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Primary adenosquamous carcinoma of ampulla of Vater—A rare case report



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ABSTRACT

INTRODUCTION: Primary adenosquamous carcinoma (ASC) of the ampulla of Vater (AmV) is extremely rare. Carcinoma of the ampulla of Vater tends to manifest early due to biliary outflow obstruction, as opposed to pancreatic neoplasms that often are advanced at the time of diagnosis. Periapillary carcinomas are treated by pancreaticoduodenectomy (PD). Adenosquamous carcinoma carries very dismal prognosis.

PRESENTATION OF CASE: Here we present a case of 58-year-old male who was presented with abdominal pain, jaundice and anorexia with no history of (h/o) pruritus and clay colored stool. All blood investigations were normal except liver function tests (LFTs). Ultrasonography (USG) of abdomen suggestive of periampillary mass with dilated pancreatico-biliary tree. Endoscopic retrograde cholangiopancreatography (E.R.C.P.) demonstrated large deformed and bulky papilla with ulcerated lesion with infiltration in to duodenum. Exploratory laprotomy proceeds Whipple's pancreaticoduodenectomy done. Histopathology revealed adenocarcinoma of the ampulla of Vater. Immunohistochemistry was confirmatory of adenosquamous carcinoma.

DISCUSSION: Adenosquamous carcinoma (ASC) is defined as a tumor in which both glandular and squamous elements are histologically malignant. Compared to adenocarcinoma, ASC of the AmV is a rare malignancy. Preoperative diagnosis is difficult because of the lack of defining characteristics in imaging studies and the difficulty in acquiring both malignant components by limited biopsy. Periapillary carcinomas are treated by pancreaticoduodenectomy.

CONCLUSION: Adenosquamous carcinoma is a very rare form of cancer of the AmV. Pancreaticoduodenectomy is the treatment of choice though early recurrence and distal metastasis may be encountered after surgery. Follow-up should be more frequent to detect possible early recurrence and distal metastasis.

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1. Introduction

Adenosquamous carcinoma (ASC) is defined as a tumor in which both glandular and squamous elements are histologically malignant. The mixed tumor was first reported in 1907. It is more common in areas where adenocarcinomas arise frequently. Incidence rate of carcinoma of AmV is 3.8 per 10,00,000 in men and 2.7 per 10,00,000 in women. ASC of the AmV has rarely been discussed in the literature. Here, we report a case of a 58-year-old male with primary ASC of the AmV. Thorough literature study shows there are only five reported cases of ASC of AmV.

2. Presentation of case

A 58-year-old male presented with abdominal pain, jaundice and vomiting for one month. He also had associated history of weight loss and anorexia with no h/o pruritus and clay colored stool. Clinical examination revealed no abnormality except icterus. The laboratory examination revealed a serum total bilirubin level of 6.7 mg/dL, an aspartate aminotransferase (SGOT) level of 102 IU/L, an alanine aminotransferase (SGPT) level of 92 IU/L and an alkaline phosphatase of 250 U/L. USG abdomen was suggestive of periampillary mass with dilated pancreatico-biliary tree. E.R.C.P. demonstrated large deformed and bulky papilla with ulcerated lesion with infiltration in to duodenum causing 1st and 2nd part of duodenal (D1D2) stenosis. Biopsy revealed squamous cell carcinoma. Metastatic work-up did not reveal any other primary focus of disease. Contrast enhanced computed tomography scan (CECT) of the abdomen revealed a soft-tissue mass in the periampillary region with dilated distal portion of the common bile duct (CBD) and main pancreatic duct.

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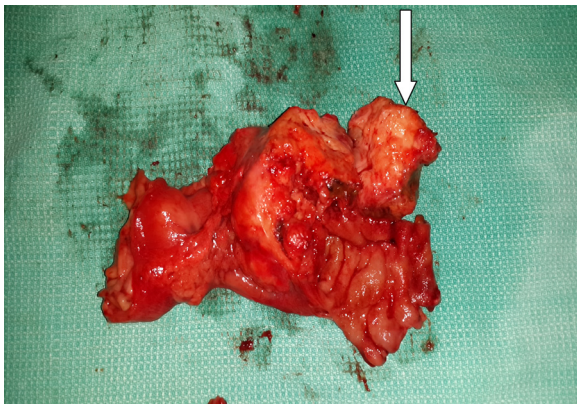


Fig. 1. Photograph showing pancreaticoduodenectomy specimen with mass in the periampullary region indicated by the arrow.

E.R.C.P. guided stent inserted in CBD. Liver function tests were repeated till the total serum bilirubin level normalized. Exploratory laparotomy proceeds Whipple's pancreaticoduodenectomy done.

On exploration there was a growth at the ampullary region with involvement of CBD leading to grossly dilated CBD and main pancreatic duct. The gross specimen revealed a growth of 4×3 cm in the AmV (Fig. 1). Portal vein and superior mesenteric vein and artery were separate from the growth. Histopathological report was of adenosquamous carcinoma of the AmV (Fig. 2). Immunohistochemistry was planned which confirmed adenosquamous carcinoma (Figs. 3 and 4). The postoperative course was uneventful.

3. Discussion

Adenosquamous carcinoma (ASC) is defined as a tumor in which both glandular and squamous elements are histologically malignant. The histogenesis of ASC remains uncertain. Four hypotheses regarding its histogenesis are summarized as follows: (1) pluripotent epithelial stem cells capable of inducing the malignant transformation of both cell types; (2) squamous metaplasia in the intestinal mucosa; (3) adenocarcinoma transforming into squamous cell carcinoma; and (4) collision of both malignant tumors. Although we do not understand the histogenesis of ASC, it exhibits a more aggressive biologic behavior and is associated with a worse

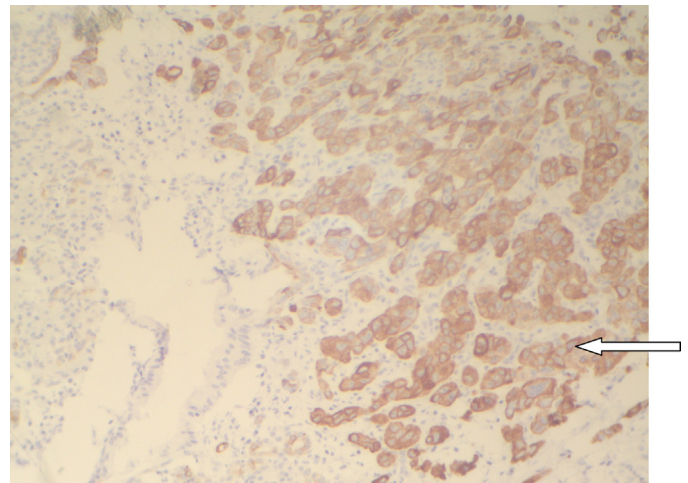


Fig. 3. Microphotograph of immunohistochemistry showing co-expression with p63 in areas of squamous differentiation.

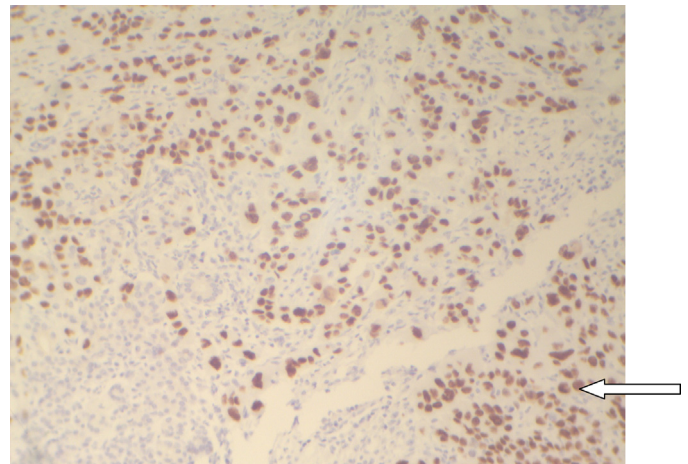


Fig. 4. Microphotograph of immunohistochemistry showing co-expression of CK5&6 in areas of squamous differentiation.

prognosis than conventional adenocarcinoma according to previous reports. Compared to adenocarcinoma, ASC of the AmV is a rare malignancy. Preoperative diagnosis is difficult because of the lack of defining characteristics in imaging studies and the difficulty in acquiring both malignant components by limited biopsy. All of the patients have a dismal prognosis, and most of them have early distal metastasis after surgery.

A review of the literature revealed only five reported cases of ASC of the AmV. We have summarized these reports, including the clinical course and final prognosis of ASC of the AmV, in Table 1.^{1–4}

ASC of the AmV is a virulent disease with a worse prognosis than that of adenocarcinoma. Majority of patients with ASC of the

Table 1

Study	Case	Age/sex	Stage	Management	Post op distal metastasis
Ueno et al. ¹	1	47/M	IIB	PD	Present
Ri et al. ²	2	62/F	IIA	PPPD	Present
Lee et al. ³	3	48/M	IB	PPPD	–
	4	80/F	IIB	PPPD	–
Song et al. ⁴	5	76/M		Stenting	Present

PD – pancreaticoduodenectomy, PPPD – pylorus preserving pancreaticoduodenectomy.

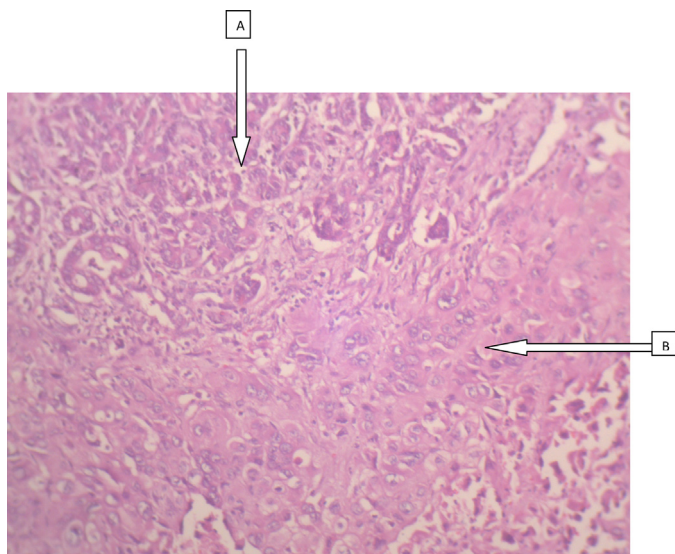


Fig. 2. Microphotograph of H&E staining showing both (a) adenomatous, (b) squamous component.

AmV are managed with operative resection. Surgery remains the mainstay therapy for this disease. A review of the data revealed that the most common procedure performed was Pancreaticoduodenectomy followed by ampullectomy. Surgical interventions do not appear to improve patient survival. Most patients with ASC of the AmV experienced early distal metastasis and short survival after surgery. Our limited experience makes it difficult to determine the clinical course of this disease and the efficacy of surgical intervention. It is hard to draw a conclusion on whether performing major surgery in these patients is beneficial, because of the potentially high morbidity. One report presented two patients with prolonged survival (19 and 46 months after surgery).³ As noted by Lee et al.³ complete surgical resection may prolong patient survival; however, this finding was not supported by other reports.^{1,2} Because of findings of early recurrence and metastasis after surgery, micrometastasis is suspected, however we could not detect it prior to surgery. Currently, there is no established diagnostic tool for detecting micrometastasis. Positron emission tomography may be another diagnostic tool for detecting micrometastasis, but the efficacy of this tool requires more evidence for confirmation.

Some authors have suggested conservative management of patients with ASC of the ampulla of Vater diagnosed before surgery, to reduce postoperative morbidity and achieve comparable survival as observed in patients undergoing surgery.⁴ Although there was no published report evaluating the efficacy of chemoradiation against this disease. In past, the diagnostic term “adenocanthoma” was used to describe tumors that contained both malignant glandular and squamous components. World Health Organization tumor classification system makes a clear distinction between an ASC, in which both glandular and squamous elements are histologically malignant and adenocanthoma which represents adenocarcinoma that contains foci of benign squamous metaplasia.⁵

4. Conclusion

Adenosquamous carcinoma is a very rare form of cancer of the AmV. E.R.C.P. is valuable to establish preoperative diagnosis. Pancreaticoduodenectomy is treatment of choice though early recurrence and distal metastasis may be encountered after surgery. Follow-up should be more frequent to detect possible early

recurrence and distal metastasis in patients with ASC of the AmV.

Conflicts of interest

None.

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None.

Ethical approval

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contributions

Dr. Ashok Y. Kshirsagar: Abstract; Dr. Nitin R. Nangare: Introduction; Dr. Mayank A. Vekariya: Presentation of case and discussion; Dr. Vaibhav Gupta: Conclusion; Dr. Akshay S. Pednekar: References; Dr. J. V. Wader: Figures; Dr. Abhishek Mahna: Figures.

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