Case report

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Fatal complication of sickle cell anemia in an immigrant patient rescued from the mediterranean sea

A. Alongi[1], E. Ventura Spagnolo[1], S. Zerbo[1], G. L. Aronica[1], F. Korte[2], E. Maresi[1], A. Argo[1]

[1]*Department Pro.Mi.Se., University of Palermo;* [2]*Italy-U.S. Fullbright Commission*

Abstract

The aim of this case report is to share with the forensic science community the experience of a rare complication of sickle cell anemia: acute chest syndrome.

In October 2016, at the port of the city of Trapani (Sicily, Italy), the landing of an ONG "Médecins Sans Frontières" ship took place with 548 non-EU citizens and a corpse on board. The man, in the hours before his death, complained of severe chest pain and respiratory difficulties, and, despite of the therapeutic aids and resuscitation maneuvers, lastly died.

The Public Prosecutor ordered that autopsy be executed on the corpse of the young Ghanaian, and it was to be supplemented by histological, toxicological, genetic investigations, and the dating of the biological age.

The autoptic examination findings were indicative of bilateral bronchopneumonia, and were also confirmed by the histological findings, which identified a condition of massive adipose pulmonary embolism, due to the presence of extramedullary hematopoietic tissue, site of endovascular thrombosis secondary to hemolysis of sickle cells. The spleen appeared of small volume, with fibrotic phenomena.

The predisposition to infections, thrombosis, extramedullary hematopoiesis are all complications of sickle cell anemia. The severe pulmonary condition characterized by vaso-occlusive findings and pulmonary inflammation with infiltrate, symptoms like fever, chest pain and severe systemic hypoxia allowed to ascribe the plausible cause of death as acute chest syndrome, a rare complication of sickle cell anemia.

The peculiarity of this clinical case is also related to the methods of medical intervention (boat with non-governmental medical support for assistance to migrants). ***Clin Ter 2020; 171 (4):e291-294. doi: 10.7417/CT.2020.2230***

Key words: acute chest syndrome, sickle cell anemia, migrant, death, rescue

Introduction

Due to the increased phenomenon of illegal immigration across the Mediterranean Sea in recent decades, Sicily has assumed the public role as the Frontier of Europe. Illegal journeys, organized by ships which come from North Africa to Italy, sow thousands of deaths every year. Rescue boats coordinated by non-governmental organizations have always worked hard to provide relief and assistance to landings. The health conditions of survivors, upon arrival at the Italian coast, are often severe, mostly due to malnutrition and infectious diseases. The difficulties of communication and the lack of knowledge of their own health conditions make it very difficult for the on-board doctors involved during the first assistance operations to obtain a medical history aimed to promptly determine the correct diagnostic-therapeutic procedure.

The Case

In October 2016, at the port of the city of Trapani (Sicily, Italy), the landing of an ONG "Médecins Sans Frontières" ship took place with 548 non-EU citizens and a corpse on board. From the statements made by the doctors on board it was gathered that the man, in the hours before his death, complained of strong chest pain and difficulty in breathing. He also claimed to have suffered blood transfusions in the past. When his respiratory function worsened and it was detected that his arterial oxygen saturation had decreased, the health professionals practiced oxygen therapy obtaining an improvement in the clinical conditions. During the morning of the following day, the respiratory difficulties and the lowering of the arterial oxygen saturation were accentuated again. After a few hours the man suffered a cardiac arrest and, after carrying out the resuscitation maneuvers, his death was recorded. The Public Prosecutor ordered that autopsy be executed on the corpse of the young Ghanaian, and it was to be supplemented by toxicological, genetic investigations, and the dating of the biological age. Subsequently the histological preparations were analyzed under the pathological-forensic profile.

Correspondence: A. Alongi, e-mail: albertoalongi@gmail.com; E. Ventura Spagnolo: e-mail: elvira.ventura@unipa.it

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Results

Upon the external examination carried out during the autopsy, multiple oval-shaped, slightly depressed scars on the lower limbs were discovered (Fig. 1).

At the superficial level, no parts of the body, including the head, showed signs of specific traumatic lesions, not even the palpatory signs of bone lesions.

At the cadaveric section, the head examination did not show osteo-meningo-encephalic lesions, with parenchymal congestion. The chest examination showed a heart of regular shape and volume in the absence of bone lesions.

The examination of the respiratory system allowed for the detection of important elements for the identification of the cause of death. The lungs were of regular shape and size, and were soft, congested, pale. Macroscopically there were modest traces of anthracosis and widespread marbling of the parenchyma; at the cut, the spontaneous outflow and in abundant quantity of yellow-brownish mucopus was detected (Fig. 2); similar findings inside the tracheal lumen were detected (Fig. 3); the alternation of emphysematous zones to zones of compaction of the parenchyma was also found.

The examination of the abdominal cavity revealed the absence of lesions on the various viscera and other bone structures, showing only widespread congestion.

The spleen was very small, intact, with a wrinkled capsule, of a greenish color (Fig. 4).

Fig. 1

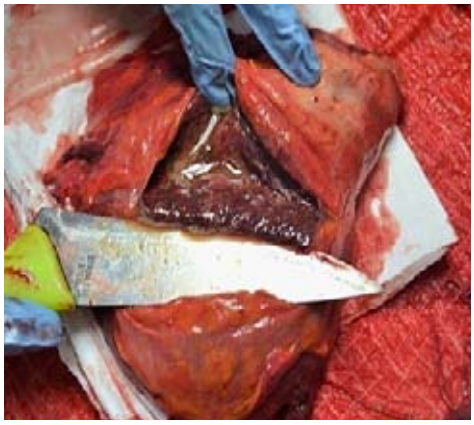

Fig. 2

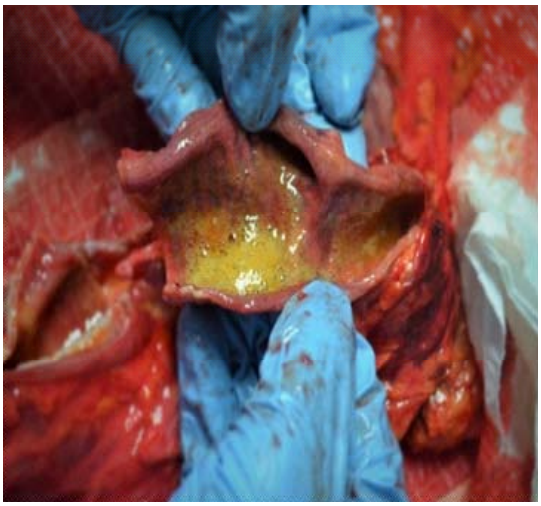

Fig. 3

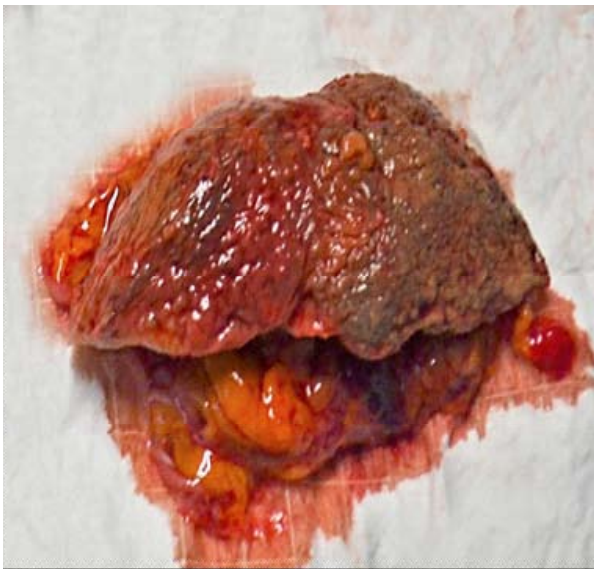

Fig. 4

Histological Findings

Routine hematoxylin-eosin (HE) was performed on all tissue specimens.

The histological examination showed:

- in the spleen, multiple outbreaks of fibrotic organization of haemorrhagic extravasations in the context of which numerous precipitates of free haemosiderin and partly phagocytized by siderocytes were observed;
- in the lungs, multiple fibrinohaematic thromboemboli in various chronohistogenic stages, some of which in organized stage, characterized by arterial vessels with partially recanalized occlusive endoluminal fibrosis. Massive adipose pulmonary embolism associated with septa rupture and desquamative alveolitis with the presence of a large number of frothy histiocytes. Acute plurifocal pulmonary edema and moderate necrotizing bronchopneumonia. In the left lung, hematopoietic medullary tissue with endovascular thrombosis secondary to hemolysis of sickle-shaped red blood cells.

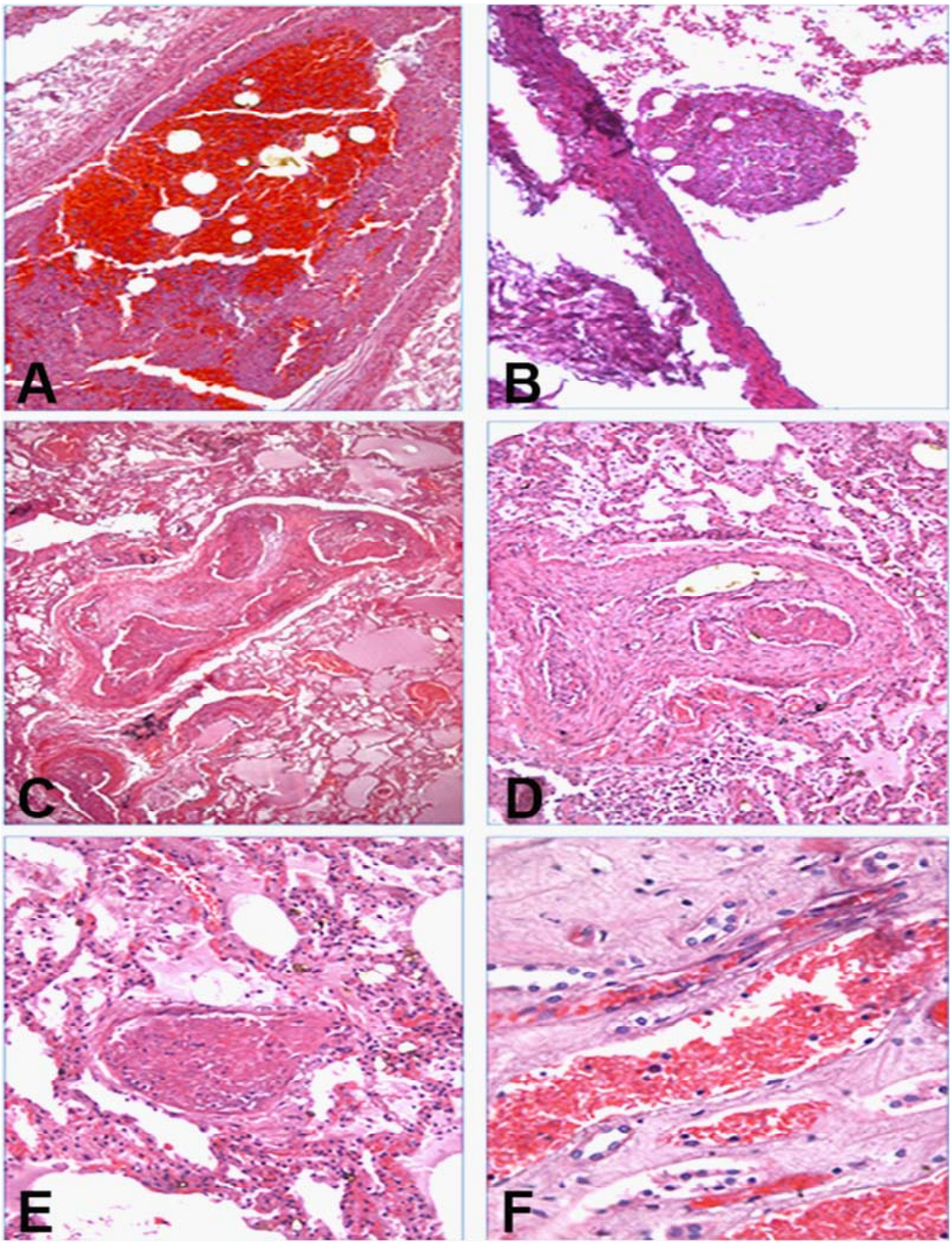

Fig. 5. A-B: Recent thromboemboli of the peripheral pulmonary arteries with droplets of lipid material inside (EE; 10x, 20x); C-D: Recent thromboemboli in peripheral pulmonary arteries with organized and partially recanalized thrombosis (EE; 20x, 40x); E-F: Recent thromboemboli with conglutinate and hemolyzed sickle cells (EE; 20x, 40x).

Discussion & Conclusion

Based on the histological findings it was possible to confirm the diagnosis of sickle cell anemia; the sickle-shaped red blood cells found were eloquent in the context of the vascular district.

It is well known that the pathological course of sickle cell anemia is characterized by several "crises." The vaso-occlusive crises, also called painful crises, represent episodes of damage from hypoxia and infarct that cause severe pain in the affected parts. The most commonly involved sites arein

the bones, lungs, liver, brain, spleen and penis. Acute chest syndrome remains a leading cause of premature mortality in SCD. In a national survey in the United Kingdom, ACS was the third most common cause of death reported in adults (1). It is a recognized risk factor for early death in HbSS patients above the age of 20 years (2).

Acute chest syndrome is a particularly dangerous type of vaso-occlusive crisis involving the lungs and typically occurs with fever, cough, chest pain and pulmonary infiltrate. An infective aetiologyis more common in children than in adults and shows seasonal variation in children, being three times more common in winter (3). Pulmonary inflammation makes blood flow as slow as that of the spleen, leading the sickle-cells to vaso-occlusion. This further compromises lung function, creating a potentially fatal cycle of lung worsening and systemic hypoxia. Predictors of acute respiratory failure include extensive lobar involvement and a history of cardiac disease (4).

The most frequent lung findings included pulmonary edema (47.6%), pulmonary thromboembolism (38.1%), fat emboli (33.3%), pulmonary hypertension, grades I-IV (33.3%), and microvascular occlusive thrombi (28.5%). Studies demonstrate percentages of acute and chronic sickle cell-related lung injury such as fat embolism (33.3%) and pulmonary hypertension (33.3%), with right ventricular hypertrophy (33.3%) (5).

In this case, the finding that appears to be unequivocally responsible for the death of man is identifiable in the pulmonary context. The initial suspicion of bilateral bronchopneumonia, caused by the release of copious amounts of yellow-brownish mucopus in the lungs, was confirmed secondarily by the analysis of histological findings that identified the presence of moderate-degree necrotizing bronchopneumonia with the presence of multiple fibrinohematic thromboemboli in various chrono-histogenic stages and arterial vessels with partially recombined occlusive endoluminal fibrosis.

The diagnosis of massive adipose pulmonary embolism, in the absence of bone fractures, is attributable to the presence of extramedullary hematopoietic tissue on the left lung, the site of endovascular thrombosis secondary to hemolysis of sickle-shaped red blood cells. In the context of the other organs it was also possible to find the presence of additional pathological damage that is typical of sickle cell anemia, and in particular in the spleen, which macroscopically appeared to be small; histologically, the presence of multiple outbreaks of fibrinous organization of haemorrhagic extravasations has been highlighted, in the context of which numerous precipitates of free haemosiderin and partly phagocytized by siderocytes were observed. These phenomena are due to the entrapment of sickle-red blood cells in the cords and splenic sinuses which, leading to a phenomenon of chronic erythrostasis, led to repeated splenic infarcts and, consequently, to fibrosis with progressive reduction of the splenic volume.

Also the presence of diffuse multiple scars in the lower limbs is a phenomenon compatible with the previous creation of ulcers due to vascular stagnation in the subcutaneous tissues.

In conclusion, the severe pulmonary pathological condition is therefore ascribable as a certain cause of death, because the respiratory difficulties, triggering a series of fast-acting processes, led to a sudden arterial desaturation leading inexorably to death.

The predisposition to infections, thrombosis, extramedullary hematopoiesis are all complications of sickle cell anemia. The severe pulmonary condition characterized by vaso-occlusive findings and pulmonary inflammation with infiltrate, symptoms like fever, chest pain and severe systemic hypoxia allowed to ascribe the plausible cause of death as acute chest syndrome, a rare complication of sickle cell anemia.

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