Squamous Cell Carcinoma of the Ampulla of Vater – A Rare Case Successfully Managed By Pancreaticodudenectomy

To the editors

A 30-year-old female patient presented with jaundice, pain in abdomen and loss of appetite. A CT scan showed a circumferential thickening in the periampullary region mildly indenting the second and third part of duodenum. Endoscopic biopsies from the ulcer were suggestive of a squamous cell carcinoma (SCC). A radical pancreaticodudenectomy (Whipple's procedure) was performed. The surgical specimen consisted of part of stomach, duodenum, common bile duct and pancreas. At the ampulla there was a 1 cm diameter chalky white ulcero-infiltrative growth, sections from which showed infiltration of ampullary region by sheets and islands of malignant squamous epithelial cells. Intercellular bridges, cellular keratinization and keratin pearl formation were seen. No evidence of glandular or adenocarcinomatous component was found on whole processing the tumor. Physical examination and a whole body CT scan did not show any other site in the body which might have given rise to a metastatic SCC of the ampulla. Final diagnosis was well-differentiated SCC of the ampulla infiltrating the duodenum.

Majority of the ampullary tumors are adenocarcinomas [1]. Rare cases of adenosquamous carcinoma, carcinoid tumor, primary malignant lymphoma, liomyosarcoma, gastrointestinal stromal tumors, rhabdomyosarcoma, sarcomatoid carcinoma and metastatic SCC of the ampulla have been described [1]. Only three cases of primary SCC of the ampulla have been reported in literature. First was reported in 1952, second in 1960 and third case was reported in 1996. The third case was reported in a 72-year-old Chinese female in whom the diagnosis was made by histologic examination after duodenoscopy. The patient developed biliary tract infection after undergoing ERCP and expired due to persistent bleeding [2]. The patient in our case was successfully managed by pancreaticoduodenectomy.

The histogenesis of ampullary SCC is not known. It has been hypothesized that tumors such as SCC of the extrahepatic bile ducts either arise from squamous metaplasia of columnar lining epithelium secondary to chronic inflammatory process or from squamous metaplasia of the tumor itself. A similar mechanism may also explain the occurrence of SCC at the ampulla. Ampullary carcinomas may extend into duodenum, pancreas and common bile duct and may show perineural and vascular invasion, nodal metastasis or distant metastasis to liver, lung and pleura. Carcinomas of the ampulla need to be differentiated from the carcinomas of distal third of the bile duct. The bile duct carcinomas produce longitudinal thickening of the bile duct and a granular appearance of the mucosa. Majority of these tumors are well differentiated and produce severe desmoplastic reaction, while ampullary carcinomas tend to be poorly differentiated without desmoplasia.. Surgical resection in the form of Whipple's pancreaticodudenectomy remains the treatment of choice in ampullary carcinomas.

To conclude, SCC of the ampulla is a very rare neoplasm and must be kept in the differential diagnosis of ampullary tumors. These tumors can be successfully managed by pancreaticodudenectomy. If a SCC is seen at the ampulla, patient must be investigated to rule out SCC of other sites in the body which may give rise to a metastasis at the ampulla.

References

[1] Talamini MA, Moesinger RC, Pitt HA, Sohn TA, Hruban RH, Lillemoe KD, et al. Adenocarcinoma of the ampulla of Vater. A 28-year experience. Ann Surg 1997;225:590–9; discussion 599–600.

[2] Chen CM, Wu CS, Tasi SL, Hung CF, Chen TC. Squamous cell carcinoma of the ampulla of Vater: a case report. Changgeng Yi Xue Za Zhi 1996;19:253–7.

Deepak Kumar Singh, Archana Rastogi, Ravi Shankar, Ranjana Gondal
Department of Pathology
Govind Ballabh Pant Hospital
New Delhi, India

Pramod K. Mishra
Department of Gastrointestinal Surgery
Govind Ballabh Pant Hospital
New Delhi, India

(Received 18 June 2008; accepted 26 June 2008)
ISSN 1365-182X print/ISSN 1477-2574 online
DOI: 10.1080/13651820802307591