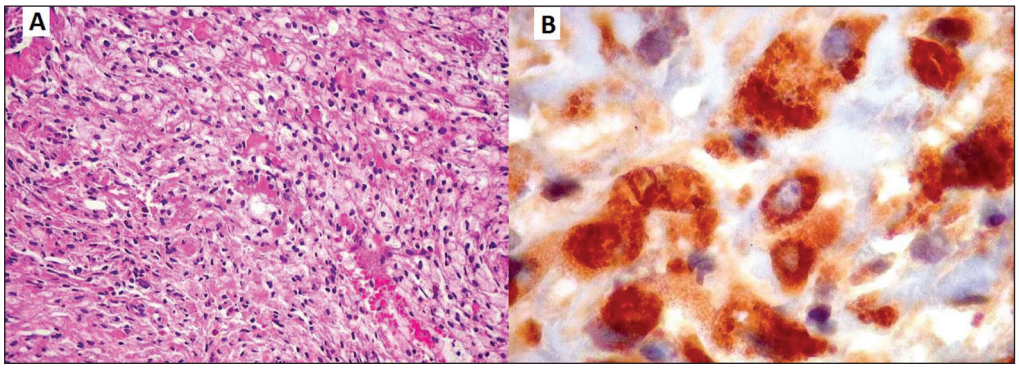

Figure 2. Non-Langerhans cell histiocytosis of the skull base. **A:** Photomicrograph showing abundant xanthomatous macrophages, in a solid arrangement, with small, dense nuclei and clear cytoplasm with lipid droplets. The cytoplasmic boundaries were more or less defined, depending on the area. **B:** Photomicrograph showing positivity for the macrophage marker CD68, which was the main antigen demonstrated in the lesion.

phages with xanthomatous cytoplasm and small nuclei, together with giant cells, as well as few lymphocytes and eosinophils. The histiocytes are positive for CD-68, negative for S-100 protein, and negative for CD1A. It is noteworthy that Langerhans cells are positive for CD1A, negativity for CD1A therefore ruling out a diagnosis of Langerhans cell histiocytosis[3].

Clinically, Erdheim-Chester disease manifests as a systemic disease, involving bone, as well as the central nervous system (CNS), eyes, lungs, mediastinum, kidneys, and retroperitoneum[4]. The most common symptoms are bone pain accompanied by progressive weakness, especially in the lower limbs, together with fever, weight loss, exophthalmos, dyspnea, and signs of neurological impairment such as diabetes insipidus.

A recent extensive systematic review of 331 articles, including a collective total of 448 patients diagnosed with Erdheim-Chester disease, showed that neurological involvement was present as an initial manifestation in 25% of the patients and over the course of the disease in 50%[5]. Exophthalmos, other eye disorders, diabetes insipidus, cerebellar syndromes, seizure, and radiculopathy were the most commonly observed CNS manifestations. The most common features seen on imaging examinations were retro-orbital masses, involvement of the cerebellar dentate nucleus and meningeal lesions of the dura mater, as well as areas of cerebellar and brain stem demyelination. Suprasellar and infundibular lesions were more often accompanied by diabetes insipidus, hypopituitarism, and hyperprolactinemia. Involvement of the spinal cord was less common than was involvement of the brain and brain stem[5].

In the present case, the neurological impairment was isolated. In the literature, we found no other reports of exclusive involvement of the CNS. The gender and age of our patient were also uncommon, given that the prevalence of Erdheim-Chester disease is highest among male patients between the 5th and the 7th decades of life[5].

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Pseudocyst in ectopic pancreas: diagnosis and percutaneous treatment guided by MDCT

Dear Editor,

A 40-year-old man presented with a 12-h history of severe abdominal pain, nausea, and vomiting. Although he reported no comorbidities, he stated that he had concomitant constipation and had consumed alcoholic beverages over the past three days. Physical examination revealed pain on palpation of the lower abdomen. Multidetector computed tomography (MDCT) of the abdomen showed a normal pancreas and tissue formation with a density of 30 HU, similar to that of the pancreatic parenchyma (Figure 1), located in the mesentery, in close contact with the proximal segment of the jejunal loop, measuring 2.8 × 2.9 × 2.9 cm, with adjacent liquid (Figures 1 and 2). The patient was hospitalized, with high levels of amylase and lipase, being treated with nutritional support and antibiotic coverage. His pain worsened, persisting for 12 more days. Another MDCT scan showed the formation of a pseudocapsule, with contrast enhancement and residual adjacent fluid. To look for infection, we opted for percutaneous drainage, smear cytology, and determination of the amylase level in the liquid (Figure 2). Cytometry showed the presence of leukocytes, a differential count with a predominance of mononuclear cells (60% lymphocytes), and the absence of malignancy. The Gram stain was negative, as were tests for fungi, acid-fast bacilli, and other bacteria. The pH was 7.79, the LDH level was 405 IU/mL, and the amylase level was 1207 IU/L. The post-drainage evolution was favorable, and the patient was discharged in good clinical condition. At this writing, he has been in outpatient follow-up for six months, during which time he has been asymptomatic.

Tumors and pseudotumors of the upper abdomen have been the subject of recent studies in the radiology literature of Brazil[1–7]. Ectopic pancreas is a rare condition that is most common in males between the fourth and sixth decades of life. It is defined as pancreatic tissue in an anomalous location, with no anatomical, neural, or vascular connection with the normal pancreas[8]. Although the pathogenesis of ectopic pancreas is unknown, there are two hypotheses: the first suggests that there is transplantation of embryonic pancreatic cells to neighboring structures during the intestinal rotation process; and the second proposes that embryonic buds remain attached to the primitive

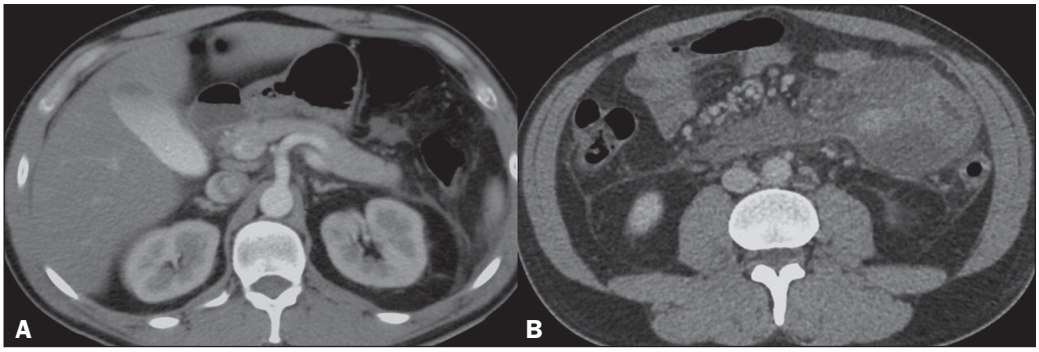

Figure 1. Axial MDCT scan of the entire abdomen, after intravenous administration of an iodinated contrast agent, in the arterial and portal phases (**A** and **B**, respectively). Note the pancreas in its usual location, without signs of inflammation (in **A**). In **B**, note the ectopic pancreatic tissue, located in the mesentery, in close contact with the proximal segment of the jejunal loop, with adjacent fluid.

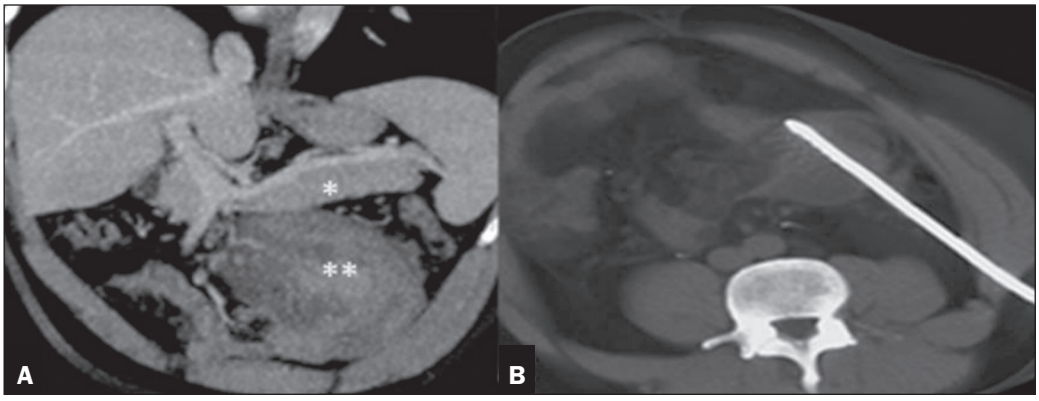

Figure 2. Oblique coronal MDCT scan of the entire abdomen, in the portal phase (**A**), in which the pancreas can be seen in its usual location (asterisk), the ectopic pancreas showing similar density and signs of inflammation (double asterisk). In **B**, puncture and MDCT-guided percutaneous drainage.

duodenum and, during the growth and formation of the gastrointestinal tract, could be carried to sites proximal or distal to the primitive duodenum[9].

The majority of patients with ectopic pancreas are asymptomatic, and the diagnosis is generally made on the basis of an incidental finding, either during an imaging examination or during exploratory surgery motivated by other diseases[10,11]. It is important to note that ectopic pancreatic tissue is susceptible to all of the same diseases that effect the native pancreas[12].

The treatment of ectopic pancreas is directly related to the symptoms and degree of malignancy. Resection is recommended for symptomatic patients with lesions greater than 3.0 cm and possible malignancy. However, when the lesion is smaller than 3.0 cm or is an incidentaloma, there is still no consensus regarding the choice between resection and conservative percutaneous treatment with periodic surveillance. Resection of an ectopic pancreas can be performed endoscopically or surgically[13].

In conclusion, we believe that, although rare, a pseudocyst[14] in ectopic pancreatic tissue should be addressed in order to avoid neoplasia and infection, facilitating clinical treatment and periodic surveillance of the patient, in which the radiologist plays an essential role.

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