CASE SNIPPETS

Psammomatous Carcinoid Tumor of the Duodenum

S A PAI, S KRISHNAMURTHY, C S SOMAN
Tata Memorial Hospital, Parel, Bombay 400 012

Abstract
We report a case of psammomatous calcification in a carcinoid tumor of the ampulla of Vater. Although the tumor was glandular, its neuroendocrine nature was proved using chromogranin immunostain

Key words: Psammoma body, duodenal neoplasia, carcinoid, somatostatinoma.

Psammoma bodies, although not usually seen in carcinoids, are a feature peculiar to duodenal carcinoids. The correct recognition of these rare lesions is of paramount importance for proper patient management. We report a patient with psammomatous calcification in a duodenal carcinoid. To our knowledge, this is the first such Indian report.

A 65-year-old man presented to our institution with complaints of abdominal pain and weight loss. His past history included surgery for peptic ulcer disease. On examination, there were no significant physical findings. Biochemical investigations were within normal limits. Review of ultrasonography and CT scan done elsewhere showed multiple gallstones, cholecystitis and dilatation of both intra-and extrahepatic biliary radicals. Endoscopic retrograde cholangiopancreatography showed a papillary tumor at the ampulla of Vater with evidence of gastrectomy and gastrojejunostomy.

Endoscopic biopsy revealed a tumor composed predominantly of glandular structures, separated by thin septae and lined by columnar to cuboidal epithelium. Nuclei were round, monotonous, with stippled chromatin. Nuclear pleomorphism and mitoses were absent. Psammoma bodies were present throughout the tumor (Fig). The tumor infiltrated the muscularis mucosae. Tumor cells showed chromogranin positivity on immunohistochemical staining. Immunostaining for somatostatin, vasoactive intestinal polypeptide, insulin and glucagon were not done. He was advised surgery but was lost to follow-up.

Although no site in the gastrointestinal tract is exempt, the duodenum, particularly the ampulla of Vater, is an unusual site for the occurrence of carcinoid tumor. Psammoma bodies have been described in meningiomas, papillary carcinomas of the ovary, thyroid, cervical and breast carcinomas and even in normal meninges. The few reported carcinoids containing psammoma bodies have all been in the duodenum.

Immunocytochemical and ultrastructural studies have shown that psammomatous carcinoids of the duodenum contain somatostatin. These do not however cause the somatostatinoma syndrome which is seen in association with pancreatic somatostatinomas. This may be because of the smaller tumor mass or due to low rate of release of biologically active peptides. Psammoma bodies have not been documented in pancreatic somatostatinomas. Their formation is believed to be related to the production of somatostatin, as well as to the actual site of the tumor.

Whipple's resection with or without post-operative chemotherapy has been used to treat patients with psammomatous duodenal carcinoids. Although the clinical outcome of these patients has varied considerably, metastases to the regional lymph nodes and liver have been reported.

Most duodenal carcinoids with psammoma bodies are of the glandular subtype. This may lead to misdiagnosis of a low grade adenocarcinoma or even a nonneoplastic lesion because of bland appearance of the glands. The presence of psammoma bodies should alert the pathologist to the fact that the lesion is a carcinoid. Chromogranin staining confirms the endocrine nature of the tumor.

Fig. Photomicrograph showing psammoma bodies within the tumor (H & E X200).

Correspondence to: Dr Shanta Krishnamurthy, Pathologist
Received May 14, 1993
Received in final revised form August 8, 1993
Accepted August 8, 1993
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