CASE REPORT

Adenosquamous Carcinoma of Ampulla of Vater: A Case Report and Review of Literature

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ABSTRACT

Introduction: Adenosquamous carcinomas are very rare tumors of ampulla of vater with only seven cases reported in literature.
Case Report: We report a case of adenosquamous carcinoma in a 40 year male that was a known case of chronic pancreatitis and presented on follow up with a contrast enhancing 2.3 x 2.1 cm lesion in the ampullary region JMS 2018; 21 (1):44-46
Key words: Adenosquamous carcinoma, Pancreaticoduodenectomy, Ampulla of vater

BACKGROUND

Adenosquamous carcinoma (ASC) is characterized by variable combinations of two malignant components, including adenocarcinoma and squamous cell carcinoma (SCC). Most of the tumors in the ampullary region are adenocarcinomas and very few adenosquamous carcinomas have been reported in the region. Although the proportion of the aforementioned two components varies, adenocarcinoma usually predominates. Such adenocarcinomas with squamoid components are considered to be clinically more aggressive, with a worse prognosis than their adenocarcinoma counterparts[1-7]. Being such a rare tumor there have been few case reports and small case series. Therefore, clinicopathological features and outcomes of this entity remain unclear and no therapeutic strategies have been established. Here, we report one such rare case of ASC the ampulla of Vater and review the reported cases of this unusual entity.

Case Presentation:

A 40-year-old male who was a known case of chronic pancreatitis with multiple acute attacks and on follow up for 3 years developed rapidly progressing jaundice. Liver function tests showed a marked increase in alkaline phosphatase (1600u/l). MRCP revealed T1 isointense/T2 hypointense lesion in the ampullary region causing proximal common bile duct dilatation. Contrast-enhanced CT abdomen revealed a 2.3 x 2.1 cm enhancing lesion in the ampullary region. Patient's CEA levels were normal but CA-19-9 levels were raised (46 IU/ml). A pancreaticoduodenectomy was performed and intraoperatively an ulcerated area was found in the ampulla, the liver was free of tumor and gallbladder was distended. The specimen we received showed an ulcerated growth in the ampulla which was grossly infiltrating into the duodenal wall. Pancreas was hard in consistency. Histopathological examination revealed a dual morphology of squamoid and adenocarcinomatous areas. Squamoid areas showed cells in syncytial nests with foci of keratinization while as adenocarcinomatous foci showed variable sized glands in fibrous stroma Squamoid areas constituted about 50% of the total tumor microscopically. The tumor was infiltrating into the duodenal wall up to muscularis propria. One of the regional nodes showed metastatic deposits of the same tumor. All the resection margins were free of tumor. Adjacent pancreas showed features of chronic pancreatitis with marked fibrosis, loss of acini and lymphoid follicle formation. The case was signed out as adenosquamous carcinoma and staged as T2 N1 M0-Stage IIIA according to the American joint committee on cancer 8th manual.

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Figure 1: Gross photograph showing external surface of a pancreaticoduodenectomy specimen with gall bladder

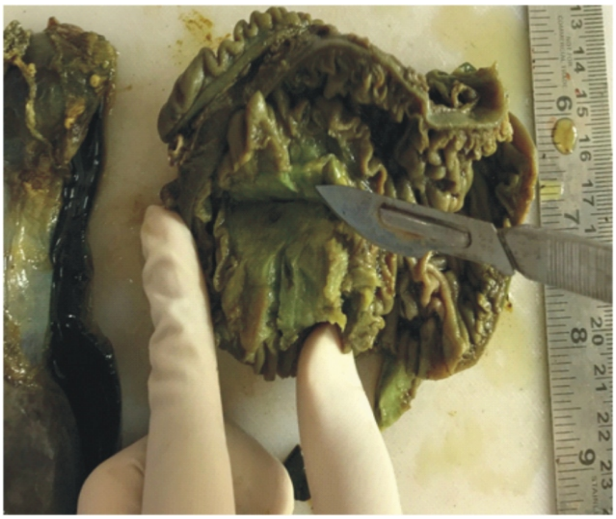


Figure 2: Gross photograph showing a cut section of growth in ampulla

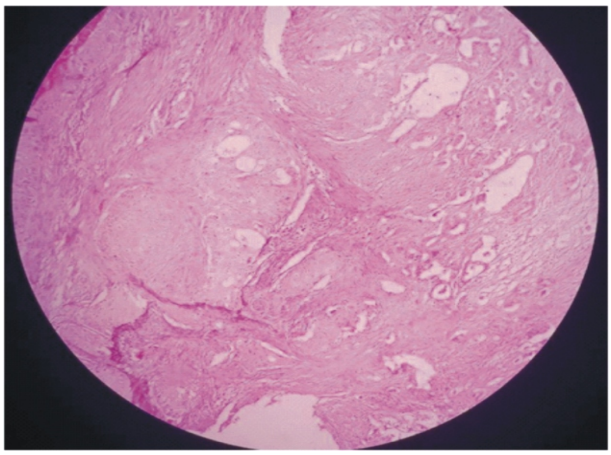


Figure 3: Low magnification photomicrograph showing tumor cells forming squamoid nests and glands (10 x 10 magnification H & E section)

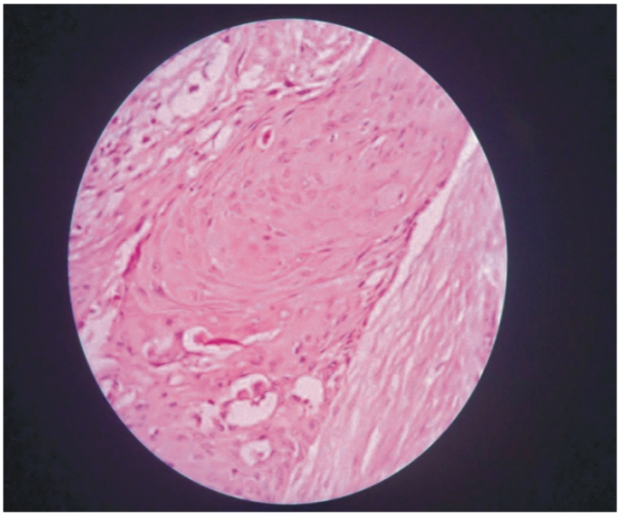


Figure 4: High magnification photomicrograph showing small nests of squamoid morphology with intercellular bridging (40 x 10 magnification H & E section)

DISCUSSION

Three diagnostic possibilities of origin of adenosquamous carcinomas that are considered include:

1. Origin from pluripotent stem cells capable of inducing the malignant transformation of both cell types
2. Squamous metaplasia in the intestinal mucosa, and
3. Collision of the two malignant tumors[8].

It has been suggested that chronic irritation due to long-standing ulcerative colitis or some other reason can cause stimulation of a pluripotential epithelial stem cell that is capable of both malignant transformation and differentiating into the squamous and glandular lines[9], thus providing evidence that tumors often originate from the transformation of normal stem cells[10]. Another hypothesis that is being considered is that adenosquamous carcinomas may occur through a metaplastic malignant squamous transformation of adenocarcinomas[11].

Demographic and clinical characteristics of all the reported cases of such tumors are summarized in Table 1[13]. The average age at diagnosis was 62 years (range, 4782), and five of seven patients (71 %) were men. Most patients presented with symptoms such as abdominal pain and jaundice. Surgical procedures performed were pancreaticoduodenectomy, including pylorus-preserving pancreaticoduodenectomy, in six cases (86 %) and ampullectomy in one case (14 %). The mean tumor size in the four cases with relevant data was 26.8±12.9 mm (range, 11-40mm), and lymph node metastases were identified in three of six cases (50 %). Overall survival in all seven cases

Table 1: Reported cases of adenosquamous carcinoma of the ampulla of Vater in the English literature

Year	Author	Age	Gender	Symptom	Size(mm)	Biopsy	Treatment	LN mets	Stage
2002	Ueno	47	M	Jaundice	22	SCC	PD	+	III
2013	Yang	64	M	Jaundice,Pain abdomen	34	ADC	PD	+	IIB
2013	Yang	82	M	Jaundice	NM	SCC	Ampullectomy	-	IB
2013	Yang	68	M	Jaundice,Pain abdomen	NM	SCC	PD	+	III
2013	Yang	34	F	Jaundice,Pain abdomen	NM	SCC	PD	-	III
2014	Kshirsagar	58	M	Jaundice,Pain abdomen	40	SCC	PD	NM	NM
2015	Hashimoto	81	F	No symptoms	11	ADC+SCC	PPPD	-	IA

SCC squamous cell carcinoma, **ADC** adenocarcinoma, **PD** pancreaticoduodenectomy, **PPPD** pylorus-preserving pancreaticoduodenectomy, **NM** not mentioned **LN** lymph node

ranged from 6 to 20 months. ASC is regarded as a more clinically aggressive tumor with less favorable prognosis than adenocarcinomas [1-7]. Imaoka et al.[12] reported that the median overall survival of patients with ASC of the pancreas was nearly half of that of patients with pancreatic ductal adenocarcinomas.

CONCLUSION

In summary, primary adenosquamous carcinoma of the ampulla of Vater is a very rare histological type of carcinoma. Its clinicopathological characteristics and optimal treatment are still not fully explored. Further documentation of this rare tumor will provide a better understanding of its pathogenesis and management for optimal survival rates.

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