Pulmonary myelolipoma containing osseous tissue: An unexpected finding at autopsy

Silvija Mašić a,⁎, Majda Vučić b, Sven Seiwerth c, d

⁎ Corresponding author.
E-mail addresses: silvijamasic57@gmail.com (S. Mašić), majda.vucic@kbscm.hr (M. Vučić), sven.seiwerth@mef.hr (S. Seiwerth).

Abstract

Pulmonary myelolipoma is a very rare benign tumor composed of mature adipose tissue and hematopoietic elements such as erythroid, myeloid and megakaryocytic. It usually represents accidental finding during autopsy or chest imaging, since most cases are asymptomatic. Larger masses can lead to hemorrhage, chest pain and chest organ compression. We present a case of incidental finding of pulmonary myelolipoma during the autopsy of an 83-year-old woman who died of abdominal aortic rupture. In the right lower lung lobe, solitary, well-circumscribed yellow-brown nodule which was 3 cm in its longest diameter was found. Pathohistological analysis revealed tumor composed of mature adipose tissue and hematopoietic cells (myeloid cells, megakaryocytes, erythroid cells) with fragments of mature bone tissue. Differential diagnosis of pulmonary myelolipoma includes lipoma, liposarcoma, hamartoma, phlebangioma, teratoma and extramedullary hematopoiesis. In majority of cases, tumor removal is not necessary, however, larger lesions should be surgically removed. No cases of malignant transformation or recurrence have so far been reported in the literature.

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aortic rupture which was 4.5 cm in diameter and was identified as the cause of lethal outcome in our patient. Besides pulmonary cyanosis, in the right lower lobe, well-circumscribed yellow-brown nodule, which was 3 cm in size and appeared calcified when cutting the tissue, was found (Fig. 1).

Routine hematoxylin & eosin staining of the nodule revealed a lesion composed of mature adipose tissue and hematopoietic cells (myeloid cells, megakaryocytes, erythroid cells) with fragments of mature bony tissue (Fig. 2) suggesting the diagnosis of pulmonary myelolipoma. Adipose tissue involved around 50% of the lesion. No foci of extramedullary hematopoiesis have been found in liver and splenic parenchyma.

3. Discussion

Etiology of myelolipoma is still not entirely clarified. More theories concerning formation of this lesion exist, reporting possible myelolipoma development from embryonic mesenchymal cells originating from adrenal gland, emboli derived from hematogenous bone marrow, reticuloendothelial cell metaplasia and proliferation of hematopoietic stem cells [1–4,6,8]. Usually no symptoms are related to myelolipoma [7], however, according to available literature hemorrhage and compression of adjacent structures can occur. Cases of pulmonary lesions have been described as being related to lumbar spine pain, fever, cough, pneumonia, bronchiectasis, pulmonary atelectasis [3,7,9]. Other locations were related to chest pain, back pain, renal pain, dysuria [6,9,11]. Also, in patients with cortisol overproduction, virilization, Conn’s and Cushing’s syndrome myelolipomas are often found [1,3,4,11].

Macroscopically, the tumor usually appears as well-circumscribed, yellowish with red-brown areas intrapulmonary lesion [3,4]. Color depends on the amount of lipid component and hematopoietic elements [3]. Microscopically, myelolipomas are described as tumors composed of mature fatty tissue and hematopoietic tissue, including myeloid, erythroid, megakaryocytic cells, lymphocytes with possible finding of hemorrhage, bone segments, cartilage and calcifications in the tumor [8,9,11].

Bone tissue represents a questionable finding. Fowler et al. presented a case of myelolipoma without bony elements and consider unusual for them to be encountered in this tumor [2]. However, Sabate and Shahian, for example, reported a case of pulmonary myelolipoma containing bony structures [1]. Also, our case represents tumor with bony segments. So far, to the best of our knowledge, clinical significance of this debatable finding has not been determined.

Diagnosis of myelolipoma is established through performing imaging techniques (chest x-rays, CT scan, MRI) [3,9], but for final diagnosis establishment, pathohistological examination is required [7,11]. CT scan reveals myeloid parts of intrapulmonary myelolipoma to be higher in attenuation than adipose segment of the tumor [6], while on MRI, myeloid segments show low signal intensity and adipose tissue high intensity on T1-weighted imaging. Adipose tissue also demonstrates high signal intensity and myeloid parts intermediate signal intensity on T2-weighted imaging [7].

Differential diagnosis includes extramedullary hematopoiesis, hamartoma, lipoma, liposarcoma, phlebangioma and teratoma. Extramedullary hematopoiesis rarely occurs in lungs, but in contrast to myelolipoma, it usually appears as multiple foci without

Fig. 1. Cut surface of the right lower lobe of the lung containing yellow-brown nodule 3 cm in size. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

Fig. 2. HaE: 2.1 Well-demarcated lesion surrounded by bony spicules (HaE x 10 Magnification). 2.2 Tumor composed of mature adipose tissue and hematopoietic cells (myeloid cells, megakaryocytes, erythroid cells) (HaE x 40 Magnification).
well-circumscribed margins, it is mostly comprised of erythroid hyperplasia and hematopoietic elements, deprived of lymphoid tissue and contains small amount of fatty tissue [3,4,8,9]. It is usually related to other hematological conditions like anaemia or myelo-proliferative conditions, which is not characteristic for myelolipoma [2,4,8,9]. Lipoma is benign tumor macroscopically characterized as well-defined, yellowish lesion histologically composed of mature fatty tissue. It is considered to be difficult to recognize in cases when myelolipoma contains mostly fat [7,8]. Liposarcoma is malignancy of adipose tissue, but unlike myelolipoma, usually demonstrates infiltrative growth [6]. Hamartoma is benign tumor, but more frequently observed than myelolipoma. Unlike to forementioned, it contains fatty tissue, fibromyxoid stroma, cartilage, bone and bronchial cells, without hematopoietic elements [7,8]. Teratomas are potentially malignant tumor composed of tissue originating from three germinal layers [7,8]. Xu et al. report that difficulty in distinguishing these two entities occurs when myelolipoma contains mostly hematopoietic cells. In those cases resolution lies in performing percutaneous needle biopsy [7]. Phlebangioma is considered to be uncommon lesion characteristic for abnormally ectasic venous vessels [12] that can be distinguished from myelolipoma by performing CT examination in order to reveal fatty tissue [7]. Myelolipomas should also be distinguished from metastasis, since cases with poorly demarcated and multiple lesions can occur and make establishing correct diagnosis difficult [4].

No surgical or other treatment of myelolipoma is required, however, in symptomatic patients and in cases of large lesions (larger than 7 cm) or ones with progressive growth, surgical removal is recommended [3,7].

4. Conclusion

Myelolipoma is a rare lesion that usually occurs in adrenal gland, while lungs and other sites represent less common finding. Although being a benign lesion and usually incidentally discovered, it is important to establish the diagnosis of myelolipoma, especially in cases of larger lesions with progressive growth [6,11], which can cause tumor rupture and eventually life threatening hemorrhage or compression of adjacent structures, since those should be surgically removed [1,7,9]. Small asymptomatic lesions (less than 4 cm in size) should only be monitored [3,9]. Also, despite the fact that no cases of myelolipoma malignant transformation have been reported [8], distinguishing it from malignant lesions is necessary in order to adequately treat the patient [7].

Conflicts of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

References