Diffuse plasmacytoma of the pancreas: a rare entity

Dear Editor,

A 50-year-old male patient, diagnosed with multiple myeloma 10 months prior and undergoing chemotherapy, presented to the emergency department with abdominal pain. Laboratory tests revealed slightly elevated pancreatic enzymes. Subsequently, contrast-enhanced computed tomography (CT) of the abdomen showed diffuse, marked enlargement of the pancreatic parenchyma, with homogeneous uptake of the iodinated contrast medium in the portal phase (Figure 1). The initial working diagnosis was acute pancreatitis. However, the expected clinical, biochemical, and radiological improvement did not occur. We chose to perform CT-guided biopsy, and the histopathological analysis of the biopsy sample revealed a malignant neoplasm composed of loosely cohesive atypical cells, with hyperchromatic, voluminous, eccentric nuclei, consistent with a diagnosis of plasma cell neoplasm (Figure 2A). A complementary immunohistochemical study revealed expression of CD138, together with monoclonal immunoglobulin deposits of kappa light chain, confirming the diagnosis of pancreatic infiltration by plasmacytoma (Figure 2B).

Multiple myeloma is characterized by proliferation of malignant plasma cells originating from the bone marrow and accounts for 10% of all hematological malignancies. Extramedullary plasmacytoma accounts for 5% of all plasma cell tumors and primarily affects males, the mean age at presentation being approximately 55 years. They can be primary, occurring as solitary masses without bone marrow involvement, or secondary, occurring as part of a multiple myeloma, the latter being the more common presentation[1–4]. The most common site of extramedullary involvement is the upper respiratory tract (80%); however, other sites, such as the gastrointestinal tract, genitourinary tract, reticuloendothelial system, thyroid, lungs, skin, and testicles, can also be involved[4].

There have been few reports of extramedullary plasmacytoma affecting the pancreas. Of the approximately 25 cases described, most have involved a focal mass and only one has involved diffuse infiltration of the pancreas[2–6], ours therefore representing only the second such case reported. The most common site of presentation is the pancreatic head, in most cases resulting in abdominal pain and obstructive jaundice[1–4]. The radiological findings of pancreatic plasmacytoma are not highly specific. In the focal presentation, the solid mass is homogeneous or heterogeneous, multilobulated, with variable enhancement[1]; in the one previously reported case with a diffuse presentation, there was diffuse volumetric enlargement of the pancreas with lobulated contours and predominantly homogenous uptake in the portal phase[6], similar to what was observed in the case reported here.

Although CT is the method of choice for the investigation of pancreatic plasmacytoma, it is not capable of excluding diseases such as adenocarcinoma, lymphoma, and metastasis, histopathology therefore being fundamental for the diagnosis[3]. In the case reported here, given the diffuse presentation, the main diagnostic hypotheses were pancreatitis and lymphoma. Lymphoma was excluded because of the clinical and laboratory findings, which indicated that pancreatitis was the most likely diagnosis. However, based on the history of multiple myeloma and the persistence of symptoms, the possibility of pancreatic infiltration by plasmacytoma was considered. Treatment for extramedullary plasmacytoma involves the combination of local radiation, chemotherapy, and, in selected cases, surgery[4].

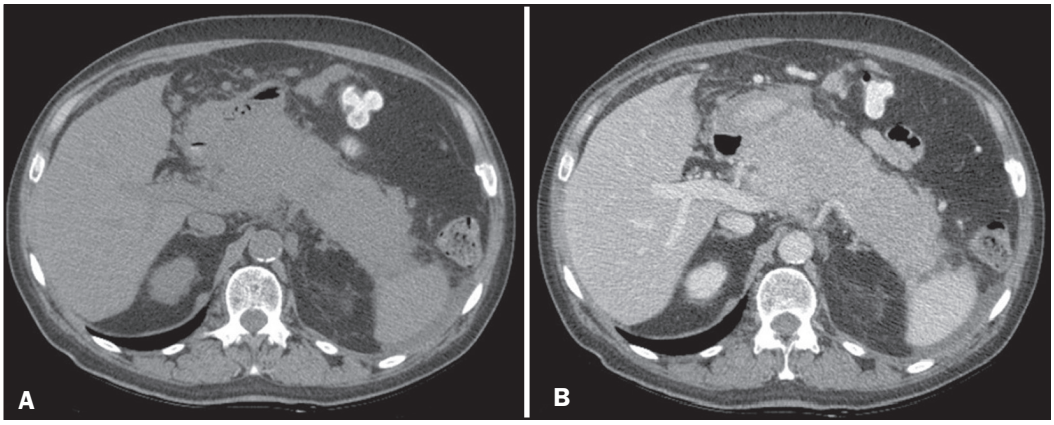

Figure 1. Axial CT scans of the abdomen, without contrast (**A**) and with contrast in the portal phase (**B**), showing diffuse, marked enlargement of the pancreatic parenchyma, with homogeneous uptake of the iodinated contrast medium.

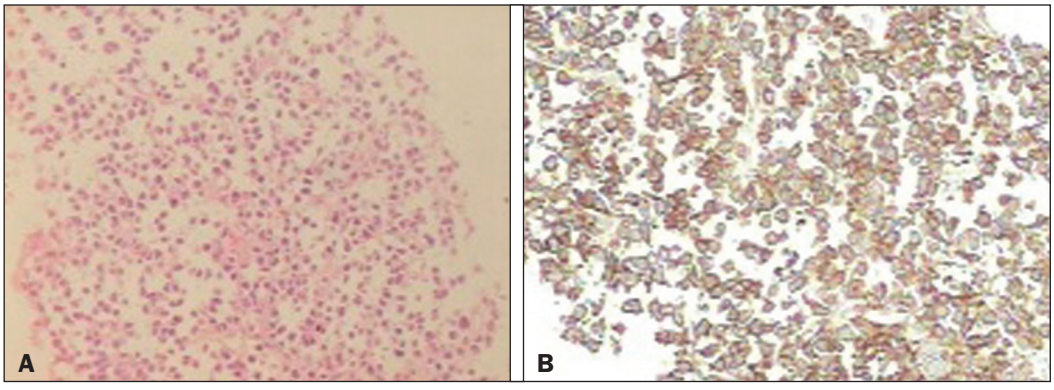

Figure 2. A: Histopathology showing malignant neoplasm composed of loosely cohesive atypical cells, with hyperchromatic, voluminous, eccentric nuclei, consistent with a diagnosis of plasma cell neoplasm. **B:** Immunohistochemistry showing CD138 expression, together with monoclonal immunoglobulin deposits of kappa light chain, confirming the diagnosis of pancreatic infiltration by plasmacytoma.

Plasmacytoma of the pancreas is a rare entity and continues to be the subject of many studies. In patients with multiple myeloma and focal or diffuse enlargement of the pancreas, the hypothesis of plasmacytoma should be considered, thus avoiding delayed diagnosis.

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A rare case of pneumorrhachis accompanying spontaneous pneumomediastinum

Dear Editor,

A 7-year-old female with dyspnea and edema of the neck, accompanied by a cough, was treated at another facility, where anti-inflammatory drugs and an inhaler were prescribed. The patient evolved to worsening of the dyspnea and cough, in addition to intercostal retraction and increased neck volume. She presented to our facility in satisfactory general health. On physical examination, the oropharynx showed no alterations, although there was bilateral edema of the neck and periorbital area, together with diminished breath sounds, sparse wheezing, respiratory rate of 30 breaths/min, intercostal retraction, and subcutaneous crackles on anterior/posterior thoracic palpation, without Hamman's sign. A chest X-ray obtained at admission (Figure 1) showed pneumomediastinum and extensive subcutaneous emphysema. She underwent computed tomography (CT) of the chest (Figure 2), which revealed pneumorrhachis, a rare finding. The patient remained in the hospital for five days under supportive care, and there was complete remission of symptoms.

Spontaneous pneumomediastinum, also known as Hamman's syndrome, is an uncommon condition in medical practice, occurring in approximately 1/30,000 hospital admissions[1] and in only 1% of asthma cases[2]. Its main causes are intense physical exercise, labor (of childbirth), pulmonary barotrauma, diving to great depths, severe paroxysmal coughing, vomiting, asthma, inhalation of narcotics, bronchial asthma, and a slender body type[1,2].

The pathophysiological hallmark of Hamman's syndrome is alveolar overdistension and rupture, which results from high intra-alveolar pressure, low perivascular pressure, or both. After the initial event, the air freely penetrates the mediastinum during the respiratory cycle, in order to balance the pressure gradients[3,4].

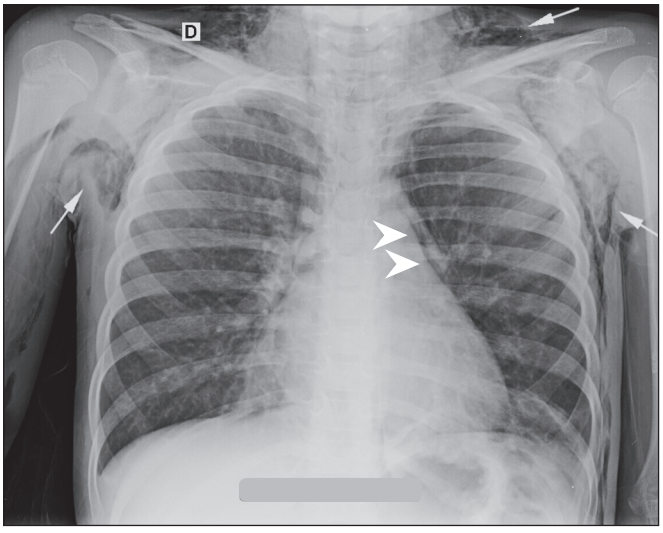

Figure 1. Posteroanterior chest X-ray showing pneumomediastinum (arrowheads), together with extensive subcutaneous emphysema in the supraclavicular and axillary regions (arrows).

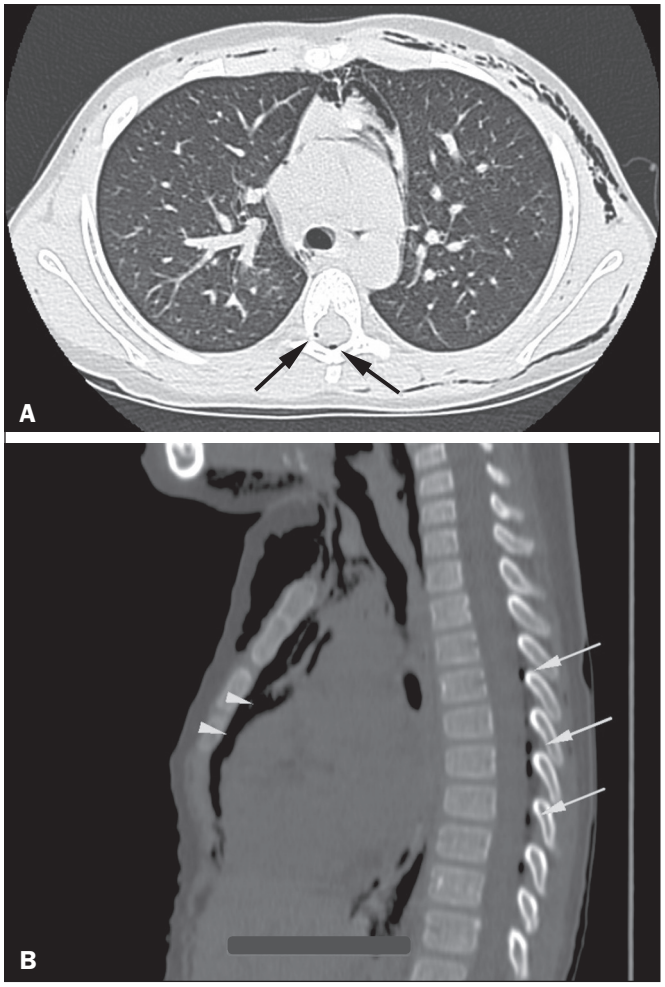

Figure 2. CT of the chest in the axial (**A**) and sagittal (**B**) planes showing pneumorrhachis (arrows) and mediastinal emphysema (arrowheads).