



Mediastinal Myelolipoma Diagnosed by Transthoracic Fine-Needle Core Biopsy

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Abstract

Extra-adrenal Myelolipoma (ML) is rare, non-functioning and generally asymptomatic tumor, composed by a mixture of adipose tissue and hematopoietic cells. It usually occurs in the adrenal glands and thoracic location is extremely unusual. We report the case of an 80-year-old woman without specific symptoms, with chest CT tomography showing incidentally a paratracheal mass (55 mm × 62 mm). Transthoracic fluoroscopic-guided fine needle biopsy was performed. Pathologic examination revealed ML, confirmed after surgical resection.

Keywords: Mediastinal mass; Myelolipoma; Transthoracic fine needle biopsy

Introduction

Described for the first time by Oberling in 1905 [1], Myelolipoma (ML) is a rare, benign, non-functioning neoplasm, of unknown origin, consisting of mature adipocytes and normal hematopoietic cells [2,3]. It is usually found as a solitary nodule or mass, accidentally detected in the adrenal glands [1], but about 15% of them are found in an extra-adrenal site [4]. Thorax localization is extremely unusual (3%) [4], and various sites may be involved, like mediastinum [2,3], lung parenchyma [5], chest wall [1] or main bronchus [6]. We report a case of mediastinal myelolipoma diagnosed by transthoracic fine needle biopsy and confirmed after surgical excision.

Materials and Methods

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An 80-year-old woman, never smoker, was evaluated in our department for a mediastinal mass diagnosed incidentally 5 months before, when the patient went to the Emergency Department for acute dyspnea. Chest-CT scan revealed the presence of pulmonary embolism associated with a well-defined round paratracheal mass (55 mm × 62 mm), with inhomogeneous aspect and enhancement after application of iodinated contrast media. Chest CT-scan performed 2 months later confirmed these finding (Figure 1). The patient was then admitted in the Pulmonary Diseases Unit of our Hospital for further investigations. Past medical history revealed: Chronic ischemic heart disease, obesity, IgM Monoclonal Gammopathy of Undetermined Significance (MGUS), hyperhomocysteinemia and mild renal failure. At the time of admission, the patient was alert and cooperative, with regular rhythm and heart rate (80 beats/min), breath rate of 13 breath/min, blood pressure of 130/70 mmHg and body temperature of 36.5°C. The oxyhemoglobin saturation on room air was 95%. Chest physical examination disclosed diffused reduced vesicular breathing without pathological sounds. Laboratory findings, respiratory function tests and electrocardiography were normal. Since the lesion was adjacent to the trachea, we proposed a diagnostic approach with Echo Bronchoscopic Guided Transbronchial Needle Aspiration (EBUS-TBNA), but the patient refused bronchoscopy. Therefore, we decided for a percutaneous fine needle core biopsy. A first sample of the mass was made using a 21G needle, under X-ray fluoroscopic guidance, with posterior paravertebral approach. On the same target biopsy with 18G needle (TSK, Japan) was performed. Specimen obtained was swiped on slides, immediately fixed on alcohol and dyed with a rapid staining method (Hemacolor Merck, Germany) and Rapid on Site Evaluation (ROSE) was done in order to assess the adequacy of the sample and the correct centering of the lesion. Since different



Figure 1: CT scan showing well-defined round paratracheal mass (55 mm × 62 mm).

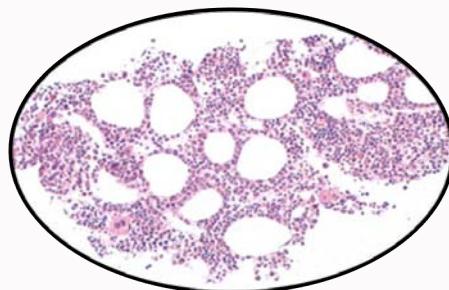


Figure 4: Biopsied fragments. There is a mixture of lipocytes and myeloid cells- all the hemopoietic lineages are recognizable (insert).

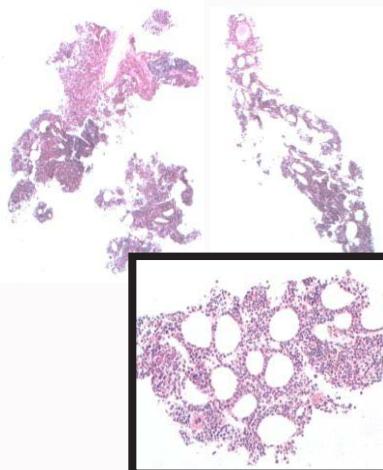


Figure 2: Biopsied fragments. There is a mixture of lipocytes and myeloid cells- all the hemopoietic lineages are recognizable (insert).



Figure 5: Two sections of the surgical specimen. The mass was yellow with brownish zones.

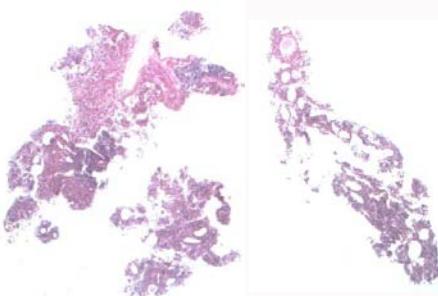


Figure 3: Biopsied fragments. There is a mixture of lipocytes and myeloid cells- all the hemopoietic lineages are recognizable (insert).

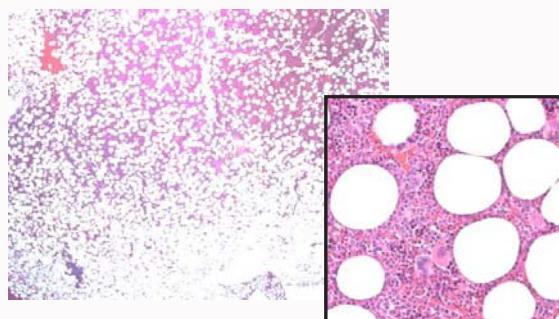


Figure 6: Surgical specimen. Adipocyte cells intermingled with hematopoietic tissue.

hematopoietic cells associated with mature adipocytes were observed, suggesting the hypothesis of ML, a biopsy with an 18G core-needle (Aeccut, TSK Japan) was done. No complications occurred. Final histological examination of the biopsy material confirmed the diagnosis of ML (Figures 2-4). Considering the size of the lesion and the close contiguity with the vena cava, a surgical treatment of the lesion was proposed, initially rejected by the patient and later accepted. She underwent Video-Assisted Thoracic Surgery (VATS) with complete resection of the mass. The histological evaluation of the surgical material confirmed the diagnosis of ML (Figure 5, 6). Follow-up one year after surgery did not show recurrence of the disease. ML is a rare, benign, mesenchymal tumor [2], commonly located in the adrenal glands (75%), and only occasionally it is symptomatic with endocrine disorders [3]. The mediastinal location is very rare

and asymptomatic. When present, symptoms are non-specific like cough, dyspnea [7] or chest pain [3]. Thoracic localizations are less common (3%) and, although bilateral lesions are reported [4], they are most commonly unilateral [4]. Usually, diagnosis is obtained after surgical excision using standard thoracotomy [2,4] or under video-assisted thoracoscopy surgery [8], but CT-guided needle biopsy [9] or endobronchial ultrasound-guided transbronchial needle aspiration [10] can be diagnostic. In our case diagnosis was suspected during rapid on-site evaluation of cytological specimen and was confirmed by histological evaluation (Figures 2-4). At CT scan ML appears as an inhomogeneous lesion, of various sizes, which may enhance after injection of iodinated contrast. A well-defined capsule is often detected [3,11]. This aspect is due to concomitant presence of a mixture of two different mature components: Fatty

and hematopoietic tissue (the latter is responsible for enhance after injection of iodinated contrast). Radiological differential diagnosis is with lymph nodes metastasis, liposarcoma, neurogenic tumors or neurofibromas, extra-adrenal pheochromocytoma, lymphoma, Extramedullary Hematopoietic Tumors (EHT) [4] and hamartoma [12]. Differential diagnosis with EHT is often difficult: chest CT-scan shows similar aspect, because both entities are characterized by presence of hematopoietic elements and adipose cells, however EHT occurs at multiple sites and they are generally associate with anemia [4,13]. In our case patient had only a solitary mediastinal mass and her blood count was normal. Histologically, mediastinal ML is similar to ML of adrenal glands and it is characterized by different ratios of mature adipose tissue and hematopoietic cell, resulting very similar to bone marrow [5,12]. Usually, hematopoietic tissue is represented by erythroid, myeloid and megakaryocytic stems, whereas lymphocytes are less common [3,12]. Pathological origin of ML is still debated. At the present there are three main theories of its pathogenesis: a) it could have an origin from embryonic mesenchymal cells, b) it could be due to an extramedullary location of bone marrow emboli [12] c) it could derive from a metaplasia of reticuloendothelial cells [1]. The theory of clonal origin of ML is confirmed by Bishop et al. [14] who describes the same pattern of X-chromosome inactivation in hematopoietic elements and fat. Furthermore, the origin from bone marrow is suggested by similar chromosomal translocation in ML and benign lipomatous neoplasms [15]. Finally, a genome defect of glands or excessive steroid production could play a role in natural history of ML, as suggested by the association with various endocrine disorders like Addison's disease, diabetes mellitus, obesity, pheochromocytoma and Cushing's syndrome [1]. The management of thoracic ML is still debate but, if the tumor is small and asymptomatic, a radiological follow-up is recommended. In case of larger and symptomatic mass or if the tumor growth may compromise the function of adjacent structures, surgical excision is the best choice [3,8].

Conclusion

Our case demonstrates that, although myelolipoma remains a rare lesion, it should be considered among differential diagnoses of asymptomatic thoracic mass. Transthoracic fine-needle core biopsy is a minimally invasive technique very useful for the diagnosis of mediastinal masses that must be considered when the lesion is not reachable by ultrasound guided endoscopic transbronchial needle aspiration or when patient refuses endoscopic approach.

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