DEATH DUE TO MULTIPLE ORGAN SARCOIDOSIS DISGUISED BY SEVERE ENDOVASCULAR DISEASE

Feodor Odzhakov [1], Todor Todorov [2], Aleksandar Apostolov[1]

[1] Department of Forensic Medicine and Deontology – Medical Faculty – Medical University - Sofia

[2] Department of General and Clinical Pathology – Medical Faculty – Medical University – Sofia

Corresponding author: Assoc. Prof. Aleksandar Apostolov, MD, PhD. Department of Forensic Medicine and Deontology, Medical University, Faculty of Medicine, 2 Zdrave St., Sofia 1431, Bulgaria, e-mails: alexa2000@mail.bg

Abstract

Introduction

Sarcoidosis is a granulomatous disease of unknown etiology, usually with multiple organ involvement. Pulmonary manifestation is the most common, clinically manifested by cough, dyspnea and respiratory failure. In about 30% of cases, the course is chronic. The chronic form causes progressive and life-threatening limitation of lung function. Fatal outcome due to the disease is observed in about 1-5% of patients.

Materials and Methods

The present study presents a case of a 62-year-old woman with undiagnosed sarcoidosis during her lifetime. She has been admitted for hospital treatment due to persistent fever and trophic changes in the limbs, diagnosed as chronic arterial and venous insufficiency 5th degree. After the death in the medical institution, a forensic autopsy of the body has been performed and a histopathological examination of intra-organ parts was performed.

Results

After analysis of the morphological finding during the autopsy, as well as the results of a subsequent histological examination, the cause of death has been established as acute respiratory failure, which has occurred due to advanced pulmonary and multiorgan chronic productive granulomatous process (sarcoidosis).

Discussion

The presented case study focuses on the possibility of chronic progressive multiorgan involvement due to sarcoidosis not to be diagnosed in time due to masking by severe endovascular disease.

Conclusion

The many unknown factors associated with the prevalence of sarcoidosis in the population, together with the ever-increasing number of reports of mortality due to complications of this disease, stress on the need to develop a program for targeted clinical recognition and diagnosis. The diagnostic challenge is even greater when another disease masks the clinical course of sarcoidosis.

Keywords: autoimmune diseases, granulomatous disease, sarcoidosis, forensic medicine, cause of death.

Introduction

The manifestation of sarcoidosis is diverse and virtually every organ in the human body can be affected (1-3). Multiorgan involvement is characteristic, but pulmonary involvement usually dominates (2-6). The skin, eyes and peripheral lymph nodes are affected in about 15-30% of patients (1-3.6). Clinically significant damage to the spleen, liver, heart, central nervous system, bones and kidneys is present in about 2-7% of cases. Asymptomatic involvement of these organs is much more common. A clinical case of multiple organ sarcoidosis is presented in this study.

In about 85-90% of patients with sarcoidosis, changes are found on radiographic examination (5-11). Pulmonary sarcoidosis usually presents with cough, dyspnea, or bronchial hyperactivity — in patients with endobronchial or pulmonary parenchymal involvement (5, 12). In about 30-60% of cases, they are asymptomatic and the disease is diagnosed accidentally by chest radiography on another occasion or by autopsy after death (5, 10, 13, 14). The clinical course is diverse, with spontaneous remission being established in about 2/3 of cases (7-11, 15). Chronic progressive pulmonary sarcoidosis is likely to cause irreversible loss of lung function and disruption of the normal anatomical structure of the parenchyma (5, 16). The deaths in which sarcoidosis is the cause of the fatal outcome vary between 1-5% (7-11, 13, 14, 17-19).

Case presentation:

The authors present a case of a forensic autopsy of a 62-year-old woman who died at the Department of Vascular Surgery and Angiology at a large hospital in Sofia. The body was admitted for autopsy to the Department of Forensic Medicine and Deontology at the University Hospital "Alexandrovska" EAD - Sofia with accompanying medical documentation - an epicrisis from the same hospital regarding the condition of the woman during her stay there. The woman was admitted for hospital treatment due to persistent fever and trophic changes in the limbs, diagnosed as chronic arterial and venous insufficiency Vth degree.

Materials and methods

The autopsy was performed according to the standard sectional technique in cases of disease-related cause of death. Materials from internal organs - lungs, myocardium, liver, spleen and kidneys - were obtained for histological examination. The materials were placed in formalin containers and stored at room temperature for 72 hours. Permanent histological specimens stained according to the standard technique for the hematoxylin-eosin protocol and additional histochemical methods such as Van Gizon, Auramin Rudomin (for tuberculosis bacteria) and Congo red (for amyloid) were developed.

Digital photographs of the established histological findings were taken using an Hp Photosmart E337 camera. Examination of the prepared histological specimens was performed on a NU-2 microscope (CARL ZEISS) using magnifications 10X10 and 10X25.

Results

The epicrisis of the deceased woman reflected the main diagnosis: Ileofemoropopliteal and subclavian phlebothrombosis on the left; chronic venous insufficiency of the lower extremities V degree; chronic arterial insufficiency of the limbs V degree; thrombocytopenia. Concomitant diseases at the time of admission were assessed as follows - arterial hypertension II degree; status post craniotomy frontalis dextra; extirpation tumoris totalis ad oculi (Simpson 1) 2.5 years ago; pyelonephritis; disc herniation. During the operation, psamomic meningioma was proven. The anamnesis indicates the presence of lymph nodes with histologically proven specific granulomatous process, without specifying their location. In the case of a ball, the facial graph of the chest visualizes persistent changes in the upper lung fields, unchanged in comparison with the graph from a year ago.

During the forensic autopsy, the external examination of the deceased woman revealed the presence of numerous bullae filled with lymphoid fluid in the area of the right lower leg and the ankle of the left leg. On the inner surface of the same ankle, there is a round atrophic ulcer with a diameter of 9 cm, uneven edges, sunken and slimy bottom. An ulcer with similar morphological characteristics and a diameter of 7 cm is also present in the area of the right medial malleolus.

An internal trepanation opening of 4 cm / 4 cm was found during the internal inspection of the head. Hydrothorax was measured 400 ml on the right and 300 ml on the left, respectively, and enlarged and compacted lymph nodes were found in the area of the mediastinum and in the area of the pulmonary hilus.

Histological examination of the lungs revealed chronic productive granulomatous inflammatory process, interstitial fibrosis and hyalinosis, as well as the presence of tuberculoid granulomas in the lung tissue (Fig. 1, 2, 3, 4, 5). Tuberculoid granulomas have been found in the spleen.

The cause of death was acute respiratory failure, which occurred on the basis of pronounced pulmonary and multiorgan chronic productive granulomatous process, which developed as a result of sarcoidosis, which took place against the background of Chronic arterial insufficiency of lower extremities (CAILE), Chronic venous insufficiency of lower extremities (CVILE) and ileofemuroplioplitеal and subclavian phlebothrombosis on the left.

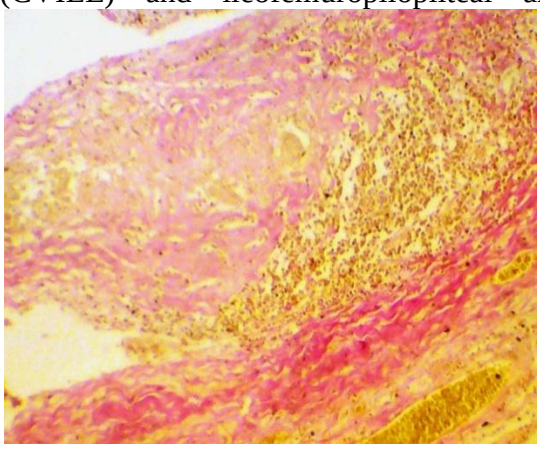

fig.1

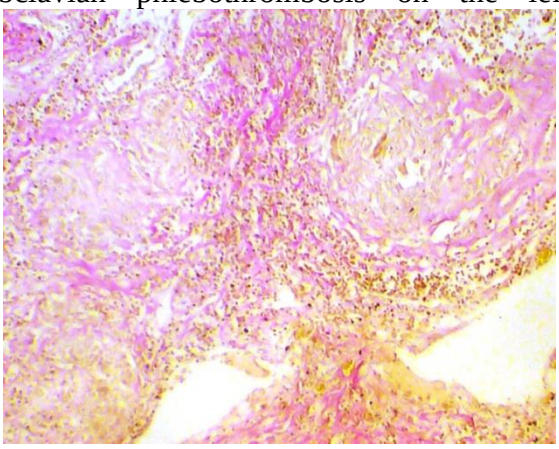

fig.2

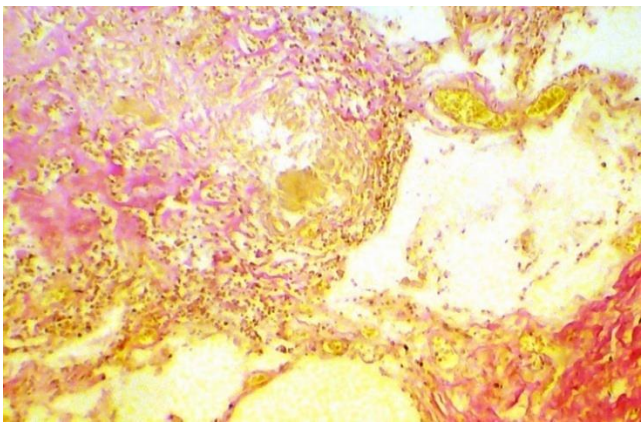

fig.3

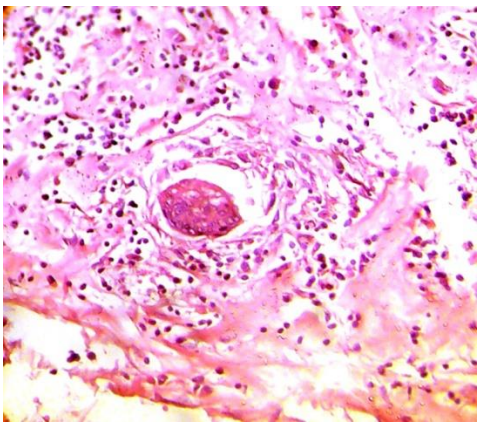

fig.4

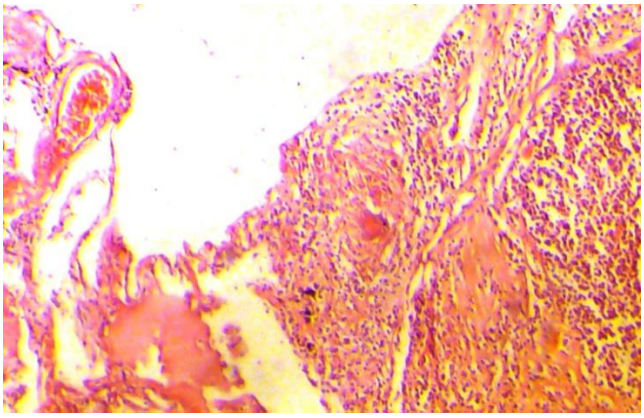

fig.5

FIG. 1- chronic granulomatous inflammatory process with areas of fibrosis in the peribronchial lymph node. Van Gizon stain, magnification 10X10.

FIG. 2 and FIG. 3- chronic granulomatous inflammatory process in the lungs. Hematoxylin-Eosin stain, magnification 10X10.

FIG. 4- chronic granulomatous inflammatory process with the presence of a giant Langhans cell in the lung. Hematoxylin-Eosin stain, magnification 10X25.

FIG. 5- chronic granulomatous inflammatory process with areas of fibrosis in the spleen. Hematoxylin-Eosin stain, magnification 10X10.

Discussion

The actual situation regarding the incidence of sarcoidosis on the territory of the Republic of Bulgaria is unknown. The study presents a clinical case where the disease was diagnosed after a forensic autopsy and subsequent histological examinations of the internal organs were performed, given the established morphological finding. The results of the tests performed in a medical institution during the woman's stay there before her death did not provide a high degree of reliability for such a diagnosis to be made. At the same time, therapeutic diagnostic efforts have focused on treating the patient's severe endovascular disease, which has masked and delayed the diagnosis of sarcoidosis. This circumstance gives grounds for a more detailed study of the standards for prevention of morbidity on the territory of the country. Some of the guidelines in this regard would be the improvement of imaging techniques (e.g. computed tomography), increasing the individual qualification of physicians of all specialties with a targeted search for the disease in clinical data on its manifestation, as well as changing personal hygiene habits of patients (reduction of stress, increase of vitamin D intake, reduction of smoking).

It is reasonable to discuss the issue of the actual number of sick people now. Given the ever-changing environmental factors, as well as the significant improvement in diagnostic methods, it is possible for sarcoidosis to be diagnosed at much earlier stages in order to improve the quality of life and increase its duration. The significance of the problem is also determined by the fact that by the onset of the first clinical symptoms and often asymptomatic course of the disease, multiorgan damage is already in an advanced stage.

Conclusion

The many unknown factors associated with the prevalence of sarcoidosis in the population, together with the ever-increasing number of reports of mortality due to complications of this disease, underline the need to develop a prophylactic program for targeted clinical recognition and diagnosis. The diagnostic challenge is even greater when the clinical

course of sarcoidosis is masked by another overt disease. It is reasonable to discuss the issue of the actual number of sick people at present. Given the ever-changing environmental factors, as well as the significant improvement in diagnostic methods, it is possible for sarcoidosis to be diagnosed at much earlier stages in order to improve the quality of life and increase its duration. The significance of the problem is also determined by the fact that by the onset of the first clinical symptoms and often asymptomatic course of the disease, multiorgan damage is already in an advanced stage.

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