

Imaging in extramedullary haematopoiesis (ECR 2011 Case of the Day)

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Section: Chest imaging

Area of Interest: Education

Procedure: Diagnostic procedure

Procedure: Education

Imaging Technique: Conventional radiography

Imaging Technique: CT

Imaging Technique: MR

Special Focus: Haematologic diseases Case Type:
Clinical Cases

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Patient: 31 years, male

Clinical History:

A 31-year-old man with a known history of β -thalassemia major, thoracic diffuse pain, dyspnoea, and migraine, was referred to our department for a standard chest evaluation followed by a thoracic and brain CT. After 1 year, a MRI cervico-thoracic spine evaluation was for paresthesia and medullar compression with a thoracic T3-T4 level.

Imaging Findings:

Poster-anterior chest radiography (Fig. 1) and lateral chest radiography (Fig. 2) followed by a thoracic CT (Fig. 3-4) and a brain CT (Fig. 5-6) were performed. After 1 year magnetic resonance imaging evaluation of the cervical and the upper part of thoracic spine in sagittal and axial plane T1 weighted was performed (Fig. 7-8). Findings on chest radiographies consists in bilateral, smooth, sharply delineated, lobulated paraspinal masses without erosion of the vertebral bodies or ribs (Fig. 1, 2). Computed tomography of the thorax and head revealed marked medullary expansion of the bony structures with the ribs showing the most pronounced involvement (Fig. 3, 4), but also skull base thickening (Fig. 5, 6). MRI of the spine revealed paravertebral and epidural mass with mass effect and compression of the thoracic spinal cord (Fig. 7, 8).

Discussion:

Extramedullary haematopoiesis (EMH) occurs as a compensatory mechanism for abnormal haematopoiesis, due to the proliferation of haematopoietic cells outside of the bone marrow in response to production of too few blood cells to satisfy the body's demand [1, 2]. EMH occurs in haemoglobinopathies, myeloproliferative disorders, or bone marrow infiltration. Myeloproliferative disorders include chronic myelogenous leukaemia, polycythemia vera, essential thrombocytosis, and myelofibrosis with myeloid metaplasia. Haemoglobinopathies include sickle cell disease and thalassaemia [3].

EMH mainly involves the RE system (liver, spleen, and lymph nodes) but is also known to occur in every organ of the body, including the thyroid, prostate, pericardium, kidney, and lungs [1]. Rarely, it can cause cord compression, pleural effusion, massive haemothorax, and respiratory failure [3-6]. EMH should be considered in the differential diagnosis in a patient with chronic severe anaemia and a paraspinal mass [6, 7].

Chest radiography reveals a sharply outlined, lobulated paraspinal and rib masses (Fig. 1-2). The thoracic CT

examination revealed well-defined bilateral paraspinal and rib masses with intralesional fine linear calcifications (Fig. 3-4). The posterior parts of the ribs showed increased trabeculation and dimensions (Fig. 4). CT of the brain showed a sphenoid osteolytic mass (Fig. 5-6). Conventional spin echo T1 MRI demonstrated that the right paravertebral masses and the mass of the epidural posterior space have the same signal characteristics with the marrow of the adjacent vertebral body (Fig. 7-8). The epidural thoracic mass compress the adjacent thoracic spinal cord (Fig. 8). Patients with haemoglobinopathies are more likely to have EMH in paraosseous locations, whereas patients with myeloproliferative disorders are more likely to have extraosseous masses [11]. More commonly observed areas of EMH include the paraspinal regions of the thorax, liver, and spleen, but it has been reported in other locations, including the adrenal gland, bowel, dura mater, and breast [11-14]. Only a few cases of perirenal and pelvicaliceal EMH have been reported in the literature [11].

Intrathoracic EMH is a rare cause of paraspinal mass and should be differentiated from other costovertebral angle masses, such as neurogenic tumours, lymphoma, metastasis, paravertebral abscess [1, 8].

EMH usually regresses or disappears after treatment with blood transfusion and hydroxyurea, splenectomy in cases of spherocytosis or radiation therapy [1, 9, 10]. The paraspinal thoracic regions, the spine, and the skull base are common sites of involvements by EMH, and familiarity with its radio-imaging findings and knowledge of the patient's clinical history are essential to avoid misdiagnosis.

Differential Diagnosis List: Extramedullary haematopoiesis, Myeloma, Lymphoma, Multiple ganglioneuroma, Metastasis, Multiple abscesses

Final Diagnosis: Extramedullary haematopoiesis

References:

- Berkmen YR, Zalta BA (2007) Extramedullary Hematopoiesis. Radiology 245:905-908 (PMID: [18024458](#))
- Georgiades CS, Neyman EG, Francis IR, Sneider MB, Fishman EK (2002) Typical and atypical presentations of extramedullary hemopoiesis. AJR 179:1239–1243 (PMID: [12388506](#))
- Chourmouzi D, Pistevou-Gompaki K, Plataniotis G, Skaragas G, Papadopoulos L, Drevelegas A (2001) MRI findings of extramedullary haemopoiesis. Eur Radiol 11:1803–1806 (PMID: [11511905](#))
- Marchiori E, Escuissato DL, Irion KL, et al (2008) Extramedullary hematopoiesis : findings on computed tomography scans of the chest in 6 patients. J Bras Pneumol 34:812–6 (PMID: [19009214](#))
- Chunduri S, Gaitonde S, Ciurea SO, et al (2008) Pulmonary extramedullary hematopoiesis in patients with myelofibrosis undergoing allogeneic stem cell transplantation. Haematologica 93:1593–5 (PMID: [18641018](#))
- Choi H, David CL, et al (2004) Extramedullary hematopoiesis. Radiology 231:52-56 (PMID: [14990806](#))
- Gumbs RV, Higginbotham-Ford EA, Teal JS, et al (1987) Thoracic extramedullary hematopoiesis in sickle cell disease. AJR 149:889-893 (PMID: [3499791](#))
- Bolaman Z, Polatli M, et al (2002) Intrathoracic extramedullary hematopoiesis resembling posterior mediastinal tumor. Am J Med 112:739–740 (PMID: [12079720](#))
- Weinschenker P, Kutner JM, Salvajoli JV, et al (2002) Whole-pulmonary low-dose radiation therapy in agnogenic myeloid metaplasia with diffuse lung involvement. Am J Hematol 69:277–280 (PMID: [11921022](#))
- Ng CS, Wan S, Lee TW, et al (2002) Video-assisted thoracic surgery for mediastinal extramedullary haematopoiesis. Ann R Coll Surg Engl 84:161–163 (PMID: [12092864](#))
- Dunnick NR (2000) The Radiological Society of North America 85th Scientific Assembly and Annual Meeting: Image interpretation session: 1999. Radiographics 20:257-278 (PMID: [10554618](#))
- King BF, Kopecky KK, Baker MK, Clark SA (1987) Extramedullary hematopoiesis in the adrenal glands: CT characteristics. J Comput Assist Tomogr 11:342-343 (PMID: [3819139](#))
- Palmer GM, Shortsleeve MJ (1998) Gastric polyps due to extramedullary hematopoiesis. AJR 171:531 (PMID: [9694500](#))

Rapezzi D, Racchi O, Ferraris AM (1997) Perirenal extramedullary hematopoiesis in agnogenic myeloid metaplasia: MR imaging findings. AJR 168:1388-1389 (PMID: [9129461](#))

Figure 1

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Description: Bilateral, smooth, sharply delineated, often lobulated paraspinal masses without erosion of the vertebral bodies or ribs. **Origin:** I.Lupescu, Radiology Department of Fundeni Clinical Department, Bucharest, Romania

Figure 2

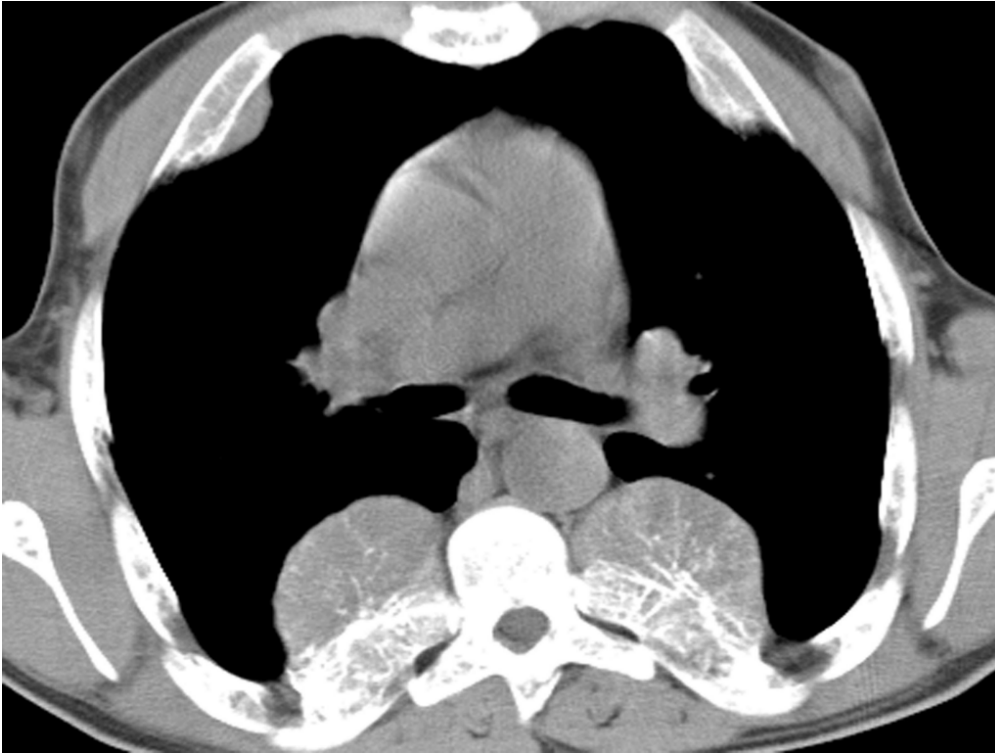
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Description: Lobulated paraspinal masses along the entire length of the vertebral column. **Origin:** I.Lupescu. Radiology Department of Fundeni Clinical Institute, Bucharest, Romania

Figure 3

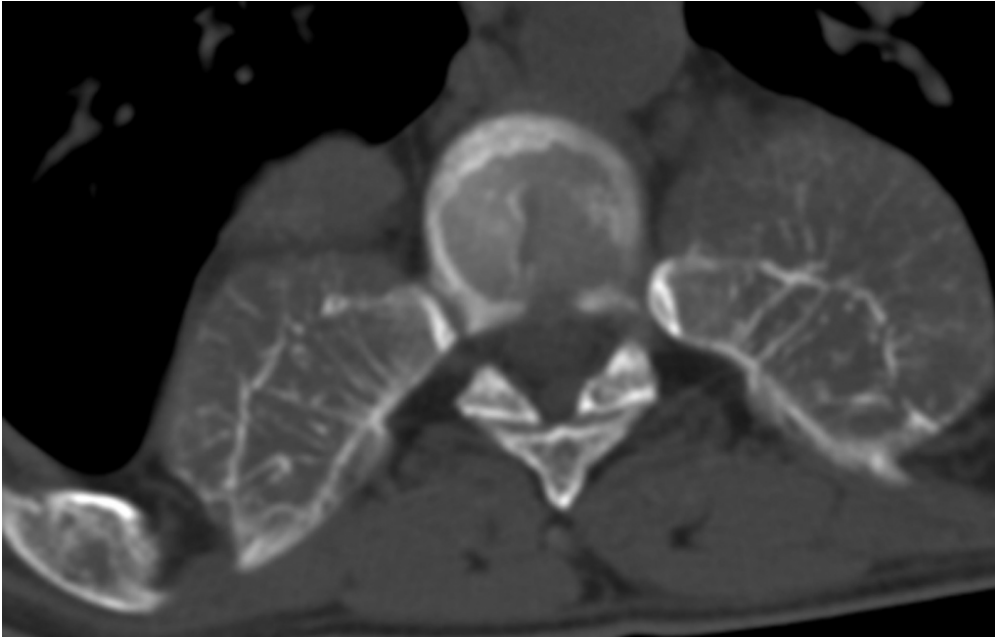
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Description: Marked medullary expansion of the bony structures with the ribs showing the most pronounced involvement. **Origin:** I.Lupescu. Radiology Department of Fundeni Clinical Institute, Bucharest, Romania

Figure 4

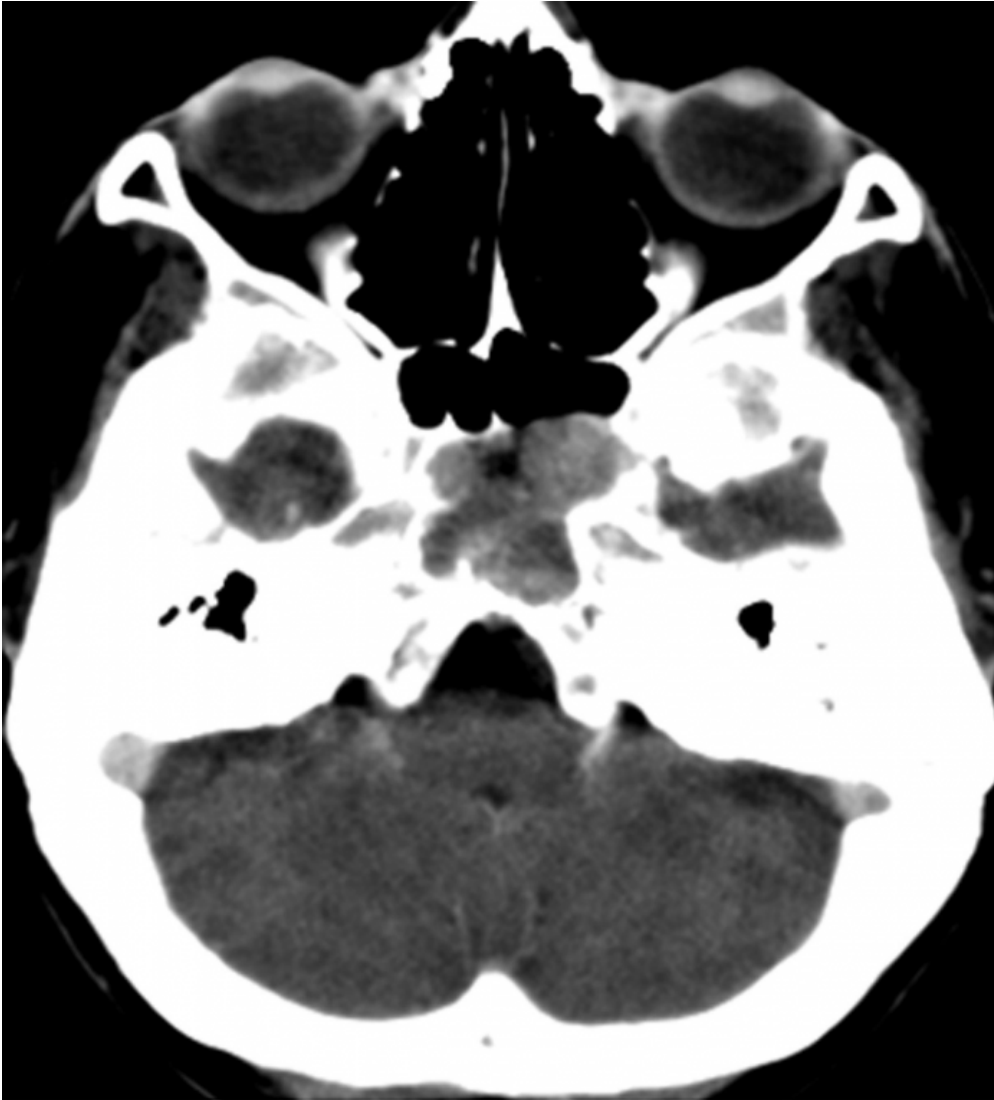
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Description: Marked medullary expansion of the bony structures with the ribs showing the most pronounced involvement. **Origin:** I.Lupescu. Radiology Department of Fundeni Clinical Institute, Bucharest, Romania

Figure 5

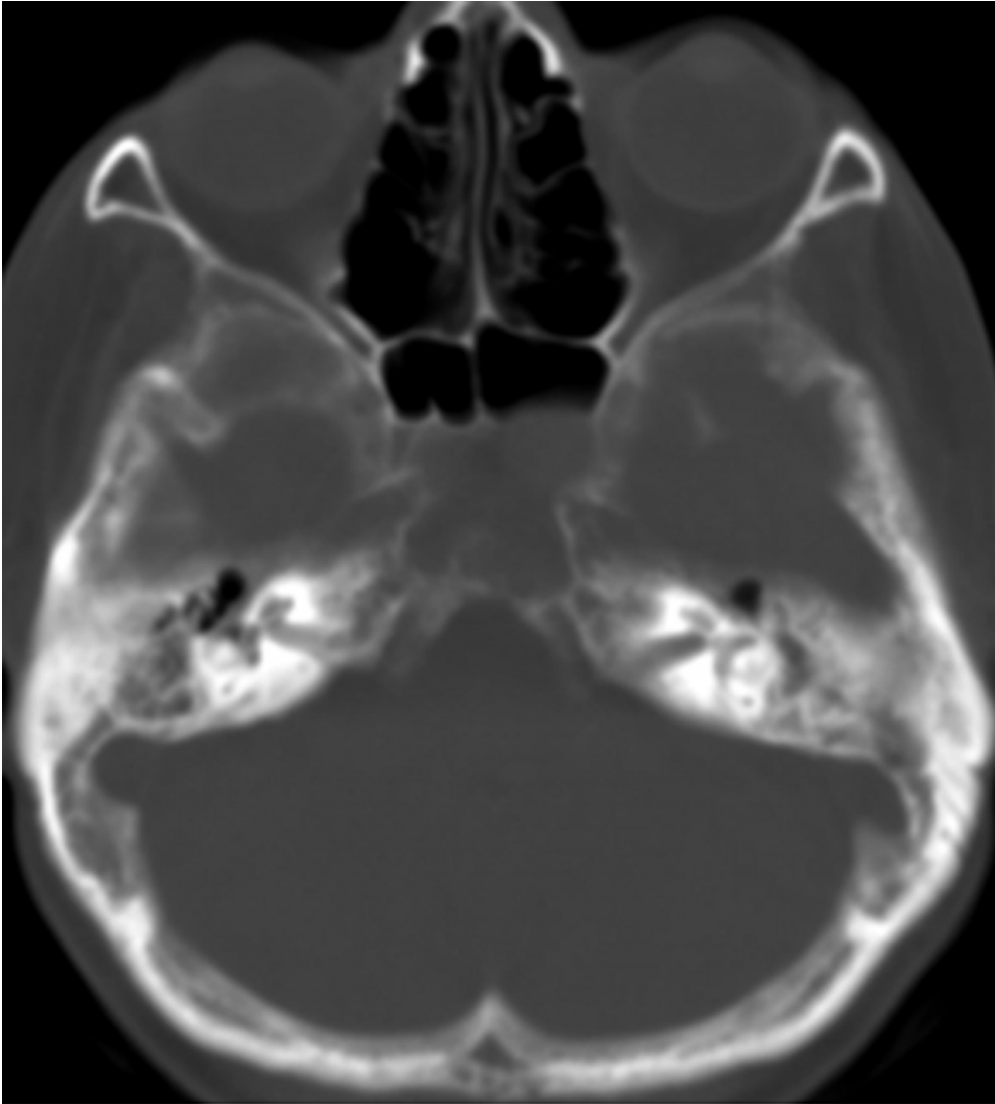
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Description: Skull base thickening and sphenoidal mass. **Origin:** I.Lupescu. Radiology Department of Fundeni Clinical Institute, Bucharest, Romania

Figure 6

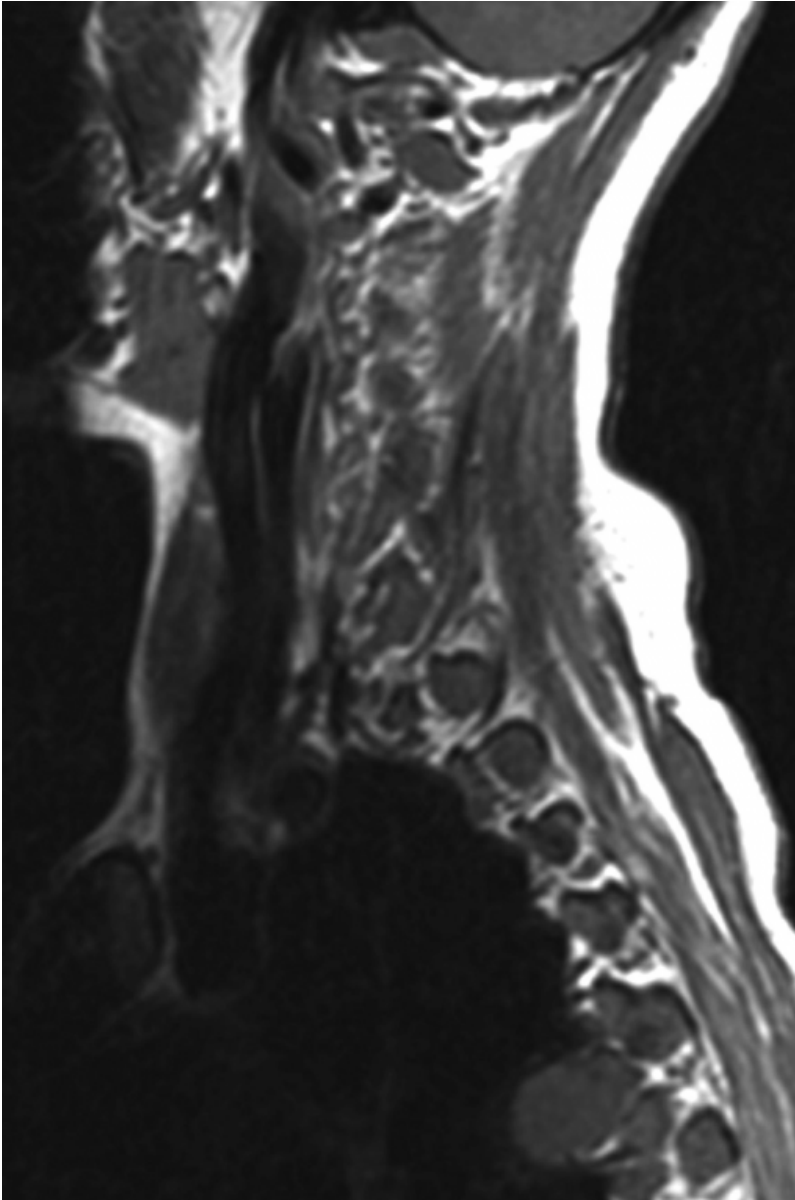
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Description: Skull base thickening and sphenoidal mass. **Origin:** I.Lupescu. Radiology Department of Fundeni Clinical Institute, Bucharest, Romania

Figure 7

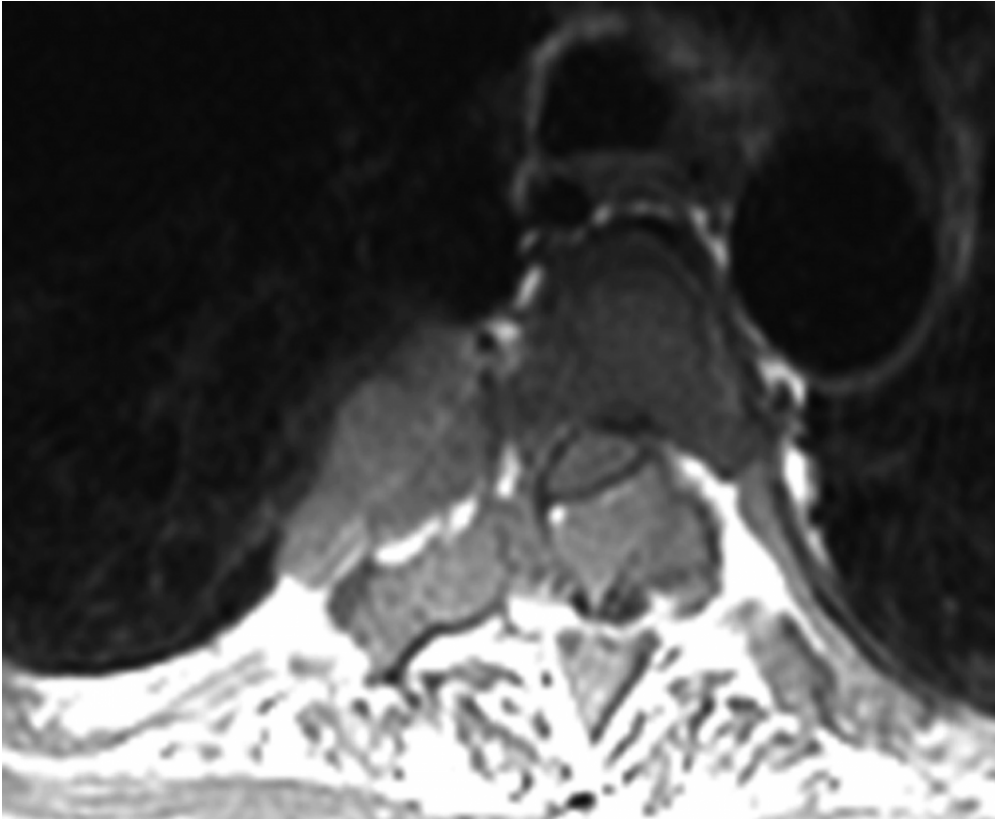
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Description: Paravertebral thoracic mass. **Origin:** I.Lupescu. Radiology Department of Fundeni Clinical Institute, Bucharest, Romania

Figure 8

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Description: Paravertebral and epidural mass with mass effect and compression of the thoracic spinal cord. **Origin:** I.Lupescu. Radiology Department of Fundeni Clinical Institute, Bucharest, Romania