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Case report

An autopsy case of an elderly man with myelodysplastic syndromes in which postmortem computed tomography showed massive splenomegaly

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ABSTRACT

Here we describe the findings of autopsy of an elderly man with several critical diseases, including myelodysplastic syndromes (MDS), who experienced sudden natural death in which postmortem computed tomography (PMCT) provided useful information in determining the cause of death.

A man in his late 80s suffered a cardiopulmonary arrest at his home. Although previously diagnosed with MDS and chronic heart failure, he had not consulted a physician for several years. PMCT images showed massive splenomegaly and consolidation in the right lung, whereas signs suggestive of pulmonary edema were not evident in the left lung. Macroscopic autopsy findings included splenomegaly (1748 g) and dark red coloring of the right lung, but no significant heart enlargement (366 g). Microscopic findings included extensive hemorrhage in the alveolar space in the right lung, hypercellular bone marrow with monotonous cell proliferation, and infiltration of those cells in various organs. Immunohistochemical staining showed that the proliferating cells were strongly positive for myeloperoxidase. Based on review of the autopsy findings, we concluded that the underlying cause of death was acute myeloid leukemia.

Review of this case suggests that PMCT may be useful in identifying the cause of death of elderly persons with several critical diseases who experience sudden natural death. Utilizing PMCT findings may allow for compilation of accurate mortality statistics regarding the elderly population, although physicians should consider the limitations of PMCT when deciding whether to perform autopsy.

1. Introduction

Myelodysplastic syndromes (MDS) are a group of heterogeneous bone marrow disorders characterized by ineffective hematopoiesis [1–6]. The natural evolution of disease in MDS consists of bone marrow failure leading to infectious and hemorrhagic episodes or anemia-related complications and transformation to acute myeloid leukemia. Because this group of disorders displays remarkable clinical, pathologic, and cytogenetic heterogeneity, the median survival varies greatly from months to years [3,4]. As MDS is a disease of older adults [3,5], the mean age at onset is over 70 years, and about one-third of MDS patients succumb to other diseases unrelated to MDS [3].

A previous report suggested that the appearance of splenomegaly in MDS may indicate a transformation to myeloproliferative disorder [7]. According to the World Health Organization classification of neoplasms of hematopoietic and lymphoid tissues, one type of neoplasm can be classified as both MDS and a myeloproliferative neoplasm (MPN), and is accordingly referred to as a MDS/MPN [8,9]. Splenomegaly is found in some patients of MDS/MPN and is generally associated with poor outcome [8].

Here we describe the case of an elderly man with both MDS and chronic heart disease who experienced sudden natural death. As the patient had been suffering from two chronic diseases, it was difficult to determine the most proximate cause of death by external examination. However, the examination of postmortem computed tomography (PMCT) images in consideration with the patients' medical history suggested deterioration of MDS instead of heart disease, and subsequent forensic autopsy and histopathology revealed the underlying cause of death to be acute myeloid leukemia.

2. Methods

Whole-body PMCT was performed before autopsy with a 64-row CT scanner (Somatom Definition AS, Siemens Healthcare, Forchheim, Germany) with the following parameters: 120 kV; quality reference mAs: 400; thickness: 64 × 0.6 mm; and field of

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view: 500 mm. Image data were analyzed by forensic radiologists using syngo.via software (Siemens AG).

3. Case report

A man in his late 80s who was living with his family suddenly lost consciousness and suffered cardiopulmonary arrest at his home, after unintentionally passing urine. He was transported to a critical care center in an ambulance, but died before he could be admitted. Although a review of his records indicated that he had been diagnosed with MDS and chronic heart failure, he had not consulted a physician for several years. According to his family, he had complained of general fatigue for three days prior to his death.

An external examination performed approximately 19 h after death did not reveal any open injuries. He had been in a supine position at the time of death, and a slightly weak postmortem livedo was observed on his back. The deceased was 151 cm tall and weighed 47 kg, with a body mass index of 20.6 kg/m^2.

Unenhanced PMCT scanning was performed approximately 36 h after death. The main findings of review of the PMCT images were massive splenomegaly and ground glass opacity/consolidation in the right lung (Fig. 1a and b). Signs suggestive of pulmonary edema were not evident in the left lung.

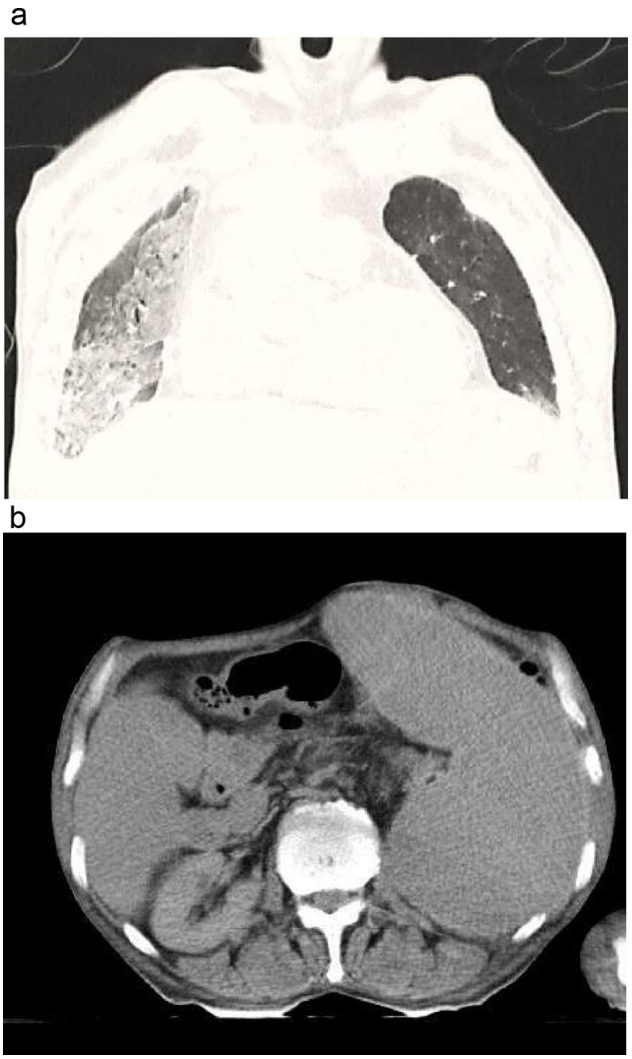

Fig. 1. Postmortem CT images (a: chest, coronal section, b: abdomen, axial section). (a) Ground glass opacity/consolidation was observed in the right lung. Signs suggestive of pulmonary edema were not evident in the left lung. (b) Massive splenomegaly was observed.

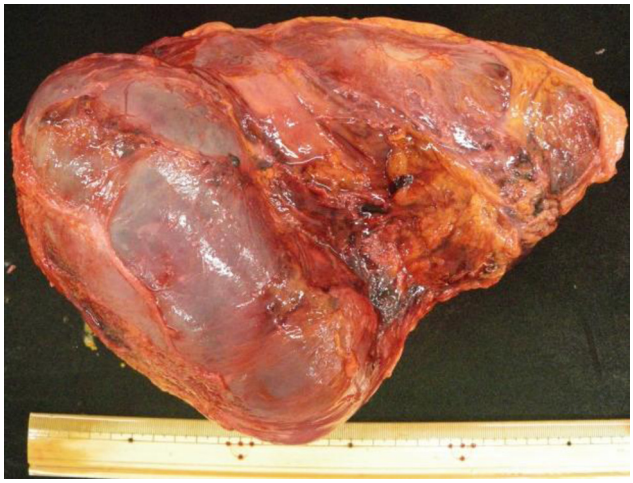

Fig. 2. Macroscopic autopsy findings. Splenomegaly (1748 g) was noted.

Forensic autopsy was performed approximately 38 h after death to determine the cause of death. Autopsy revealed massive splenomegaly (1748 g; Fig. 2), but no significant enlargement of the heart (366 g) was observed. Neither significant stenosis of coronary arteries nor fibrotic lesions were observed in the heart. The lungs weighed 426 g (left) and 698 g (right) and were colored dark red, especially the right lung. Other findings included hemorrhage in the hilum of the right kidney and adhesion of the pleura and the peritoneum.

Microscopic examination indicated hypercellular bone marrow with monotonous cell proliferation (Fig. 3a). The proliferating cells had convoluted nuclei and marked nucleoli. Infiltration of those cells was also observed in various sites, including the spleen, the sinusoidal space of the liver, the heart (Fig. 3b), and the interstitial space of the kidney and the pituitary gland. Massive hemorrhage was observed in a wide range of alveoli in the right lung, as well as occupation of microvessels by proliferating cells. Microthrombi were also observed in the microvessels of the right lung (Fig. 3c). Moreover, proximal tubular necrosis was observed in the kidney on microscopic examination. Immunohistochemical staining of the bone marrow revealed the proliferating cells to be negative for CD3 and CD79α (lymphocyte marker) and strongly positive for myeloperoxidase (Fig. 3d), indicating that the proliferating cells showed myeloid differentiation. Based on the findings of autopsy and immunohistochemical staining, we concluded that the cause of death was multiple organ failure including massive alveolar hemorrhage caused by acute myeloid leukemia.

4. Discussion

The rate of autopsy of elderly persons is generally low in many countries [10–12]. Japan is no exception, where the death investigation system is currently in the developmental stage [13,14]. If external examination indicates the possibility of death due to external causes is low, determination of probable cause of death is performed by review of the patient's medical history. However, elderly persons often have several critical diseases, as had the present case. When autopsy is not performed, physicians tend to report circulatory system disease as the cause of sudden death [14–16]. Accordingly, there is a risk that chronic diseases other than circulatory diseases may be underreported as causes of death in cases of sudden natural death.

Review of the PMCT images in this case clearly showed massive splenomegaly and consolidation in the right lung, which may be