CASE REPORT

Mediastinal myelolipoma with leukocytosis

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SUMMARY

Myelolipoma is a benign tumour consisting of mature fat with scattered foci of haematopoietic elements resembling bone marrow. Extra-adrenal myelolipomas are an infrequent pattern of presentation. We report the case of a 64-year-old woman who presented a heterogeneous 1.4×2.5×3 cm paravertebral thoracic mass detected by chest tomography during the study of a leukocytosis of unknown aetiology. The CT findings and pathology revealed the diagnosis of myelolipoma.

BACKGROUND

Myelolipoma is a rare benign tumour consisting of mature fat with scattered foci of haematopoietic elements resembling bone marrow.¹ It was first described in 1905, and the term myelolipoma was minted in 1929. There are four proposed types of myelolipoma according to clinical patterns and pathology: Isolated adrenal myelolipoma, adrenal myelolipoma with haemorrhage, extra-adrenal myelolipoma and myelolipoma associated with other adrenal disease.¹ These tumours usually appear in a single location although multiple or even simultaneous adrenal and extra-adrenal myelolipomas have been described.² We describe a case of a thoracic myelolipoma located in the posterior mediastinum.

CASE PRESENTATION

A 64-year-old woman with a history of ischaemic heart disease, non-toxic nodular goitre, asthma and type 2 diabetes (T2D), presented with an asymptomatic leukocytosis of unknown origin. The patient was non-alcoholic and non-smoker. There was no relevant family history. Physical examination revealed no remarkable alterations.

INVESTIGATIONS

During its study a heterogeneous, 1.4×2.5×3 cm left paravertebral thoracic mass was detected by chest tomography. It was predominantly fatty, and probably mesenchyma-derived, with an average value of -90 HU (Hounsfield Units) (figure 1A,B). There were no other alterations such as pleural or pericardial effusion or mediastinal adenopathies. There were no respiratory symptoms. Laboratory studies revealed haemoglobin of 11.8 g/dL, mean corpuscular volume 85.4 fL, platelet count 428 000/μL and white blood cell (WBC) 12 470/μL with 7300 neutrophils, 3700 lymphocytes and 400 monocytes.

To determine the mass nature, a left video-assisted thoracic surgery (VATS) was planned. The removed tumour was brown, fatty and had an elastic texture. It was divided into two pieces, one of 2.5×2 cm and another of 1.5×1 cm. The resected pieces were fixed in formalin, embedded

in paraffin and processed for routine histological examination in H&E. A microscopic study demonstrated predominant mature adipose tissue with interspersed islands of trilinear haematopoietic cells, including megakaryocytes and lymphocytes without any signs of atypia or malignant transformation, consistent with the diagnosis of myelolipoma (figure 1C,D).

DIFFERENTIAL DIAGNOSIS

Mediastinal myelolipomas are generally posterior mediastinum tumours that have to be differentiated from neurogenic tumours such as schwannomas, neurofibromas, ganglioneuromas, ganglioneuroblastomas paragangliomas and from gastroenteric cysts too.³

Extra-adrenal myelolipomas such as mediastinal entities should be differentiated from extramedullary haematopoiesis (EMH), and this cannot be made exclusively by histology. Mediastinal EMH usually occurs at multiple places such as the thoracic paravertebral area and they feature with anaemia, hepatosplenomegaly and many other signs of haematological disease and generally missing adipose tissue, while myelolipomas are usually round well-circumscribed fatty lesions without any other alterations,⁴ even if there are some cases associated with blood findings.

Considering their location, differential diagnosis of both adrenal and extra-adrenal myelolipomas may vary. Relating to abdomen, it is a challenge due to the difficulty in differentiating them from other lipomatous masses such as well-differentiated liposarcomas,² which are the most frequent fatty tumours after benign lipomas.

TREATMENT

AVATS was made in order to remove the tumour.

OUTCOME AND FOLLOW-UP

Postoperatively the patient's recovery was uneventful with a correct radiological follow-up, so she was subsequently discharged. However, WBC remained high after surgery with a total count of 11 880.

DISCUSSION

Myelolipomas occur most frequently in women rather than in men, and more often in elderly patients than in the young ones.²⁻⁴ While adrenal myelolipomas are by far the most frequent entities (they make up from 0.08% to 0.4% of adrenal incidentalomas), extra-adrenal myelolipomas are not a rare finding at autopsies. The most common location of extra-adrenal myelolipomas is the presacral region (50%), while they have been described in the gastric region, liver, lymph nodes, spleen and thorax.⁵⁻⁸

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Figure 1 (A and B) Axial and sagittal CT images of the thorax revealing a heterogeneous lipomatous mass in left paravertebral area. (C) Histological images of the mass. There are fatty areas with interspersed haematopoietic nests (H&E $\times 4$). (D) Details of haematopoietic series, with megakaryocytes, granulocytes and erythroid precursors. (H&E $\times 40$).

Most of the thoracic myelolipomas (3% of myelolipomas) are mediastinal myelolipomas as in the case we describe (and there are only 27 cases in literature). Other thoracic locations such as lung have been reported.⁹

The origin of myelolipomas is not well determined, although there are several theories explaining their aetiology for instance derivation from bone marrow tissue is discussed.^{10–11} Another hypothesis is that this tumour may derive from adrenal metaplasia due to stimuli like necrosis, infection or cell stress that would cause a hamartoma or from migrating haematopoietic stem cells during intrauterine period.^{2–12–13} Some authors believe it to be a neoplastic phenomenon,¹⁴ In fact, some true neoplasms, such as leukaemia located in myelolipomas, have been described.¹⁵ Another possible explanation we propose, and considering that these tumours are attached to vertebral bodies, is that haematopoietic tissue may project from them to paravertebral space, and this could be caused by microfractures. Ectopic haematopoietic tissue may include stem cells that would be the origin of myelolipoma formation.

Radiographic findings are normally similar both in adrenal and extra-adrenal myelolipomas.^{1–9} CT and MRI are the most accurate techniques for diagnosis, even though CT may be a better option in the case of a myelolipoma associated with haemorrhage.¹ Imaging findings show low attenuated lesions due to the lipomatous nature, with isolated foci of high attenuation that maximise with contrast representing the haematopoietic constituent of myelolipomas.^{1–15}

A pathological study reveals a mixed appearance formed by abundant fatty areas with scattered regions of haematopoietic elements. At histological inspection there is a variable blend of adipocytes, myeloid, erythroid precursors and mature cells and even megakaryocytes and lymphoid cells resembling all together bone marrow.¹

Fine needle aspiration or even imaging diagnosis is usually enough to diagnose asymptomatic mediastinal myelolipomas, and there is no need for surgery.⁴ In the case we describe although there were no symptoms the patient had a leukocytosis of unknown origin, hence the tumour was finally removed so as to determine if it had originated that condition.

Learning points

- Myelolipoma is a benign tumour.
- Myelolipomas are bone marrow masses.
- Thoracic location of myelolipomas is uncommon and they make up a differential diagnosis of mediastinal masses.
- Surgical removal of myelolipomas is indicated when they are symptomatic.

Contributors All the authors have contributed for conception and design, gathering of data and analysis and the interpretation of data, and drafting the article and revising it critically for important intellectual content.

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