

Which is your diagnosis? • Qual o seu diagnóstico?

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A female, 34-year-old patient followed-up in the neurological care unit for a clinical picture of distally initiated peripheral sensorimotor polyneuropathy. The patient attended the emergency department with sudden dyspnea, and was referred to the unit of imaging diagnosis with clinical suspicion of pulmonary thromboembolism. The occurrence of other previous significant medical conditions was denied by the patient.



Figure 1. Chest computed tomography: axial image acquired after intravenous iodinated contrast injection (arterial phase).

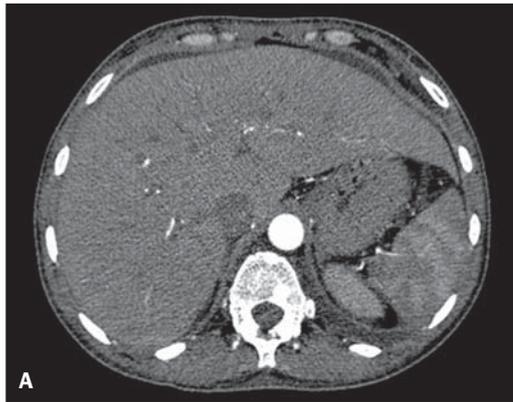


Figure 2. Upper abdominal computed tomography: axial images acquired after intravenous iodinated contrast injection (arterial phase).

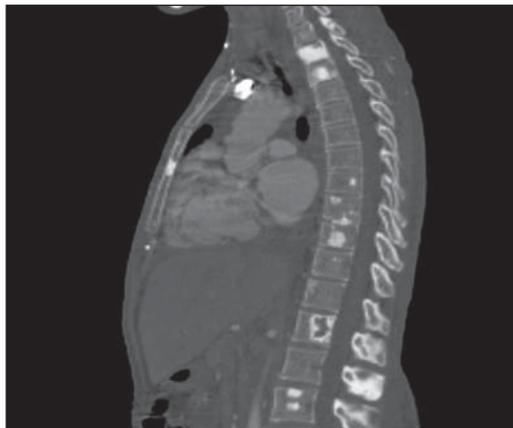
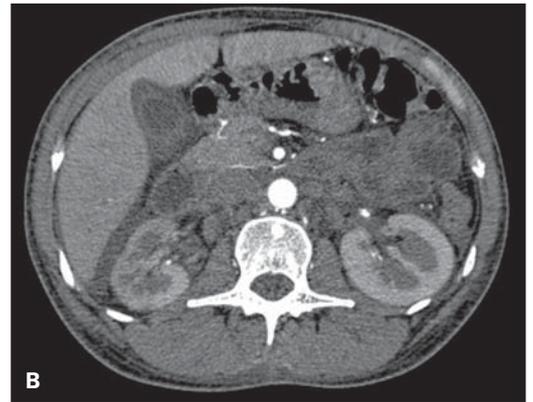


Figure 3. Thoracic spine computed tomography: sagittal reformatting (bone window).

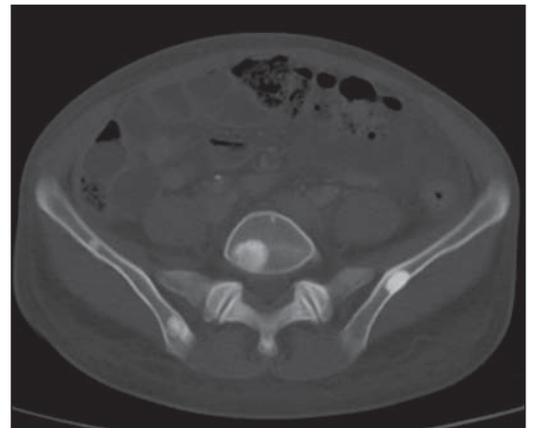


Figure 4. Axial computed tomography image of the hip (bone window).

Images description

Figure 1. Cardiomegaly and moderate bilateral pleural effusion are observed. There is no evidence of pulmonary thromboembolism.

Figure 2. Presence of homogeneous hepatomegaly, free perihepatic and perivesicular fluid (ascites) and retroperitoneal lymphadenomegaly can be observed.

Figure 3. Multiple focal sclerotic lesions sparsely scattered over the thoracic vertebral bodies, posterior elements and sternum. Also, a lytic lesion with irregular sclerotic margins on T10.

Figure 4. Multiple focal sclerotic bone lesions on the iliac bone and lumbar vertebral body.

Diagnosis: POEMS syndrome.

COMMENTS

POEMS syndrome is a rare disorder that was firstly described by Bardwick et al.⁽¹⁾ in 1980, and is characterized by a predominantly sensorimotor polyneuropathy, organomegaly (hepatomegaly, lymphadenomegaly), endocrinopathy (particularly hypothyroidism), monoclonal gammopathy and skin abnormalities.

The diagnosis is based on clinical findings, and the knowledge on radiological alterations allows the radiologist to suggest this diagnostic hypothesis.

The patient presented monoclonal

gammopathy and subclinical hypothyroidism compatible with the diagnosis of POEMS syndrome.

POEMS syndrome is a rare disorder preferentially affecting men in the fifth or sixth decade of life. It is characterized by a range of clinical alterations and the pathophysiology is not well understood. Patients with POEMS syndrome essentially present plasma cell dyscrasia which may range from monoclonal gammopathy up to bones involvement (sclerotic myeloma)⁽²⁾.

All the patients affected by this syndrome present peripheral neuropathy and monoclonal gammopathy considered by some authors as the two major criteria required for diagnosis of the disease. Minor criteria are the following: presence of sclerotic bone lesions, organomegaly (hepatomegaly, splenomegaly, lymphadenomegaly), edemas (pleural effusions, ascites), endocrinopathy (involving adrenal glands, thyroid, parathyroid, gonads, hypophysis and pancreas) and skin changes (hyperpigmentation, hypertrichosis, hemangiomas)⁽³⁾.

The main radiological findings in POEMS syndrome are the presence of organomegaly (particularly hepatosplenomegaly and lymphadenomegaly) and bone lesions. The lesions are mainly sclerotic and, less frequently, mixed. Lytic lesions are rarely found. Preferentially, the axial skeleton is affected, with frequent

presence of lesions on elements of the posterior vertebral arch, which increases the difficulty in the differential diagnosis with secondary lesions. Additionally, abnormal bone proliferation may be observed, with lesions on intervertebral discs and costovertebral joints. In these cases, the sudden appearance of calcifications with irregular, spiculate borders is quite suggestive of this disease⁽⁴⁾.

Generally, this syndrome progresses as a chronic disease, and the prognosis is directly related to the presence of bone marrow involvement and to the number and distribution of the bone lesions, hence the significant role played by the radiologist in the establishment of the prognosis for these patients⁽³⁾.

REFERENCES

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