Autopsy Case Reports

Article / Clinical Case Report

Myelolipoma of the posterior mediastinum in a patient with chronic dyserythropoietic anemia

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ABSTRACT

Myelolipoma (ML) is an uncommon benign mesenchymal neoplasia composed of mature adipose and hematopoietic tissues of uncertain etiology. Less than 3% of MLs occur in the mediastinal topography. The main differential diagnosis involves extramedullary hematopoiesis; therefore, pathological evaluation is essential for the definitive diagnosis. The authors report the case of a 50-year-old man diagnosed with congenital dyserythropoiesis and secondary hemosiderosis, who presented a posterior mediastinal tumor. The tumor was resected. It was macroscopically characterized by mature fat tissue with fibrous areas and soft consistency, which was yellowish at the cut surface. Histology revealed a well-defined nodule composed of adipocytes and hematopoietic tissue represented by erythroid, granulocytic, and megakaryocytic series, which was consistent with the diagnosis of ML located in the posterior mediastinum. There was no recurrence of the lesion during the 3-year follow-up. The aim of this report is to show the diagnosis of an unusual mediastinal lesion in the context of a chronic hematologic disease.

Keywords
Myelolipoma; Mediastinal Neoplasms; Mediastinum; Anemia

INTRODUCTION

Myelolipoma (ML) is a benign mesenchymal neoplasm composed of hematopoietic and mature adipose tissue.[1] The incidence of this tumor is estimated in up to 0.2% of mesenchymal tumors, and it can occur in various parts of the body, but predominantly in the adrenals. The extra-adrenal location is unusual (15%) and includes the retroperitoneum, pelvis, pre-sacral region, stomach, and liver. In the lungs and mediastinum, the occurrence of ML is quite infrequent accounting for 3% of all cases.[1,2] This report shows an rare case of ML located in the posterior mediastinum in a patient with congenital dyserythropoiesis.

CASE REPORT

A 50-year-old Caucasian male patient, previously diagnosed with congenital dyserythropoiesis, secondary hemosiderosis, and systemic arterial hypertension,

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presented a nodule in the right posterior mediastinum. Despite the diagnosis of chronic anemia, the patient was asymptomatic and was in steady control of the hemoglobin and the hematocrit levels. In a routine thoracic and abdominal computed tomography (CT), a nodule with soft tissue density, which measured 3.5 cm in diameter, was detected with enhancement after intravenous contrast injection localized in the right posterior mediastinum, in paravertebral topography (Figure 1).

The patient was a former smoker and underwent a splenectomy and a liver biopsy 5 years ago. The spleen was congested and weighed 1280 g (reference value 150 g). Histologic review of the spleen and liver slides revealed splenic congestion, and hemosiderin deposits in both the spleen and the liver (Figure 2A to 2D). Immunohistochemistry failed to show myeloid metaplasia in the splenic samples. The patient did not have a bone marrow biopsy.

In this clinical and radiological context, the video-assisted thoracic lesion resection was indicated considering the diagnostic hypothesis of paraganglioma, schwannoma, and nodal hematopoietic tumor infiltration. The gross examination of the surgical specimen revealed an oval-shaped tumor mass with a lobular surface, covered by a thin membrane, with soft consistency measuring 5.6 × 3.7 × 1.9 cm and weighting 10 g (Figure 3A). Histopathology revealed a well-defined nodular lesion (Figure 3B) predominantly consisting of adipose tissue, intermingled with hematopoietic tissue, which was represented by an erythroid, granulocytic, and megakaryocytic series.

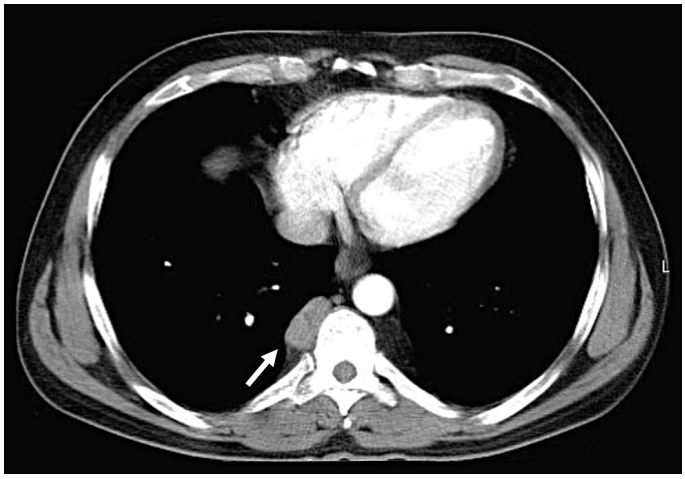

Figure 1. Thoracic axial computed tomography showing a paravertebral nodular lesion in the right posterior mediastinum with soft tissue density (arrow).

Immunohistochemistry positivity for glycophorin A, myeloperoxidase, and Factor VIII confirmed the presence of erythroid cells, granulocytic lineage, and megakaryocytes, respectively, in the surgical specimen, which was consistent with the diagnosis of ML (Figure 4A-C). The patient was followed-up for 3 years without relapses; however, he was submitted to monthly bloodletting.

DISCUSSION

MLs are benign and nonfunctioning tumors composed of mixed hematopoietic and mature adipose tissue, in varying proportions, which are usually located in the adrenal glands.[3,4] Since the first descriptions in the early 20th century,[1] other topographies have been reported in (i) the soft tissue; (ii) the retroperitoneum; (iii) the spleen; (iv) the liver; (v) the stomach; and (vi) the nasal cavity.[4] The extra-adrenal MLs occur in up to 15% of cases.[5,6] The lungs and mediastinum are unusual sites for such occurrences. In a literature review, Xu et al.[5] reported 10 cases of intrapulmonary ML: 8 had isolated lesions, and 2 had a multifocal clinical presentation. The mediastinum is the site of occurrence in approximately 3.0% of cases.[4]

In a recent study, Xiong et al.[3] described the clinical and pathological aspects of 28 cases of mediastinal MLs.[3] In this series, the symptoms, when present, were nonspecific and were represented by changes in the respiratory system (10%), such as dyspnea, hematopoietic abnormalities (9%), endocrine disorders (6%), and cardiovascular disorders (5%). Most cases (93%) occurred in the posterior mediastinum as a single lesion. The mean age of these 28 patients with mediastinal MLs was 64 years, with a slight male predominance. The series reported by Shen et al.[2], which comprised 16 patients with thoracic ML, showed singular chest lesions in 13 cases.[2] Patients with extra-adrenal MLs were older than those with adrenal lesions.[6]

The diagnosis of ML is often made incidentally by imaging studies, such as CT and magnetic resonance, due to the scarcity of typical symptoms. Similarly, imaging findings are also nonspecific.[4] The reported tumoral mass of our patient was described as a nodular

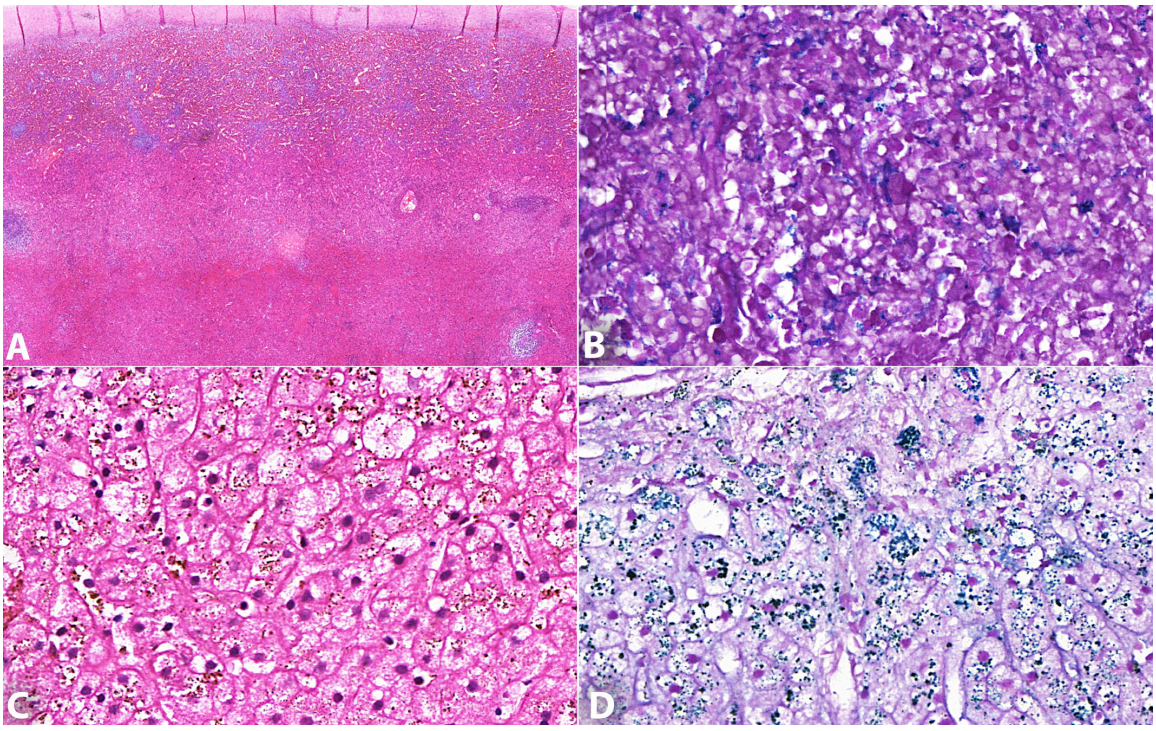

Figure 2. A and **B** - Photomicrography of the spleen; **A** - Splenic congestion, hypoplastic white pulp, and an absence of myeloid metaplasia (H&E, 200X); **B** - Hemosiderosis: hemosiderin deposits stained in blue (Perls, 400X); **C** and **D** - Photomicrography of the liver; **C** - Hepatocellular ballooning, and hemosiderin deposits (H&E, 400X); **D** - Hemosiderosis: hemosiderin deposits stained blue (Perls, 400X).

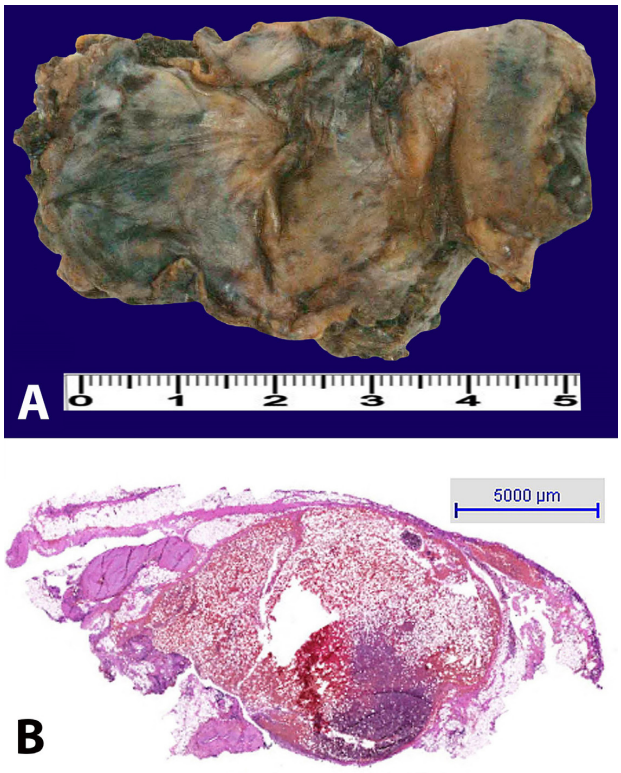

Figure 3. A - Gross appearance of the oval shaped and lobular mediastinal tumor; **B** - Photomicrography of the myelolipoma: a panoramic view of the histological section of the specimen showing a circumscribed nodular area primarily consisting of adipose tissue (H&E, low magnification view).

lesion with soft tissue density, which showed some high-intensity regions with attenuation enhancement after intravenous contrast injection. This nonspecific imaging pattern of presentation supports the differential diagnosis of bronchial malignancies, lymphoproliferative disorders, neurogenic tumors, and lipomatous and metastatic lesions.[1,4] In this setting, the gold standard examination for the diagnosis of ML is the histological examination.[1,4] Macroscopically, generally, the ML are well-defined tumors that are often enclosed by a thin capsule measuring approximately 4-5 cm in diameter.[2] The cut surface can be variegated, depending on the ratio between fat and hematopoietic tissues. In our patient, the lesion surface had a pale yellowish aspect due to the increased adipose tissue component. The histological examination showed the varied proportion of hematopoietic tissue intermingled with adult adipose tissue. The hematopoietic component was recognized by the presence of erythroid, granulocytic, and megakaryocytic series. In our patient, there were no bone spurs or hyperplasia of any hematopoietic component, and the adipose component did not present atypia.[2,4]

The pathogeny of ML is unknown. Shen et al.[2] and Xiong et al.[3] compiled some possible explanations for

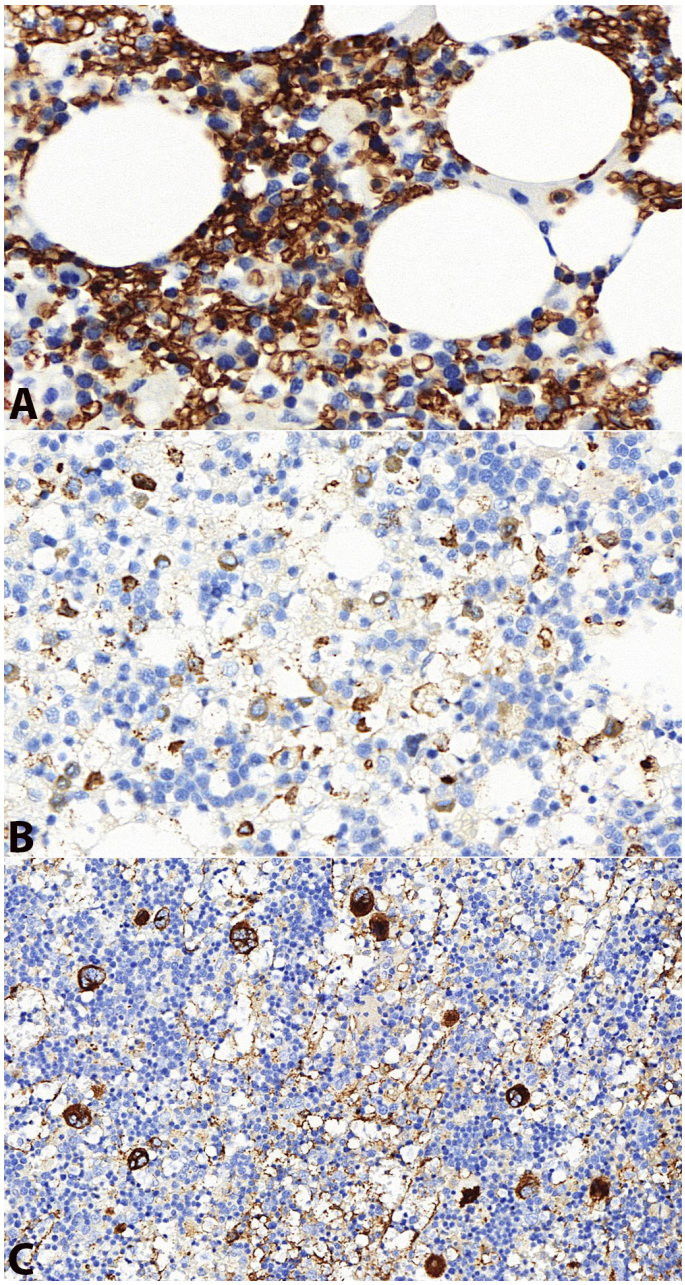

Figure 4. Photomicrography of the surgical specimen. **A** - Glycophorin positivity in the erythroid series colonies (Immunohistochemistry, 400X); **B** - Granulocytic lineage confirmed by positivity for myeloperoxidase (Immunohistochemistry, 400X); **C** - Megakaryocytes highlighted by the positivity for Factor VIII (Immunohistochemistry, 200X).

the genesis of ML: (i) it could be derived from bone marrow emboli that may lodge in different regions of the organism; (ii) it could be derived from embryonic primitive mesenchymal cells; (iii) it could be derived from the metaplastic transformation of embryonic stromal cells after chromosomal translocation, similar to that observed in myelogenous leukemia; and iv) some disorders, such as the endocrine diseases like Cushing's syndrome, diabetes mellitus, Addison's disease, Cohn's syndrome, pheochromocytoma, obesity, and even hypertension that accompany ML may play a role in the genesis of this neoplasm.[2,3,] Fonda et al.[7] proposed that ML may derived from projection of hematopoietic tissue (including stem cells) by bone microfractures. Some authors emphasize that the development of ML may be associated with prolonged and excessive steroid production or genome defects of the endocrine glands responsible for multiple endocrine neoplasia type 1.[7,8]

In general, patients with ML do not present with hematopoietic disorders. The presence of anemia or blood disorders does not exclude the diagnosis. The patient in this report presented the diagnosis of congenital dyserythropoiesis with hemosiderosis due to repeated blood transfusions, having undergone a splenectomy. Congenital dyserythropoiesis is a rare, inherited anemia that leads to the failure of erythroid maturation accompanied by hemosiderosis and extramedullary hematopoiesis.[9] In this patient's case, it is possible that the persistent erythropoietin stimulation, due to chronic anemia, may have played an important role in tumor development. It is interesting to note that extramedullary hematopoiesis occurs as mediastinal and paravertebral masses.

In this clinico-pathological context, an important differential diagnosis is extramedullary erythropoiesis. This is a commonly multifocal lesion that, when presented in the chest, develops in the perivertebral and mediastinal topography.[9] It is histologically characterized by an irregular shape, with a predominance of hematopoietic elements, especially erythroid and myeloid series; however there also may be hyperplasia of erythroid series.[6,10] In contrast, our patient had chronic anemia due to the underlying blood disease and had a well-defined singular nodular lesion discovered incidentally on routine imaging. Additionally, the microscopy showed an abundant adipose component and all the hematopoietic series without atypia. These aspects were consistent with the diagnosis of ML.

The most adopted therapeutic approach is to remove the lesion by conventional thoracotomy or a video-assisted method, depending on the size of the lesion. The prognosis is usually excellent.[1-3]

CONCLUSION

This report presents the case of a patient with congenital dyserythropoiesis, with secondary hemosiderosis, whose tomographic follow-up examination revealed the presence of a tumor in the posterior mediastinum. The tumor was removed, and the pathology confirmed the diagnosis of ML—a rare tumor that was found in an unusual topography.

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