Pathophysiology, Incidence and Implications in Intraductal Papillary Mucinous Neoplasia of the Pancreas

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Pathophysiology of IPMN

Intraductal papillary mucinous neoplasms (IPMNs) are uncommon tumors arising from the main pancreatic duct. Branch duct and main duct IPMN have distinct features. The overall incidence of invasive carcinoma arising in IPMN is approximately 5–10% in patients with main duct IPMN and approximately 40% in patients with branch duct IPMN (Mohri et al., 2011).

Type 1 IPMNs consist of a cystic dilatation of the main pancreatic duct and additional ductal dilatations, resulting in a so-called “giant cyst” according to Gallucci et al. (2012). The cause of the dilatation of the main pancreatic duct in a Type 1 IPMN can vary from slight expansion to large cystic spaces, and the epithelium of the duct lining can range from columnar to papillary epithelium.

Type 2 IPMNs, also referred to as “mucinous cystic neoplasms,” are characterized by a cystic dilatation of the main pancreatic duct associated with a mural nodule, which is a key feature of this type of neoplasm (Gallucci et al., 2012). The mural nodule is typically smaller than 2 cm and is often found in association with a cystic lesion in the body or tail of the pancreas (Howell & Kent, 2014). Histologically, Type 2 IPMNs are characterized by the presence of mucinous cysts lined by columnar or papillary epithelium, with or without a mural nodule. The cyst fluid is typically mucoid, with occasional proteinacious fluid.

Type 3 IPMNs, also referred to as “intracystic papillary mucinous neoplasms,” consist of a cystic dilatation of the main pancreatic duct associated with a mural nodule, which is considered a “true” neoplasm. These neoplasms are characterized by the presence of a mural nodule, which is usually greater than 2 cm in size, and the cyst fluid is typically mucoid, with occasional proteinacious fluid. Type 3 IPMNs are often associated with a papillary projection into the cyst lumen, which is a key feature of this type of neoplasm (Gallucci et al., 2012).

Pathophysiology of IPMN

IPMNs arise from the pancreatic ductal epithelium and can be associated with adenocarcinoma. The main duct of the pancreas is the site of origin for Type 1 IPMN, whereas the branch ducts are the site of origin for Type 2 IPMN. Type 3 IPMNs are characterized by the presence of a mural nodule, which is considered a “true” neoplasm.

The cystic spaces in the pancreatic ducts appear to be present in the main pancreatic duct.

The cystic space is the differential diagnosis to be considered, illustrating the need for surgical resection of main duct IPMN. In this photograph, you can see the remnants of the mural nodule and the cystic dilatation of the main pancreatic duct.

Possible Molecular Abnormalities in IPMN

Mutations in the Kras gene are frequent in IPMN, and Kras mutations are associated with increased risk of progression to invasive malignancy (Mohri et al., 2011). Kras mutations are also associated with increased risk of developing invasive ductal carcinoma in patients with IPMN (Mohri et al., 2011).

The presence of Kras mutations in IPMN is associated with increased risk of developing invasive ductal carcinoma in patients with IPMN (Mohri et al., 2011). Kras mutations are also associated with increased risk of developing invasive ductal carcinoma in patients with IPMN (Mohri et al., 2011).

References


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Implications for Nursing Care

As advanced practice nurses, it is important to consider incidental imaging findings and their clinical significance. In the case of incidental pancreatic findings, referral to pancreatic surgeons or surgical oncologists is critical, as early detection of premalignant lesions will lead to increased survival outcomes. Awareness of the clinical significance of a lesion is crucial to prevent persistent abnormalities from being followed closely and IPMN is significant because the cellular changes are often asymptomatic and can progress to an invasive malignancy. According to Gallucci et al. (2012), the 5-year survival rate of patients with IPMN is 1-14%.

Signs & Symptoms

Patients with main duct and side branch IPMNs are asymptomatic. Symptoms of IDMCs include jaundice, weight loss, diabetes, and chronic abdominal pain. The symptoms are often vague and nonspecific, and patients with main duct IPMN are at a higher risk for developing gastrointestinal symptoms than patients with branch duct IPMN (Mohri et al., 2011).

Mucin type 1-1 is associated with malignancy. MUC1 and MUC2 are related to mucin type 1-1, and MUC3 is associated with the development of mucinous cystic neoplasms (MCNs). MUC1 and MUC2 are associated with malignancy in patients with IPMN. The pathophysiology of IPMN is significant because the cellular changes are often asymptomatic and can progress to an invasive malignancy. According to Gallucci et al. (2012), the 5-year survival rate of patients with IPMN is 1-14%.

Conclusion

IPMNs are precursor to invasive pancreatic adenocarcinoma. Recognizing the clinical significance is essential in practice and educating patients about their implication is a responsibility of the practitioner. Management of these precancerous lesions varies depending on subtype, patient characteristics, and availability of required resources. The use of a multidisciplinary approach may be the preferred method in managing these patients, particularly those with worrisome symptoms or findings (Reich et al., 2014).

Additional Sources


