Case report

Gigantic primary lipoma of the diaphragm presenting with respiratory failure

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Abstract

Primary diaphragmatic lipomas are rare. A 66-year-old female case is reported, with the unusual presentation of progressive dyspnoea leading to respiratory failure, mainly because of a large fatty mass occupying the base of the left hemithorax. The completely resected mass (16.14.5.5.5 cm) was a primary diaphragmatic lipoma. The literature is discussed. We recommend surgical exploration since the possibility of liposarcoma cannot be excluded. © 1998 Elsevier Science B.V. All rights reserved

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1. Introduction

Lipomas are common, however they are rarely located in the diaphragm. Eighteen cases were reported until 1976 [1]. In a 71-patient series with primary diaphragmatic neoplasms, only 9 were lipomas [2]. Primary diaphragmatic lipomas (PDLs) are usually incidental findings [1]. They occasionally cause manifestations including coughing, dyspnoea, chest pain, heart disturbances or even death from diaphragmatic rupture [1,3,4]. We present the following case of a large PDL, with an unusual clinical presentation.

2. Case report

A 66-year-old white female, ex-smoker, was urgently referred because of respiratory failure with progressive dyspnoea, and left pleuritic chest pain coming on over a period of 3 weeks prior to admission. On clinical examination she was overweight and there was loud stridor and expiratory wheeze; the left base was quiet and dull on percussion; BP was 170/95 mmHg. Other clinical signs were not found.

Past medical history included two operations 2 and 7 years earlier. Both preoperative chest radiographs (CXR) had been reported to be normal at that time, but retrospectively they were evaluated as abnormal. Her mother died aged 39 (laryngeal cancer).

On CXR the left hemidiaphragm appeared markedly elevated, the mediastinum widened and the thoracic aorta unfolded (Fig. 1). Chest and abdomen computed tomography (CT) scanning (Fig. 2) revealed the presence of a 10.15 cm mass at the base of the left hemithorax, homogenous, with attenuation coefficient of adipose tissue (−103 to −121 Hounsfield units (HU)), that had been reported as ‘subdiaphragmatic’. Two smaller ‘subdiaphragmatic’ fatty masses were present on the right side.

ESR was 55 mm/1st h. Arterial blood gases on air were: pO2 = 51.5 mmHg, pCO2 = 49 mmHg, pH = 7.417; base excess = 6.5, bicarbonates = 30.6, saturation = 87.4%. FEV1 = 0.8 l (39% of the predicted value) and FVC = 1.03 l (40%). The rest of laboratory investigations were normal.

The patient was treated with nebulised bronchodilators, oxygen and oral steroids, so that her preoperative condition was optimised. Then a left exploratory thoracotomy was undertaken and a 16 × 14.5 × 5.5 cm smooth, mushroom-shaped, well defined extrapleural mass was found arising from the left hemidiaphragm posterolaterally. It was not adherent to the lung, pericardium or the phrenic nerve. Blood supply was provided by a wide vascular pedicle con-
necting the mass to a locally weakened part of the dia-
phragm. Complete resection of the mass, its pedicle and 
the weakened part of the diaphragm was carried out. The 
diaphragmatic defect (4 × 2 cm) was easily repaired with 
non-absorbable sutures.

Histologically the mass was a lipoma, representing a pro-
liferation of mature adipose cells. The patient recovered 
uneventfully and left hospital 12 days later. She remains 
well 10 months postoperatively; CXR and blood gases are 
satisfactory. Respiratory function tests are significantly 
improved with FEV\(_1\) = 1.82 l (89%) and FVC = 2.34 l 
(91% of the predicted value).

3. Discussion

In 1976 Ferguson and Westcott [1] reviewed 17 PDL 
cases. The male/female ratio was 4:5. All patients were 
above 45 years of age and with a preference to the left 
side (11:7). Fourteen cases (78%) were asymptomatic (inci-
dental post mortem findings), whereas four (22%) experi-
cenced either chest pain or coughing and dyspnoea or fatal 
rupture of the diaphragm at the site of the lipoma whilst 
coronary insufficiency with minimal ECG-changes, disap-
pearing after resection. Ferguson and Westcott [1]suggest 
that trauma may be responsible for the generation of a PDL; 
however this was neither the case in our patient nor con-
firmed in the literature. Finally, bilateral lipomas have also 
been reported [6]. Our case appears to be one of those.

Lipomas arising from the diaphragm are much more rare 
than other purely intrathoracic lipomas. They have been 
classified [7] according to their anatomic site in: (1) ‘intra-
trathoracic lipomas’ (lying entirely within the thoracic 
cage) and (2) ‘hourglass thoracic lipomas’ (having intra-
and extrathoracic portions, further classified into (a) cervi-
comediastinal and (b) transmural lipomas).

DPLs grow slowly [10] and our patient’s lipoma probably 
existed for a long time. However diagnosis was missed 
twice, when CXRs were carried out as part of routine pre-
operative investigations 2 and 7 years earlier. We found no 
records of recurrence of DPL after resection in the literature.

Malignant transformation of lipomas is rare [11].

Radiologically PDLs appear as smooth rounded masses 
in continuity with the diaphragm, usually arising postero-
laterally [1]. They are homogenous, faintly-shadowed with 
clear borders and less dense than expected on the basis of
their size [1]. CXR findings alone are not specific, hardly distinguishing lipomas from cysts, hernias and other diaphragmatic tumours.

CT scanning has improved the accuracy of qualitative diagnosis of PDL by providing a low attenuation coefficient –80 to –130 HU (that of fatty tissue) [5]. However visualisation of the diaphragm cannot be definitely achieved even by means of CT scanning, because of its thinness, domed contour and contiguity with abdominal soft tissues. Distinguishing between a PDL and herniated omental fat is not easy [8].

The differentiation from Bochdalek’s hernia depends on two elements. Firstly, a DPL is located posterolaterally (rather than posteromedially). Secondly, the integrity of the diaphragm is demonstrated in PDLs, compared to a V-shaped discontinuity of the diaphragmatic musculature in Bochdalek’s hernias [5].

Differentiation between PDLs and malignant tumours (such as liposarcomas) often relies on the assumption that the latter should cause symptoms and would often be associated with a pleural effusion [1]; however such criteria are not safe and we believe that only the histological examination of a completely resected specimen can securely make the final diagnosis. Therefore, we agree [1,3] that surgical resection is indicated, not adopting the also advocated [9,10] conservative management by close observation alone. Additionally, PDLs can gradually become larger, so that they can be expected to cause symptoms after all. Finally, the larger they are the more laborious their resection is.

References