

Thoracic myxoid lipoblastoma (ECR 2018 Case of the Day)

Published on 06.07.2018

DOI: 10.1594/EURORAD/CASE.15917

ISSN: 1563-4086

Section: Paediatric radiology

Area of Interest: Thoracic wall Soft tissues / Skin

Procedure: Radiation safety

Procedure: Diagnostic procedure

Imaging Technique: CT

Imaging Technique: MR

Imaging Technique: MR-Diffusion/Perfusion

Special Focus: Neoplasia Case Type: Clinical Cases

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Patient: 11 years, female

Clinical History:

An 11-year-old male patient with a two-week history of chest pain.

Imaging Findings:

Chest radiograph shows a round-shaped lesion in the upper right hemithorax with “incomplete border” sign, suggesting an extrapulmonary origin.

Contrast-enhanced thorax CT scan in axial and coronal plane with lung and soft tissue window confirms the presence of a homogeneous, slightly hypodense (16 UH) extrapulmonary mass with obtuse angles. In bone window the adjacent rib shows a very subtle medial margin spiculation without osteolysis. Lungs and mediastinum were normal.

Chest MR showed a high T2 and T2 FS signal extrapulmonary chest soft tissue mass. Extent from the 5th to the 7th rib, with mass effect and increased intercostal space and posterior growth, evident on the sagittal T2 WI. The adjacent ribs show normal signal intensity.

On T1 WI the lesion is slightly hyperintense compared to muscles. Post-contrast shows heterogeneous intense enhancement.

Hyperintensity on DWI was related to T2 effect given the hyperintensity on the ADC map (average $2.4 \times 10^{-3} \text{mm}^2/\text{sec}$).

Discussion:

Background: Lipoblastomas are rare benign mesenchymal tumours of infancy and early childhood that arise from

abnormal proliferation of immature fat cells. [1] Depending on their growth pattern these tumours are considered as lipoblastoma when they are well-circumscribed and lipoblastomatosis when they have an infiltrative growth. [2]

Clinical Perspective: Symptoms related to lipoblastoma vary depending on localisation, in our case, chest pain might be related to compression of adjacent intercostal nerves or pleural involvement. Chest plain film is usually one of the first imaging techniques for evaluation of chest pain in children, and CT and/or MR were indicated to evaluate a thoracic mass.

Imaging Perspective: Chest plain film is highly suggestive of an extrapulmonary lesion. MR appearance of lipoblastoma varies according to histological components. Adipocytes are accompanied by fibrous septa and frequently a plexiform myxoid matrix that comprises large portions of the tumour [1]. This is the case in our patient where no mature fat tissue was evident on MR.

Lesions that are predominantly composed of mature fat appear much like lipomas, hyperintense on T1- and T2-weighted images and hypointense on fat-suppressed images. Lipoblastoma and myxoid liposarcoma cannot be reliably differentiated on imaging alone. Previous studies mentioned that liposarcomas are exceedingly rare in children younger than 10 years of age with the myxoid variant representing the most frequent histologic subtype [3]. However, recent studies established a younger age than 5 years to comfortably rule out liposarcoma. [4] In this case the patient is 11 years old, so liposarcoma should be considered in the differential diagnosis.

Imaging techniques, histopathology and immunohistochemistry remain the gold standard in differentiating a lipoblastoma from a myxoid liposarcoma. [5]

Outcome: Total excision is the treatment of choice with preservation of vital organs. The prognosis is excellent despite large tumour size and local invasion. Recurrence rate is high in incompletely resected tumours. Regular follow-up is important for early detection of recurrences. [1]

Take-Home Message, Teaching Points: Chest plain film suggests an extrapulmonary mass in this case. We review the MR appearance of a rare paediatric adipocytic tumour. Differential diagnosis with liposarcoma should be considered in children over 5 years of age.

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Thoracic myxoid lipoblastoma, Lipoblastoma, Askin tumour, Ganglioneuroma, Neurofibroma, Myofibroma

Final Diagnosis: Thoracic myxoid lipoblastoma

References:

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- Krishnan J, Hathiramani V, Hastak M, Redkar RG. (2013) Myxoid lipoblastoma. *Indian Pediatr* 8;50(6):603-5. (PMID: [23942405](#))

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Figure 1

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Description: Chest PA radiograph **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain

Figure 2

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Description: Thorax axial CT with lung window. **Origin:** Department of Radiology , Hospital Sant Jan de Deu , Barcelona , Spain

Figure 3

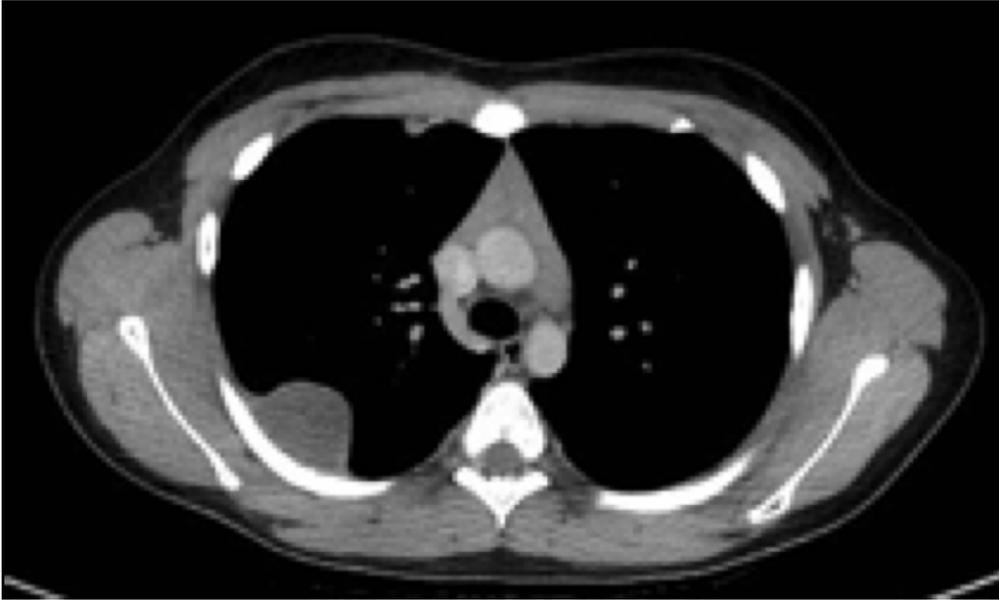
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Description: Thorax axial CT with bone window. **Origin:** Department of Radiology, Hospital Sant Joan de Deu , Barcelona , España.

Figure 4

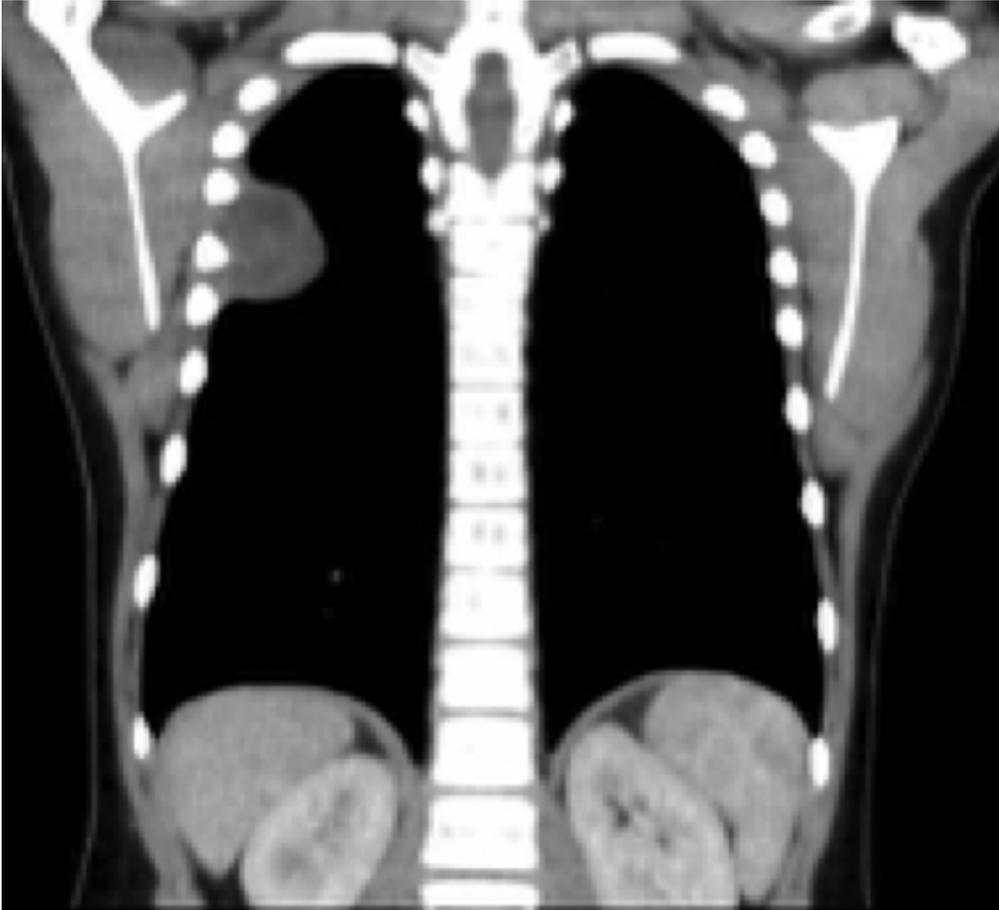
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Description: Thorax axial CT with soft tissue window. **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain.

Figure 5

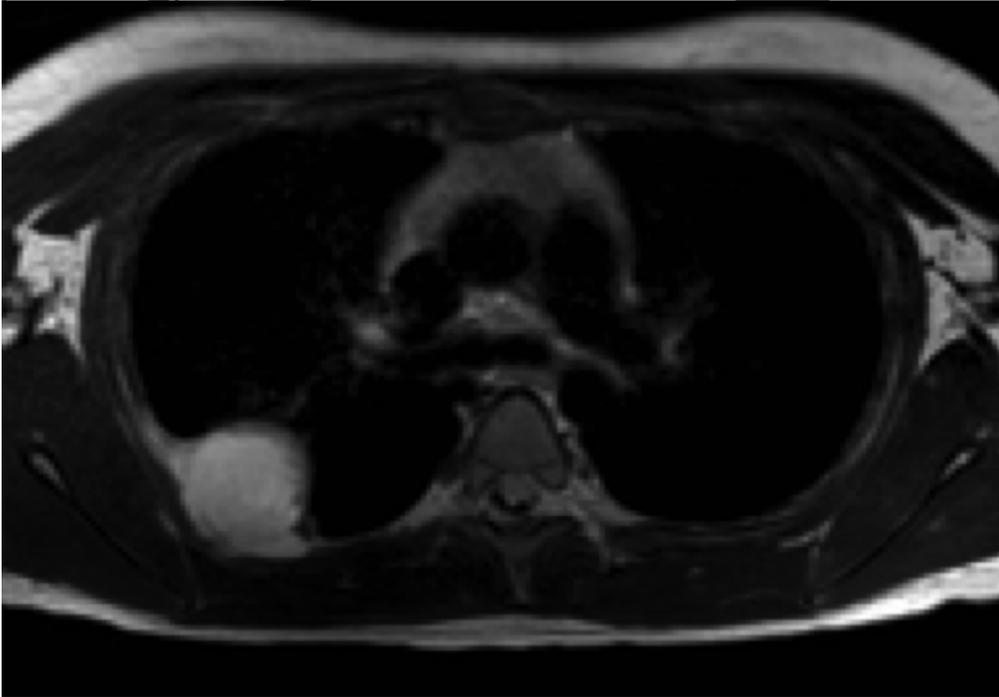
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Description: Thorax CT coronal reconstruction with soft tissue window. **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain.

Figure 6

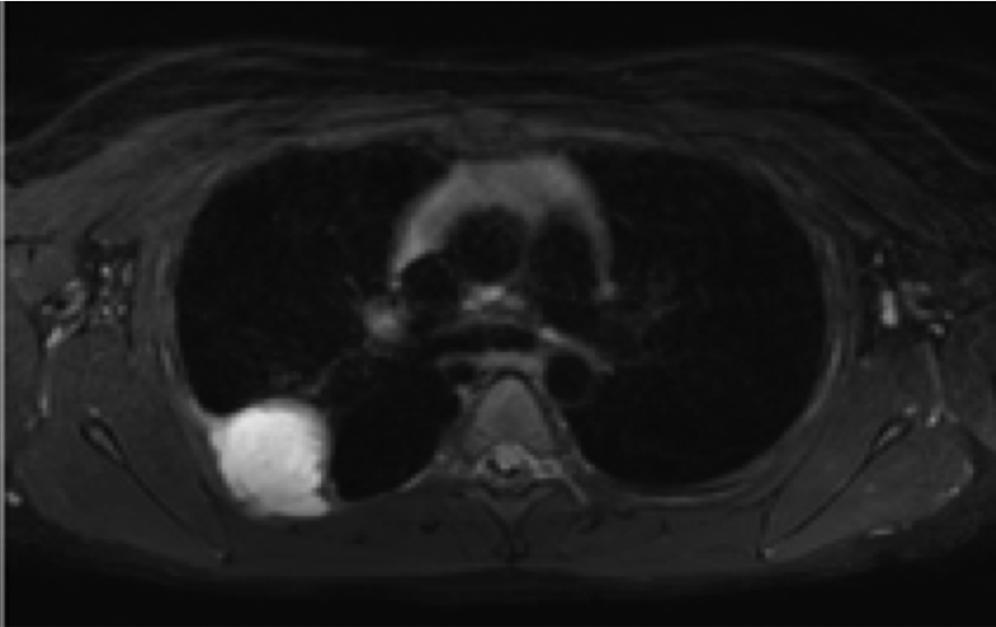
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Description: Thoracic MRI axial DIXON T2 **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain.

Figure 7

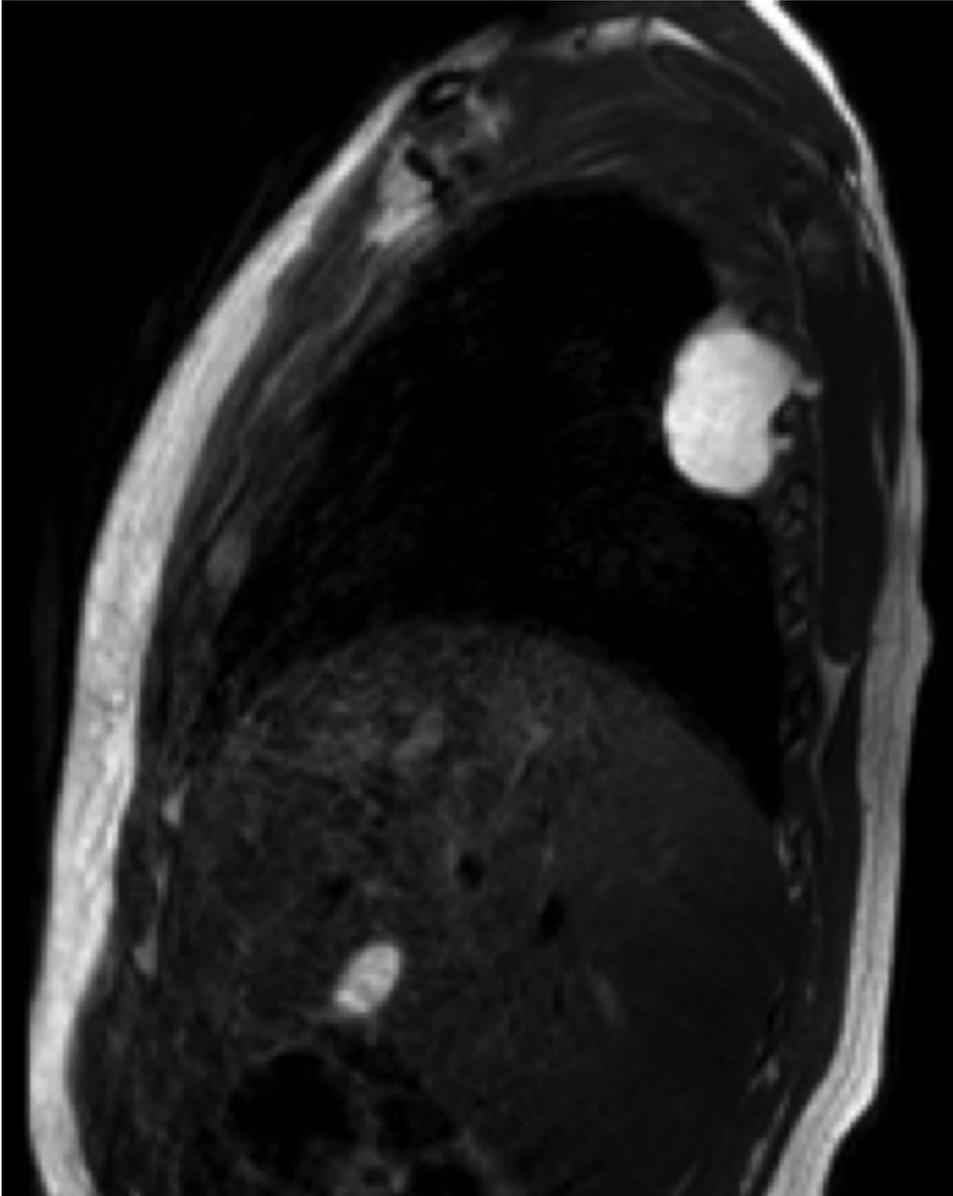
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Description: Thoracic MRI axial DIXON FS T2 **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain.

Figure 8

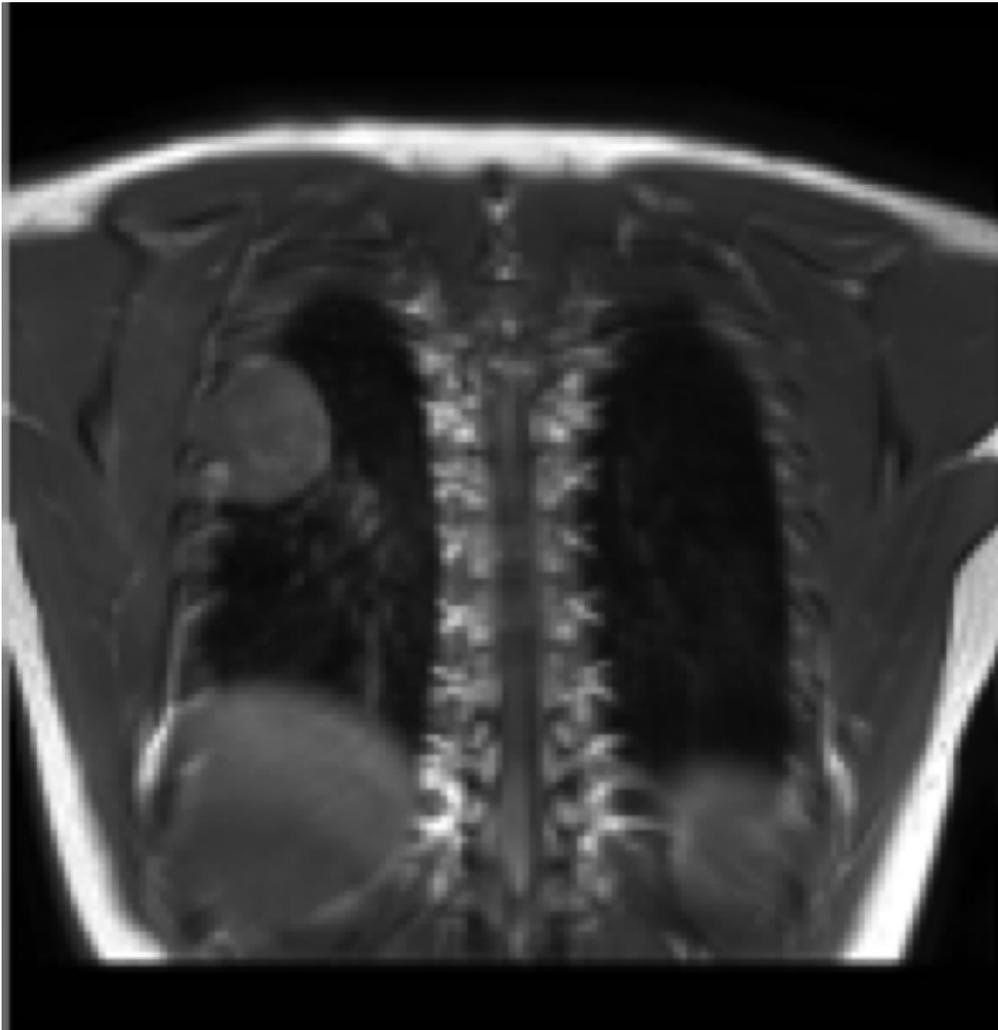
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Description: Thoracic MRI sagittal T2 **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain.

Figure 9

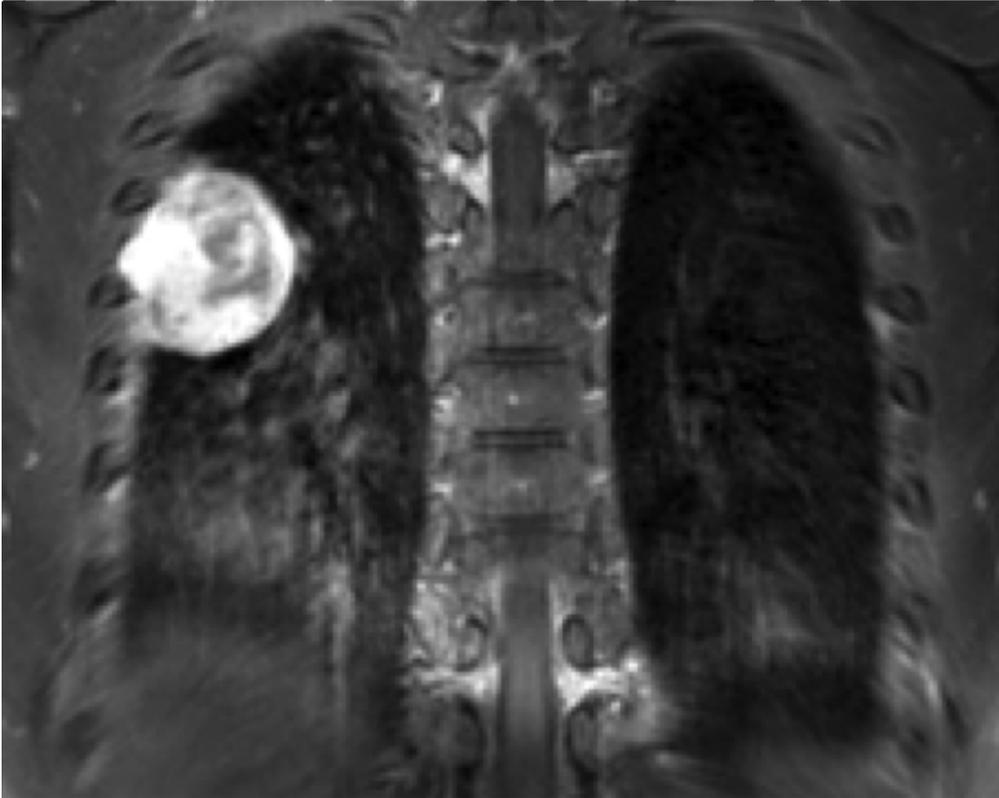
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Description: Thoracic coronal MRI T1 SPAIR **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain.

Figure 10

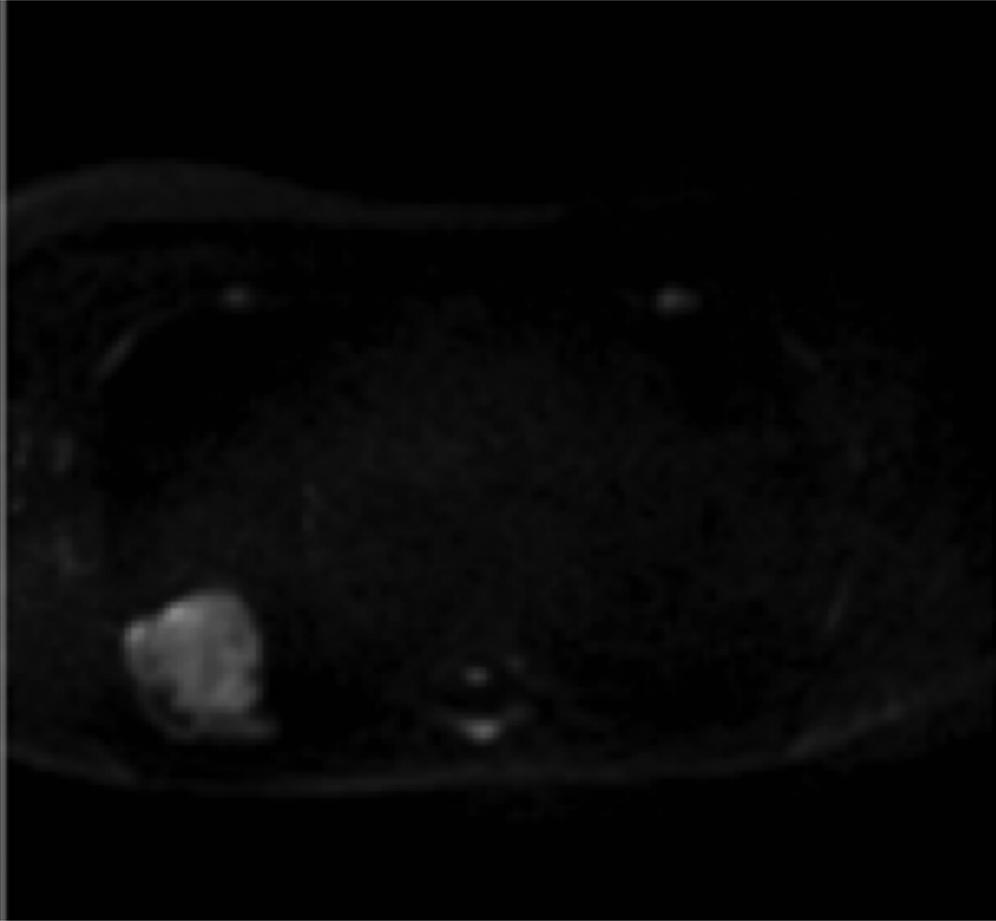
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Description: Thoracic coronal MRI T1 SPAIR with gadolinium **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain.

Figure 11

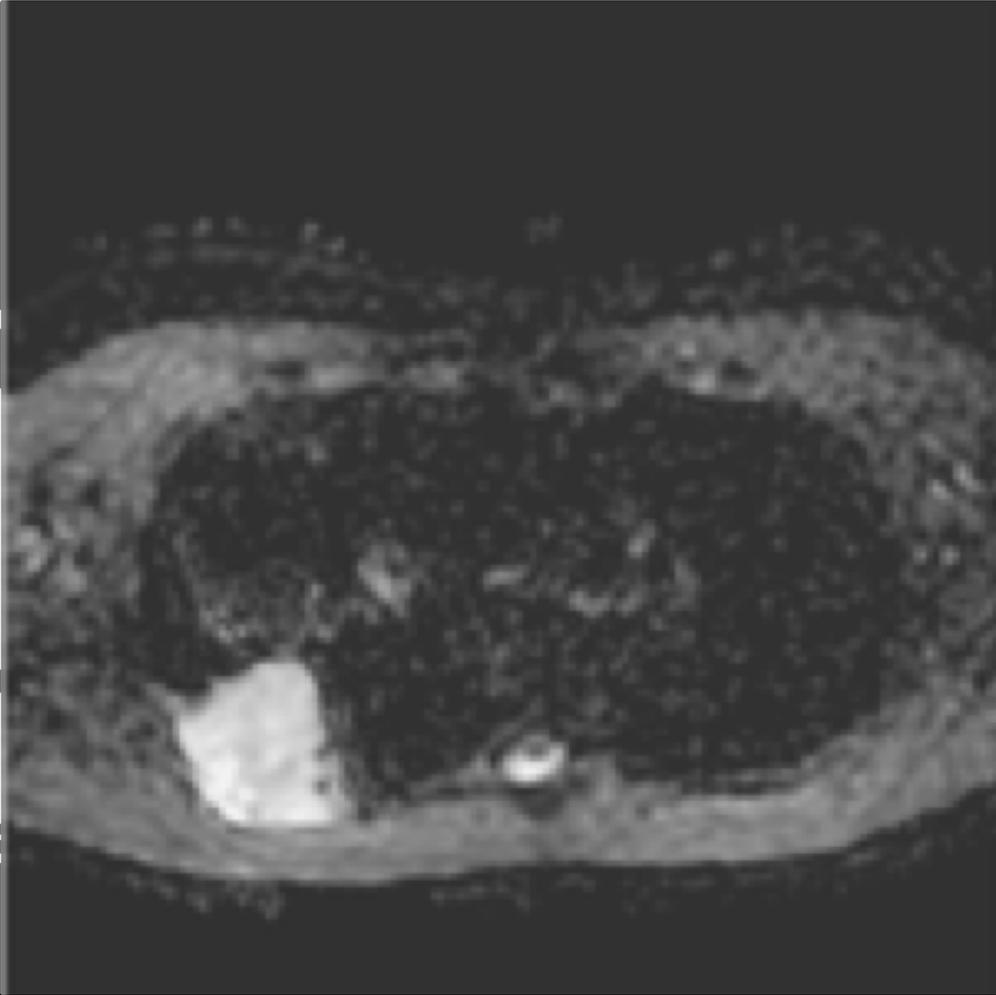
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Description: Thoracic MRI DWI **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain.

Figure 12

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Description: Thoracic MRI ADC map. **Origin:** Department of Radiology , Hospital Sant Joan de Deu , Barcelona , Spain.