan rarely reveal the diaphragmatic origin of this tumour. On the other hand, according to Blondeel and al. [5], magnetic resonance imaging appears to be more accurate and specific.

Angiography and scintigraphy argued against the diagnosis, suggesting an atypical presentation, but they did not exclude another origin of that mass [6].

The diagnosis of leiomyosarcoma of the diaphragm is principally anatomopathologic. Macroscopically, the tumour could be sessile or pediculated, firm in consistency, whitish in colour and highly vascular. Histopathological examination reveals fusiform cells with pleomorphic nuclei, organised in bundles [8]. The tumour cells are immunoreactive for vimentine and desmine [5]. Leiomyosarcomas metastasize by vascular embolization to the liver, lung, brain and seed to the peritoneal cavity [9].

Total surgical resection remains the treatment of choice and is predictive of a prolonged survival in patients with solitary leiomyosarcoma [5, 6, 9, 10, 11]. Surgical excision must be wide in accordance to the classic oncologic principles. The surgical approach is achieved through thoracotomy and or laparotomy depending on the extent of the tumour.

The place of radiotherapy and chemotherapy is controversial in post-operative management of leiomyosarcoma. Blondeel and al. treated a patient with combination chemotherapy (ifosfamide and adriamycin) for relapse after surgery resulting in very good partial remission and this patient was alive after 51 months [5]. Radiotherapy and chemotherapy containing ifosfamide or ifosfamide and doxorubicin have been used in the treatment of patients with metastatic sarcomas with acceptable toxicity [12]. Cho and al. consider chemotherapy for relapse after surgery or incomplete resection [11].

Little is known about the long-term prognosis of these tumours. The prognosis at long term is not yet established due to the rarity of cases. Patients die after extensive tumour dissemination at a mean of 13 months after the diagnosis [6].

CONCLUSION

Leiomyosarcoma of the diaphragm is an exceptional entity. Diagnosis is more difficult in the early stages and prognosis is very poor. Radical surgery represents the only reasonable treatment and is predictive of a prolonged survival in patients with solitary tumour. Radiochemotherapy appears to have only a palliative purpose for relapse after surgery or incomplete resection.

CORRESPONDENCE TO

Abdoulahed LOUZI (Professor) Faculté de Médecine et de Pharmacie de Marrakech BP 7010 – Sidi Abbad 40000 – Marrakech Tel.: +21210774091 / +21261410400 Email: louzi1@hotmail.fr

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Author Information

A. Louzi

Department of General Surgery, CHU Mohamed VI

R. Benelkhaïat

Department of General Surgery, CHU Mohamed VI

B. Finech

Department of General Surgery, CHU Mohamed VI

B. Belaabidia

Department of Pathology, CHU Mohamed VI

M. Hakkou

Department of Pathology Zohor, CHU Mohamed VI

A.D. Elidrissi

Department of General Surgery, CHU Mohamed VI