Eur Radiol (2009) 19: 2315–2319
DOI 10.1007/s00330-008-1221-5

INTERPRETATION CORNER

Luciano Cardinale
Francesco Ardissone
Ubaldo Familiari
Mariaelena Perna
Cesare Fava

A case of intermittent lumbar pain radiating to the right shoulder in a 76-year-old woman (2009: 6b)

Received: 22 July 2008
Revised: 5 September 2008
Accepted: 12 October 2008
Published online: 22 July 2009

L. Cardinale · M. Perna · C. Fava
Department of Radiology, S. Luigi Hospital, University of Turin, Turin, Italy

F. Ardissone
Department of Thoracic Surgery, S. Luigi Hospital, University of Turin, Turin, Italy

U. Familiari
Department of Pathologic Anatomy, S. Luigi Hospital, University of Turin, Turin, Italy

L. Cardinale (✉)
Ospedale San Luigi - Radiologia, Regione Gonzole 10, Orbassano, 10043, Italy
e-mail: luciano.cardinale@gmail.com
Tel.: +39-0119026881
Fax: +39-0119026884

Abstract Malignant fibrous histiocytoma (MFH) is a pleomorphic sarcoma, occurring most frequently in the deep soft tissues of the extremities, and it is most frequently seen in elderly patients. A primary MFH of the diaphragm is very rare, and to the best of our knowledge, a multi-phased spiral CT appearance of this tumour has not been previously reported. In this report, we describe the clinical and multi-phase CT features of a primary MFH of the diaphragm.

Keywords Diaphragmatic tumours · Multi-detector CT · Malignant Fibrous · Histiocytoma

Introduction

Malignant fibrous histiocytoma (MFH), described by O'Brien and Stout in 1964, is a pleomorphic sarcoma, occurring most frequently in the deep soft tissues of the extremities. It is most frequently seen in elderly patients [1].

A primary MFH of the diaphragm is very rare [2], and to the best of our knowledge, a multi-phased spiral CT appearance of this tumour has not been previously reported.

In this report, we describe the clinical and CT features of a primary MFH of the diaphragm.

Case report

A 76-year-old, non-smoking woman presented with a 3-month history of intermittent right lumbar pain radiating to the right shoulder, increasing on deep inspiration. The patient's past medical history was characterised by mild hypertension and gastroesophageal reflux. Physical examination and routine laboratory investigations were unremarkable.

Computed tomography of the chest showed a large, lobulated, well-defined extra-pulmonary soft-tissue mass (about 8 cm in diameter) located in the base of the postero-lateral part of the right chest wall. The mass had an aggressive appearance, being confluent with the diaphragm and compressing the liver. It invaded ribs and muscles of the chest wall and caused thickening of the latero-conal fascia. On unenhanced multidetector CT, it appeared hypodense compared with the chest wall muscles (Fig. 1). After injection of iodinated contrast agent, in the arterial phase, it showed intense early nodular contrast enhancement, especially just next to peripheral nodules of the lesion (Fig. 2). During the 4-min-late phase, the lesion showed a more homogeneous and slightly increased contrast enhancement than in the arterial phase (Fig. 3).

A fine-needle aspiration biopsy was performed, and cytological results confirmed a malignant, low-grade soft tissue tumour.

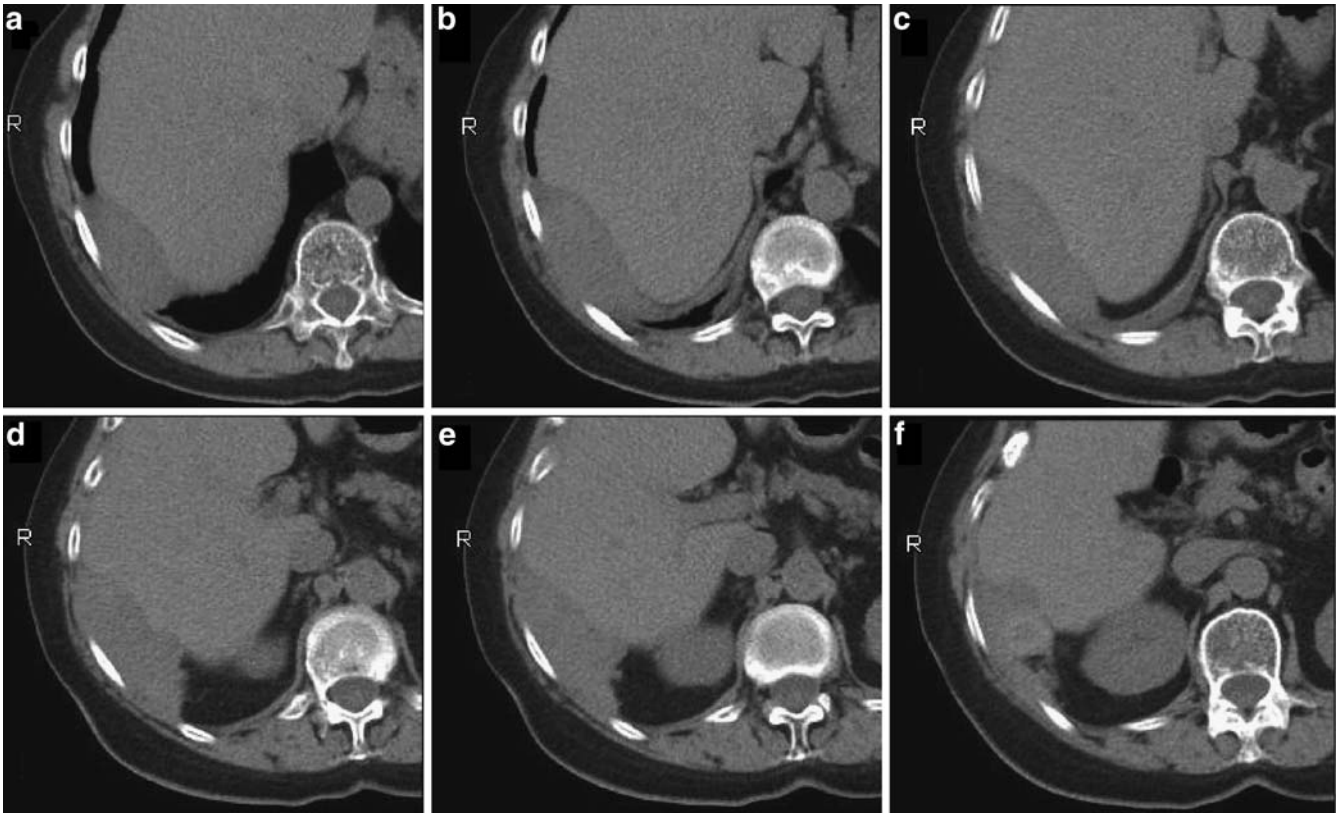

Fig. 1 From A (cranial) to F (caudal). Large, lobulated, well-defined extra-pulmonary soft-tissue mass located in base of the postero-lateral part of right chest wall, indissociable from the diaphragm and compressing the liver. On basal CT the tumour appeared slithly hypodense to the chest wall muscles

Removal of the tumour was performed through a standard right posterolateral thoracotomy, through the fifth intercostal space. During the operation, the tumour presented as a fibrous lobulated mass, which originated in the right hemidiaphragm, involved the adjacent chest wall, and compressed the liver parenchyma, adhering to Glisson's capsule. Through a second incision in the eighth intercostal space, the patient underwent a wide (4-cm margin) en-bloc radical resection of the right posterolateral chest wall, including the 9th to 11th ribs, extended resection of the right hemidiaphragm, and adherent Glisson's capsule. Microscopic confirmation of negative margins was achieved by frozen section. Reconstruction of the chest wall and diaphragm defects was performed with two polytetrafluoroethylene patches. Recovery from the operation was uneventful.

The results of histological and immunohistochemical studies confirmed a definitive diagnosis of pleomorphic MFH.

Eight months after surgery, the patient developed a devastating thromboembolic stroke and died shortly thereafter. At autopsy, the patient was found to have local recurrence appearing as multiple nodules (less than 1 cm in diameter) on the surface of the residual pleomorphic MFH of the right hemi-diaphragm, bordering the lung.

Discussion

Diaphragmatic masses present a radiological challenge because of their rarity and the difficulty of establishing exact anatomical relationship with respect to neighbouring structures, such as the pleura, lungs, spleen or liver where disease is much more frequent.

Grancher was the first to describe a primary tumour of the diaphragm, a fibroma, in 1868 [3]. Weiner and Chou reviewed all reported cases of primary diaphragmatic tumours from 1868–1963 [4], Olaffson from 1963–1968 [5] and Wekslerfrom from 1968 to 1998 [6].

Diaphragmatic tumours may be benign or malignant. Diaphragmatic mesothelial cysts are congenital lesions that derive from coelomic remnant (the coelom is the primitive cavity that lies within the developing embryo that will form

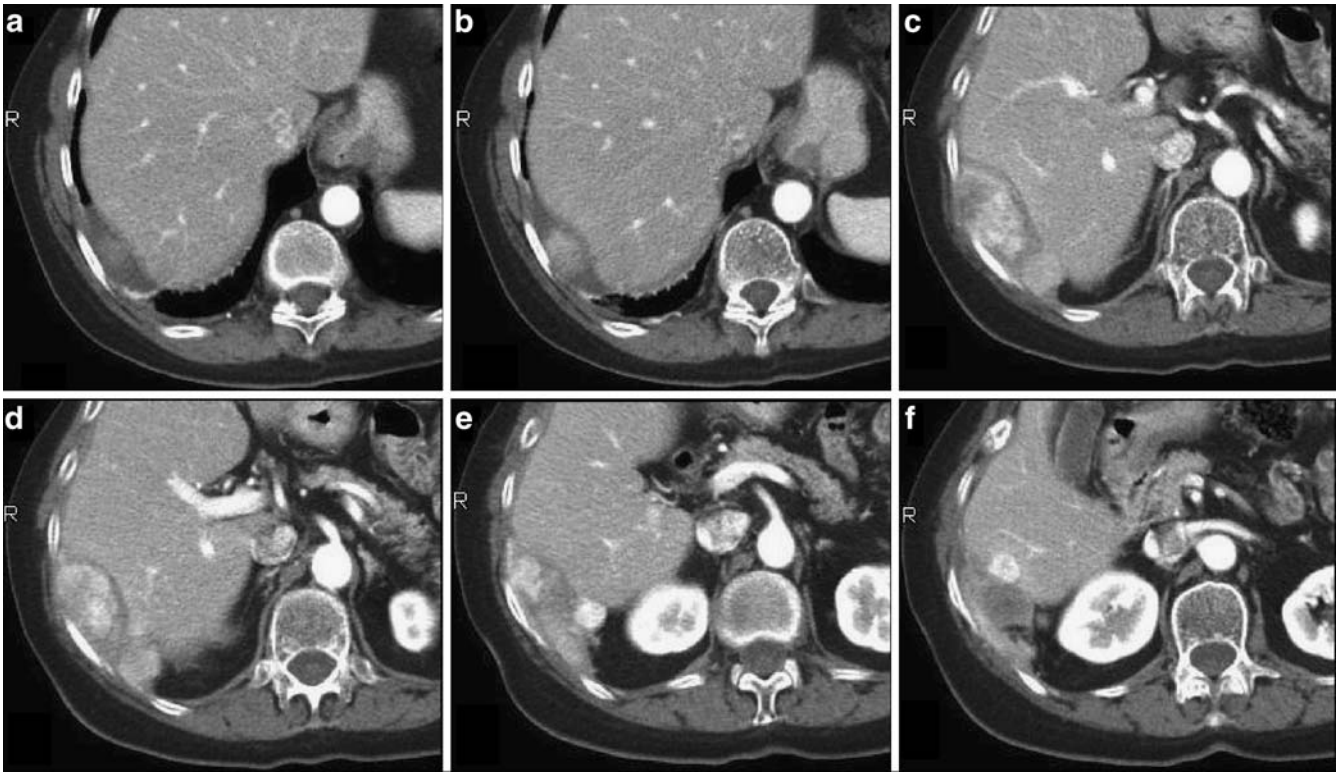

Fig. 2 From A (cranial) to F (caudal). After injection of iodinated contrast agent, in the arterial phase, contrast enhancement is seen within peripheral lesion nodules

the three major body cavities: pericardial, pleural and peritoneal). They represent the most common benign tumours, followed by lipomas and neurolemnomas [7].

As far as malignant tumours are concerned, fibrosarcomas are the most common, followed by leiomyosarcomas and rhabdomyosarcomas.

In the literature so far, there are reports of only five cases of primary malignant fibrous histiocytoma (MFH) originating in the diaphragm [2, 8]; they all presented as intrathoracic masses.

MFH is a deep-seated pleomorphic sarcoma of adults that occurs most frequently in the deep fascia and skeletal muscles of the extremities and the trunk. But there are reports of MFH occurring also in the head and neck region, brain, chest wall, lung, heart, intestinal tract and mesentery [9].

It was first documented in 1964 by O'Brien and Stout [10], who considered it to have a histiocytic origin. There has been a lot of controversy over the histogenesis for several decades, but it still remains uncertain [9]. One of the theories advanced is that it might originate from primitive mesenchymal cells, which have the capacity for multidirectional differentiation. This could also explain why MFH can occur at any location and has a diversified histological morphology [11, 12].

MFH is aggressive with a propensity for local recurrence and distant metastasis [13, 14]. Tumour size is a factor that seems to influence the rate of local recurrence or metastasis, the prognosis of MFH being worse in patients with tumours larger than 5 cm in diameter [2, 14, 15].

The majority (60%) of MFHs are classified as the storiform-pleomorphic subtype; they are pleomorphic sarcomas that contain both fibroblast-like and histiocytic-like elements in varying proportions, thereby manifesting a spectrum of histological appearances [13, 16].

Characteristically, the plump spindle cells are arranged in a cartwheel-like appearance around vessels forming peripheral nodules that are typical of the pleomorphic form [1]. On a pretherapeutic CT study calcifications are uncommon prior to chemotherapy.

After administration of contrast material, there is often nodular and peripheral arterial enhancement of the solid (pleomorphic) elements [1, 18] due to an abundant capillary network. Our case also, in late venous phase (4

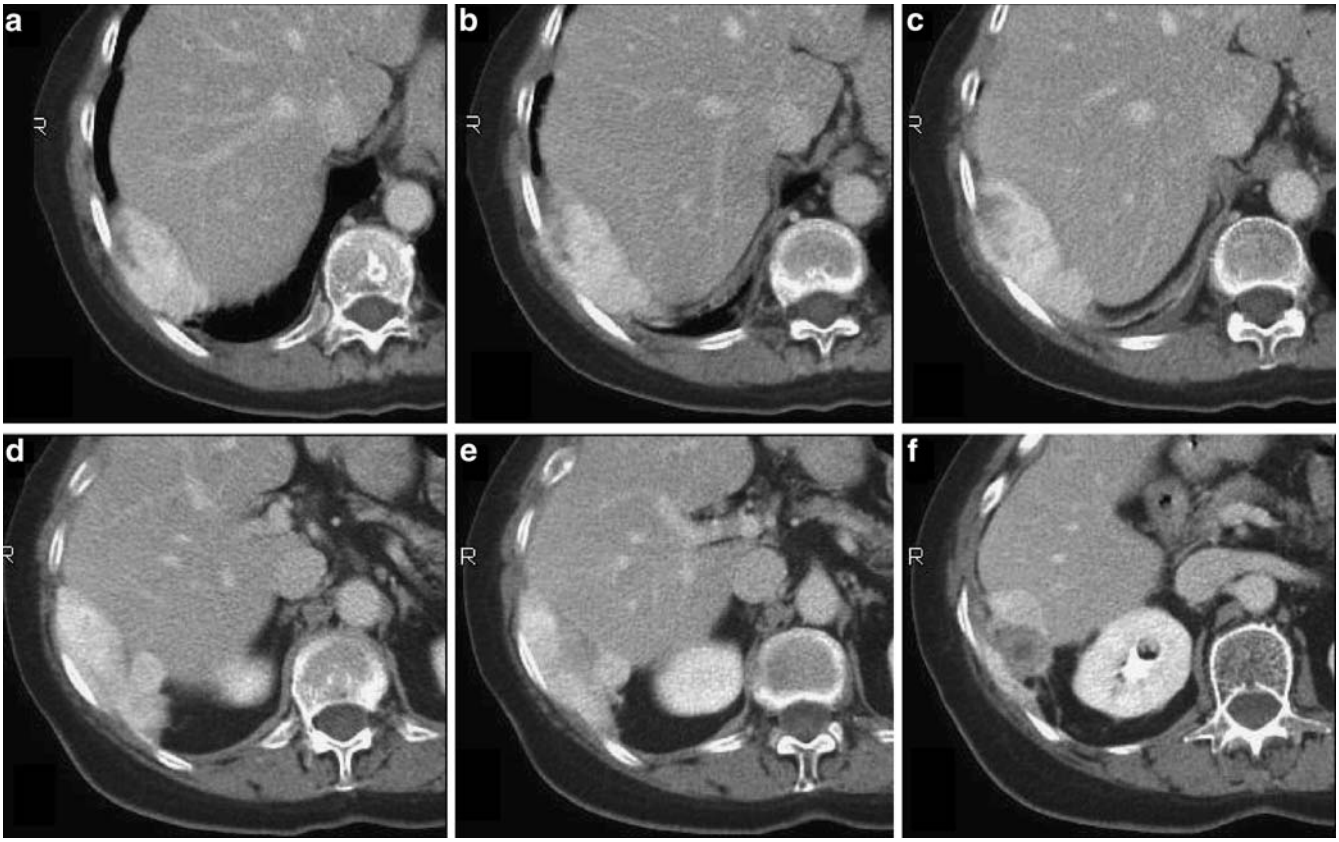

Fig. 3 From A (cranial) to F (caudal). During the 4-min-late phase, the lesion showed a more homogeneous contrast enhancement than in the arterial phase

min after contrast agent injection), shows persistence of slight and homogeneous contrast enhancement.

The second most common subtype (25%) is the myxoid variant. It is characterised by a prominent myxoid appearance to the stroma. Myxoid tissue is generally hypocellular. Typically, on CT scans it presents a low density central area because of a large amount of mucous material, such as haemorrhage or necrosis [1, 17, 18].

Occasionally the appearance of myxoid MFHs, particularly those with more vascular and cellular elements, is less specific on CT images, with a more homogeneous soft-tissue appearance being seen both before and after contrast material administration.

Diaphragmatic MFHs, which occur in older adults, have to be distinguished from the other most common malignant diaphragmatic tumours, such as fibrosarcoma, which is isodense to the muscles in plain CT.

Leiomyosarcoma has a spindle shape and after intravenous contrast material typically exhibits an enhanced peripheral rim and a quite large central area of low attenuation. Differential diagnosis with pleomorphic MFH is quite easy due to the absence of peripheral enhanced nodules in leiomyosarcoma. Otherwise, it is similar to myxoid MFH because of central large hypocellular areas, but leiomyosarcoma typically occurs in young people.

Rhabdomyosarcoma occurs usually in people younger than 45 years of age and has areas of necrosis that do not enhance after contrast injection and that can alternate with areas of marked enhancement [17].

A diaphragmatic lesion has to be distinguished from a localized pleural malignant mesothelioma, which has calcifications and shows a slight contrast enhancement.

Finally, the appearance of diaphragmetic liposarcoma varies from a predominantly fat-containing mass to a solid mass: low attenuation values around −50 Hounsfield units are consistent with a tissue composed of fat; greater values are related to the necrosis, heterogeneity and soft tissue component in liposarcomas [19].

As far as we can ascertain, this is the first report of a dynamic multi-phase multidetector CT study of a primary diaphragmatic MFH.

Although the definitive diagnosis of MFH is histological, multi-phase CT is useful for diagnosing the presence of a tumour and its characterisation in the diaphragm because detection of peripheral nodules with intense contrast enhancement in arterial phase could be useful for diagnosing MFH.

References

1. Grancher M (1868) Tumeur vegetante du centre phrenique du diaphragme. Bull Soc Anat Paris 4385–4386
2. Weiner MF, Chou WH (1965) Primary tumours of the diaphragm. Arch Surg 90:143–152
3. Olafson G, Rausing A, Holen O (1971) Primary tumours of the diaphragm. Chest 59:568–570
4. Weksler B, Ginsberg RJ (1998) Tumours of the diaphragm. Chest Surg Clin N Am 8:441–447
5. Yamamoto H, Watanabe K, Takayama W, Yamada S, Honda I, Fujita Y, Obata S, Komatsu T (1994) Primary malignant fibrous histiocytoma of the diaphragm: report of a case. Jpn J Surg 24:744–748
6. Puls R, Kreissig R, Hosten N, Gaffke G, Stroszcynski C, Felix R (2002) Tumour of the diaphragm mimicking liver lesion. Eur J Radiol 41:168–169
7. Estaùn JE, Alfageme AG, Banuelos JS (2003) Radiological appearance of diaphragmatic mesotelial cysts. Pediatr Radiol 33:855–858
8. Huang CC, Ko SF, Ng SH et al (2001) Cystic malignant fibrous of the gastrocolic ligament. Br J Radiol 74:651–653
9. O'Brien JE, Stout AP (1964) Malignant fibrous xanthomas. Cancer 17:1445–1455
10. Kim MH, Chung J-J, Choi YJ, Park KS, Park S, Yang HC (2006) Malignant fibrous histiocytoma of the common hepatic duct: a case report. J Comput Assist Tomogr 30(6):903–905
11. Kempson RL, Kiriakos M (1972) Fibroxanthosarcoma of the soft tissues: a type of malignant fibrous histiocytoma. Cancer 29:961–976
12. Weiss SW, Enzinger FM (1978) Malignant fibrous histiocytoma. An analysis of 200 cases. Cancer 41:2250–2266
13. Pezzi CM, Rawling MS, Esgro JJ, Pollock RE, Romsdahl MM (1992) Prognostic factors in 227 patients with malignant fibrous histiocytoma. Cancer 69:2098–2103
14. Bertoni F, Capanna R, Biagini R, Bacchini P, Guerra A, Ruggieri P, Present D, Campanacci M (1985) Malignant fibrous histiocytoma of soft tissue. An analysis of 78 cases located and deeply seated in the extremities. Cancer 56:356–367
15. Enzinger FM, Weiss SW (1988) Malignant fibrohistiocytic tumours. In: Enzinger FM, Weiss SW (eds) Soft tissue tumours, 2nd edn. Mosby, St Louis, Mo, pp 269–300
16. Murphey MD, Gross TM, Rosenthal HG (1994) Musculoskeletal malignant fibrous histiocytoma: radiologic-pathologic correlation. From the archives of the AFIP. Radiographics 14:807–826
17. Tateishi U, Kusumoto M, Hasegawa T, Yokoyama R, Moriyama N (2002) Primary malignant fibrous histiocytoma of the chest wall: CT and MR appearance. J Comput Assist Tomogr 26 (4):558–563
18. Tateishi U, Gladish G, Kusumoto M, Hasegawa T, Yokoyama R, Tsuchiya R, Moriyama N (2003) Chest wall tumours: radiologic findings and pathologic correlation: part 2. Malignant tumours. Radiographics 23(6):1411–1508
19. Evans HL (1988) Liposarcomas and typical lipomatous tumors: a study of 66 cases followed for a minimum for 10 years. Surg Pathol 1:41–54

Precisely correct answer was received by closing date from:

Marc De Baets, Lugano, Switzerland