Neuroendocrine tumor of the pancreas with cystic appearance mimicking a progressive intraductal papillary mucinous neoplasm: pitfall in medical imaging

Pancreatic cystic lesions are increasingly being recognized on imaging studies. Among these, intraductal papillary mucinous neoplasms (IPMNs) frequently exhibit a spectrum of neoplastic transformation and follow-up is recommended according to the Sendai [1] and Fukuoka criteria [2], with the preference being for magnetic resonance imaging (MRI) [3]. Endoscopic ultrasound (EUS) can help to verify the absence of thickened walls or mural nodules [2].

We report on a 75-year-old woman who was referred to our emergency department with nausea and chest pain. Her physical examination was unremarkable and her laboratory parameters were within the reference ranges, except for a slightly elevated lipase. Computed tomography (CT) of the abdomen revealed a cystic lesion between the body and tail of the pancreas (Fig. 1). On EUS, a connection between the cystic lesion and the main pancreatic duct (MPD) was seen (Fig. 2) and therefore a branch duct IPMN was diagnosed. MRI showed a 1.8 × 1.5-cm cyst with a connection to the MPD (Fig. 3 and Fig. 4).

After 2 months the patient was re-examined with a contrast-enhanced EUS. The cyst showed an increased size with a persistent contrast-enhancing thickened wall (Fig. 5; Video 1). Because of these “worrisome features” of the lesion and its rapid growth (now 24 mm) a 19-gauge needle biopsy was performed. The fluid showed a carcinoembryonic antigen (CEA) level of 212.3 U/mL and a positive string sign [4], suggestive of mucinous cyst content [5]. A lipase value of 114812 U/mL proved that there was involvement of the pancreatic duct.

Histology unexpectedly revealed cells of a well-differentiated neuroendocrine tumor (Fig. 6) with no evidence of a mucinous epithelium. Immunohistochemistry showed positivity for synaptophysin and somatostatin receptor. The patient was referred for a surgical resection.

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References