Kahler's disease revealed by acute pancreatitis: A case report

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Abstract

Background: The circumstances of the discovery of Kahler's disease are multiple but acute pancreatitis has rarely been described as a pathology revealing this disease. **Case report:** We report the case of a patient who presented with acute pancreatitis, revealing Kahler's disease. **Conclusion:** Kahler's disease is rarely manifested as acute pancreatitis, hence the importance of complementary examinations in this case, especially when no one of its common etiological factors is found and when other nonspecific signs are present.

Key words: Acute Pancreatitis; Kahler's Disease; Hypercalcemia

INTRODUCTION

Kahler's disease or multiple myeloma is a malignant medullary proliferation of plasma cells secreting a monoclonal immunoglobulin. This is the second most common hematological disorder (10%) after non-Hodgkin's lymphoma.

The circumstances of the discovery of Kahler's disease are multiple. Rheumatologic manifestations are the most frequent due to tumor infiltration of bone tissue. Nevertheless, pancreatitis has rarely been described as a pathology revealing this disease.

We report the case of a patient who presented with acute pancreatitis, revealing Kahler's disease.

CASE REPORT

A 58-year-old patient without a past medical history was admitted at the emergency department for an altered conscious state which appeared a week previously and which was accompanied by cough and vomiting. At the examination, the respiratory rate was 24 cpm, and the peripheral oxygen saturation at the ambient air was 88%. The pulmonary auscultation objective crackles on the right pulmonary base. The blood pressure was 100/60 mmHg, the heart rate was 100 bpm. The Glasgow scale was 14 with agitation. The abdomen examination revealed a tender but soft abdomen in all areas. The capillary blood glucose was amounted to 1.19 g/L. The patient was febrile with a temperature amounted to 38.2°C. The chest X-ray showed a right basal alveolar syndrome (Figure 1).

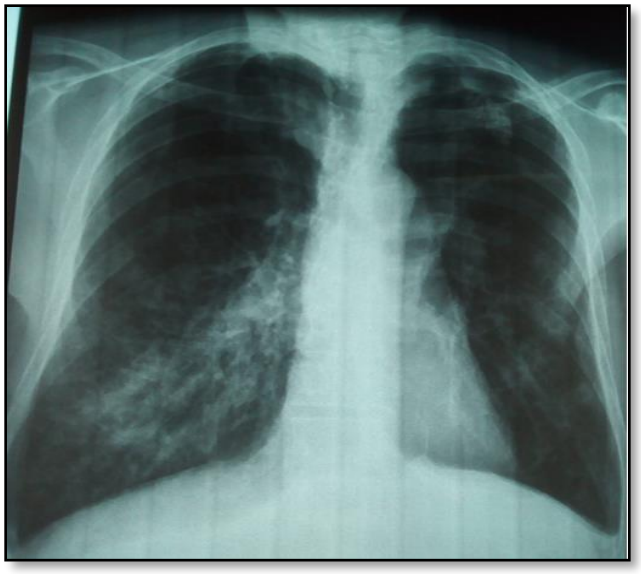

Figure 1: The chest X-ray showing the right basal pneumonia

The cerebral CT scan showed a heterogeneous aspect of the bone structure with no other abnormalities. The Blood tests showed: Leucocytes=16930/mm3, Hb=8.1 g/dL, Platelets=389000/mm3, Creatinine=658 µmol/L, Direct Bilirubin=7.9 umol/L, AST=61 UI/L, ALT=27 UI/L, Lipase level >1200 UI/L.

The abdominal ultrasound examination showed a hyperechogenic appearance of the renal cortex. The diagnosis of pulmonary sepsis associated with acute pancreatitis has been made and adequate treatment was performed. Twenty-four hours later, the patient had a persistent altered conscious state and renal function. A second cerebral CT scan was then achieved and showed the presence of limited gaps of non-specific appearance in the bone structure of the head. The serum calcium concentration was then measured and its level was 4.07 mmol/L.

The diagnosis of acute pancreatitis due to hypercalcemia was then established. A crane radio was performed. It showed multiple geodes combined with demineralization suggestive of Kahler's disease (Figure 2).

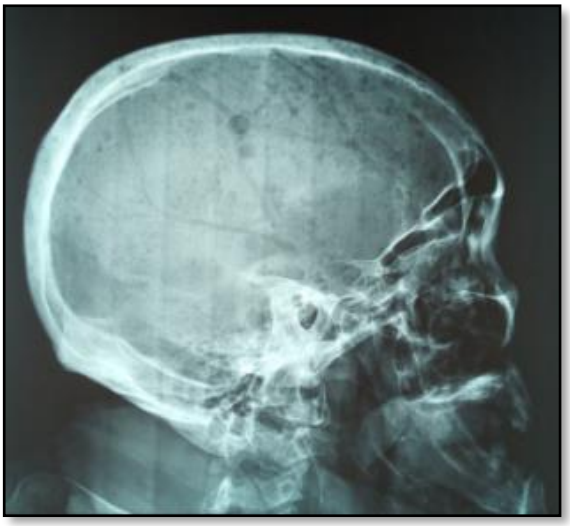

Figure 2: The crane radio showing the holes consecutive to bone destruction

The medical team decided to expand the patient's water intake to 6L/24h and to administer corticoids and diuretics.

The consciousness state of the patient has normalized in parallel with the calcium blood level normalization. The patient was then addressed to the hematology consultation for further explorations to confirm the diagnosis

Thereafter, a serum protein electrophoresis was performed and revealed a marked gamma globulin spike (Figure 3).

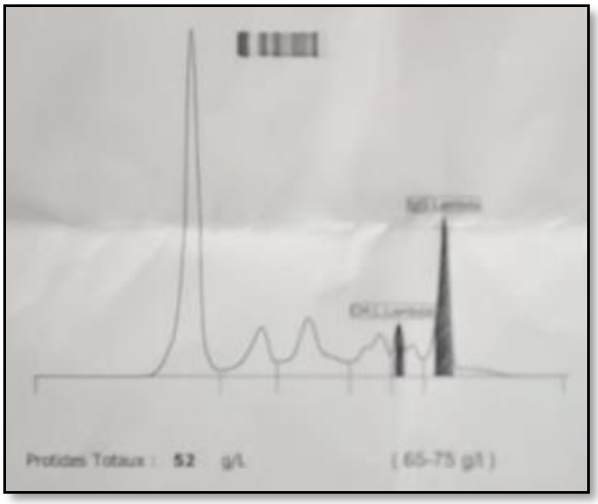

Figure 3: The serum protein electrophoresis revealing a marked gamma globulin spike

Immunofixation of serum proteins was also achieved showing a peak of Lambda light chains associated with an IgG Lambda monoclonal peak.

The immunofixation of urinary proteins revealed proteinuria with the presence of a peak of Lambda light chains.

The patient was hospitalized in the hematology department and he had a good outcome.

DISCUSSION

The common etiological factors of acute pancreatitis are alcohol, gallstone disease, drugs, trauma, viral infection, and hyperlipidemia, but those etiologies are not always identified, and acute pancreatitis is often classified as idiopathic.

Thus, hypercalcemia must be systematically sought in the presence of acute pancreatitis, when there is no obvious cause.

The presentation of multiple myeloma as acute pancreatitis is rarely reported in the literature [1-2].

Multiple myeloma is a complex disorder that causes a multitude of clinical symptoms and signs mediated through a variety of mechanisms.

It is a cancer of the plasma cells in which abnormal plasma cells multiply uncontrollably in the bone marrow, and sometimes in other parts of the body.

It usually evolves from an asymptomatic premalignant stage of clonal plasma cell proliferation termed “monoclonal gammopathy of undetermined significance” (MGUS) [3].

MGUS is present in more than 3% of the population above the age of 50 years and progresses to myeloma or related malignancy at a rate of 1% per year.

The cause of multiple myeloma is unknown. However, there are no known risk factors for multiple myeloma. Researchers suggest that genetic abnormalities, such as c-Myc genes or environmental exposures, may play a role.

The most frequent symptoms of multiple myeloma are bone pains and weakness [3]. Its manifestations are summarized by the "CRAB symptoms": hypercalcemia, Renal failure, Anemia, and Bone lesions. Hypercalcemia is related to osteolysis phenomena [4].

Increased infection susceptibility is also one of the features of this disease [5].

The multiple myeloma is diagnosed with serum or urine protein electrophoresis or immunofixation and bone marrow aspirate analysis.

Skeletal radiographs are important in staging multiple myeloma and revealing lytic lesions, vertebral compression fractures, and osteoporosis.

Magnetic resonance imaging and positron emission tomography or computed tomography were emerging as useful tools in the evaluation of patients with myeloma [6].

Multiple myeloma is considered treatable, but generally incurable. Remissions may be brought about with steroids, chemotherapy, targeted therapy, and transplants. Bisphosphonates and radiation therapy are sometimes used to reduce pain from bone lesions [6-7].

Five-year survival rates approach 33 percent, and the median survival rate is 33 months [6].

In the case of our patient, the diagnosis of Kahler's disease was suspected based on biological and radiologic findings. Once the diagnosis was made, we were able to link each of the unexplained patient's disorders to one of the mechanisms of this disease, such as his confusion, his pneumonia, his renal failure, his anemia, and especially his acute pancreatitis. Hence the originality of our case where the initial clinical presentation of Kahler's disease was atypical and included all of the signs which was rarely described in the literature.

CONCLUSION

Patients with Kahler's disease have hypercalcemia, but it's rarely manifested as acute pancreatitis. Hence the importance of complementary examinations in case of acute pancreatitis, especially when no one of its common etiological factors is found and when other nonspecific signs are present.

This case shows the role of the emergency physician in the early recognition and management of malignant hypercalcemia signs. Its etiologies are multiple; Khaler's disease is one of them.

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