58-year old female with posterior mediastinal mass

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Clinical history

The patient is a 58-year-old female who is radiologically followed for a posterior mediastinal mass discovered incidentally in 2014. The last imaging studies reveal a right paravertebral 2,0 x 1,5 cm mass at T9, with SUV of 5.0 (Figure 1), and the mass is surgically resected.

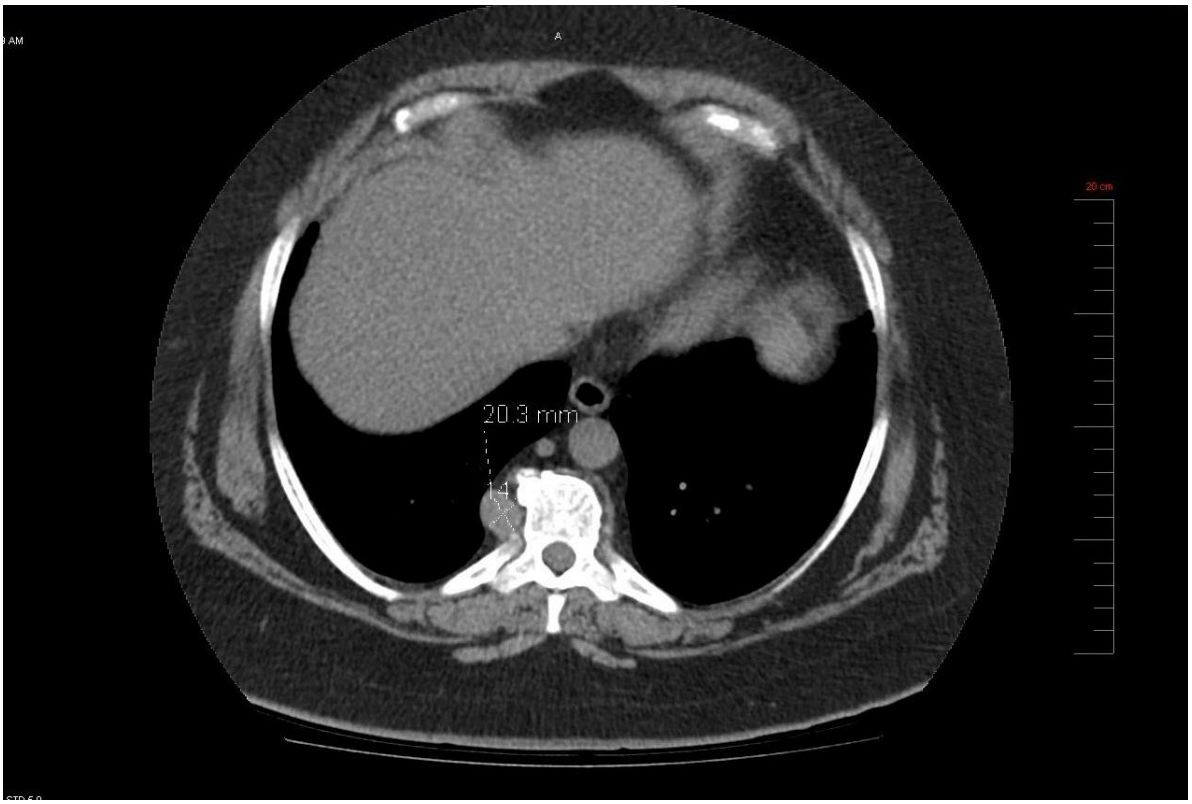

Figure 1: Right paravertebral mass, computed tomography scan.

Histology:

The mass is mostly composed of benign hematopoietic tissue with normal trilineage maturation, and with appropriate myeloid to erythroid ratio (M:E ratio of 3:1). The megakaryocytes are appropriate in number and morphology (Figures 2-4). Fibroadipose tissue, nerves and ganglion cells (likely originating from the sympathetic trunk) are also present (Figure 5). Of note, the adipocytes are not admixed with the hematopoietic tissue (Figure 6).

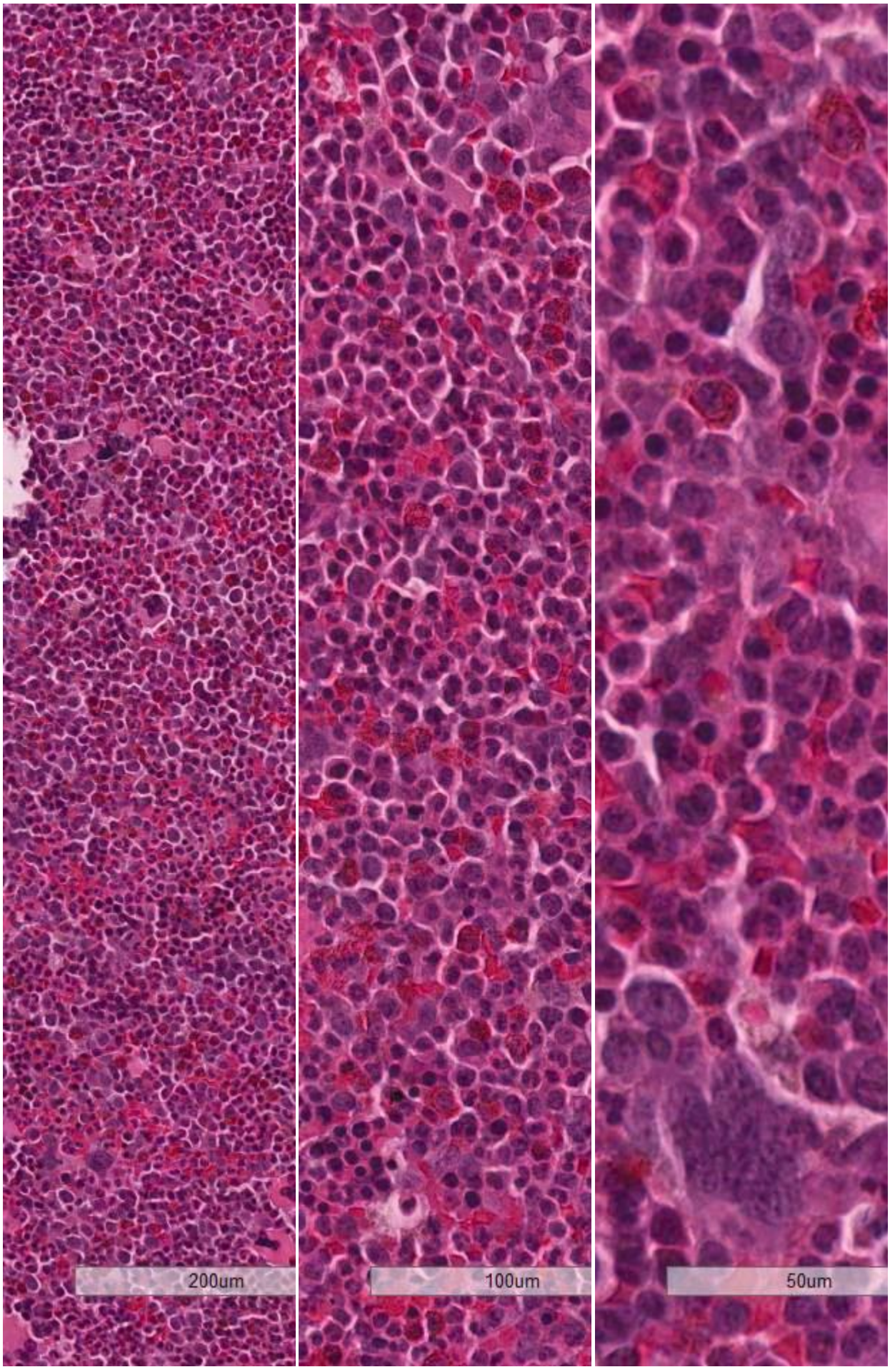
200um
100um
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Figure 2: Benign hematopoietic tissue, H&E, 100x.

Figure 3: Benign hematopoietic tissue, H&E, 200x.

Figure 4: Benign hematopoietic tissue, H&E, 400x.

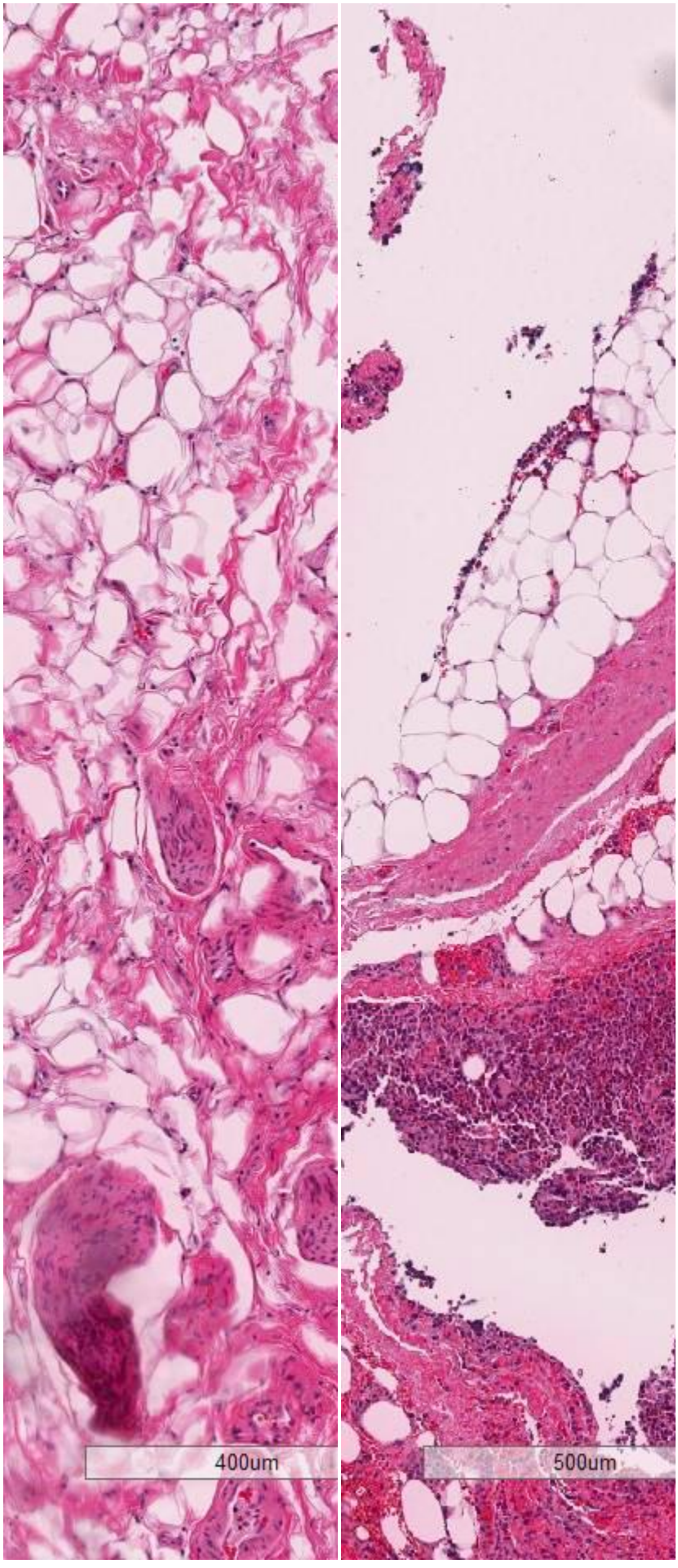

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Figure 5: Benign ganglia and fibroadipose tissue, H&E, 50x.

Figure 6: Relation between adipose and hematopoietic tissues, H&E, 40x.

Discussion

The differential diagnosis of tumoral (mass-forming) hematopoiesis includes myelolipoma and extra-medullary hematopoiesis (EMH), two entities that have very similar histology but different clinical contexts.

Myelolipomas

Myelolipomas are usually found in the adrenal glands but have also been described in other sites (kidney, liver, mediastinum, retroperitoneum, presacral, testis)[1,2]. The pathophysiology of myelolipomas remains unclear, although later findings point towards a clonal process[1]. Hypotheses on stem cells and hormonal factors have also been formulated[2]. Those tumours are classically described as incidental masses in patients older than 40-years-old, and are round, small, and solitary on imaging[2,3]. The most helpful distinction is that myelolipomas are typically not associated with hematological disorders[1]. It is a benign entity, although it has been seen with adrenal cortical tumours, congenital adrenal hyperplasia and pheochromocytomas[1,2]. On histology, myelolipomas are composed of benign hematopoietic tissue and mature adipocytes[1,2]. The hematopoiesis contains trilineage maturation and has been described with "markedly increased megakaryocytes"[4]. The adipocytes are usually admixed within the hematopoietic foci[1].

Extra Medullary Hematopoiesis

This entity also known as myeloid metaplasia is usually seen in the liver and the lymphoid tissues but has been described in other sites (mediastinum, kidney, bladder, uterus, breast, skin)[1]. EMH is usually a microscopic finding, which can occur essentially in any organ as well as in body cavity fluids. However, it may occasionally present as a mass-forming lesion, which can simulate a neoplasm clinically and radiologically, as in this case. In the mediastinum, it is usually radiologically described as large, solitary, irregular and paravertebral mass[5,6].

EMH occurs when there is an overproduction of one or more types of bone marrow elements or whenever the bone marrow is replaced by either tumor or fibrosis. Therefore, the clue to the diagnosis of EMH is that it is usually seen in the context of hematological disorders, such as thalassemia, hereditary spherocytosis, and other conditions such as metastatic carcinoma effacing the marrow space and drug therapies[1]. It is a benign entity, but has been described with certain tumours (hemangioblastomas)[1]. The pathophysiology of EMH also remains unclear.

On histology, it appears as benign hematopoiesis with trilineage maturation. Histological clues to extra-medullary hematopoiesis have been described, including increased erythroid precursors[3,5], and higher hematopoietic tissue density[7]. The trilineage should theoretically be reflective of the underlying hematological disorder, but this is difficult to appreciate in practice. When presenting as a mass-forming lesion or in body cavity fluids, EMH may also be diagnosed by (fine needle aspiration) cytology. The diagnosis is relatively easy when trilineage hematopoiesis is

recognized, and no special stains are required in the appropriate clinical context (ie, known hematological disorder)[8]. However, due to the bizarre giant cells (megakaryocytes) and accompanying inflammatory background, especially in the absence of a known hematologic abnormality, the cytomorphologic differential diagnosis may include metastatic carcinoma, Hodgkin lymphoma and myeloid sarcoma.[8]

Final diagnosis

Right mediastinal mass, excision: Most consistent with extra-medullary hematopoiesis (see comment) with benign fibroadipose tissue, nerves and ganglia. Negative for malignancy

Comment: The favored diagnosis is a nodule of extra-medullary hematopoiesis, which has been described in the posterior mediastinum. A differential diagnosis would include a myelolipoma, which can also be found in this location but in these lesions, there is typical infiltration of the hematopoietic tissue by adipose tissue, a feature that is absent in our case. Extra medullary hematopoiesis is associated with underlying hematological disorders (hereditary spherocytosis, thalassemia, myelofibrosis, chronic anemia amongst others) and as such, clinical correlation is recommended.

Note: unfortunately, no follow-up was available in our case.

References

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