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A Rare Case of Intraductal Papillary Mucinous Carcinoma of the Pancreas

Marielda D'Ambra[1], Carlo Alberto Pacilio[1], Riccardo Panzacchi[3], Giovanni Taffurelli[1], Salvatore Buscemi[1], Claudio Ricci[1], Maurizio Cervellera[1], Valeria Tonini[1], Donatella Santini[3], Raffaele Pezzilli[2], Riccardo Casadei[1], Francesco Minni[1]

Departments of [1]Surgery, [2]Digestive Diseases and Internal Medicine, and [3]Pathology, S.Orsola-Malpighi Hospital. Bologna, Italy

Context Intraductal papillary mucinous neoplasms (IPMN) of the pancreas represent an evolving pancreatic disease. **Case report** A 68-year-old asymptomatic woman, mild smoker, with clinical history of gallstones, arterial hypertension and surgical treatment for breast carcinoma, was admitted to our surgical unit in January 2012. An ultrasound examination, performed as a follow-up for gallstones, showed a cystic lesion of the pancreatic head. Laboratory tests revealed an increase of pancreatic amylases (114 U/L; reference range: 8-52 U/L) and lipases (89 U/L; reference range: 8-78 U/L), while tumor markers were within reference range. A contrast-enhanced CT scan showed the presence of a 22 mm diameter, multilocular, cystic lesion of the pancreatic head, with contrast-enhanced mural nodules. An endoscopic ultrasonography confirmed the lesion, the mural nodules and showed no dilatation of the main pancreatic duct (MPD); a fine needle aspiration (FNA) revealed an increase of CEA (416.6 ng/mL; reference range: 0-192 ng/mL) in the cystic fluid and cytological analysis showed mild to high grade dysplasia. The CEA levels were suggestive of a mucinous cystic neoplasm, while the presence of mural nodules suggested a malignant type II IPMN. Thus, a pancreaticoduodenectomy was performed. The postoperative course was uneventful and the patient was discharged in postoperative day 18th. The pathological specimen macroscopically showed a solid 14 mm diameter microcystic lesion with endoluminal micropapillae. Microscopically, the lesion was characterized by a carcinoma with tubulo-papillary pattern of growth, atypical cube-shaped cells without cytoplasmatic mucin and intralesional necrotic foci. Moreover, in the pathological specimen, clusters of IPMN gastric-subtype, with low to moderate dysplasia, were detected. These findings suggested the diagnosis of a well differentiated tubulo-papillary carcinoma associated to a mild dysplasia type II IPMN. All lymph nodes were negative. The patient is well and alive at 6 months from surgery. **Conclusion** To our knowledge, in literature only ten cases of ITPN were reported and its peculiar characteristic seems to be a more aggressive natural history.