CONCURRENT OCCURRENCE OF PERIAMPULLARY DISTAL CHOLANGIOCARCINOMA AND MUCINOUS CYSTADENOCARCINOMA OF THE OVARY: A RARE CASE REPORT

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ABSTRACT

In this case report, the clinical characteristics, diagnostic approach, and outcome of a patient with coexisting periampullary distal cholangiocarcinoma and mucinous cystadenocarcinoma of the ovary were described. This case underscores the importance of considering rare tumor combinations and highlights the diagnostic challenges and limited treatment options in such cases.

Keywords: Periampullary cholangiocarcinoma, Mucinous cystadenocarcinoma, Ovary, Concurrent malignancies.

INTRODUCTION

Primary bilateral ovarian involvement occurs as the most common neoplasms. For invasive micropapillary serous carcinoma, bilaterality seems to be more prevalent with about 80–90% [1]. Cystadenocarcinoma signifies an infrequent entity, accounting for approximately 20% of all periampullary cancers [2]. However, insufficient literature exist concerning the ideal therapeutic approach for this carcinoma because of rarity, and physicians frequently encounter significant difficulties in the management of these malignancies. The coexistence of multiple primary malignancies in a single patient is a rare occurrence, often posing diagnostic and therapeutic challenges. In this case report, we aim to present the clinical characteristics, diagnostic approach, and outcome of a patient with coexisting periampullary distal cholangiocarcinoma and mucinous cystadenocarcinoma of the ovary. By highlighting this rare occurrence, we hope to contribute to the existing literature and raise awareness among clinicians about the potential coexistence of these malignancies, which may have implications for diagnostic evaluation and treatment planning.

CASE PRESENTATION

A 62-year-old female presented with severe abdominal pain, abdominal fullness, and yellowish discoloration of the skin. Clinical examination revealed that a palpable, thrill, and ascitic fluid was tapped and sent for cytological evaluation. Laboratory investigations showed elevated bilirubin levels (6 mg%), raised alkaline phosphatase levels (1000 U/L), and increased Cancer antigen 125 (CA-125) levels (120,000 u/L). Ascitic fluid analysis revealed malignant cells.

Ultrasonography revealed an ill-defined heteroechoic mass lesion in the periampullary region with involvement of the extrahepatic common bile duct. The mass is causing gross upstream central and peripheral dilatation of the left and the right biliary radicles

CECT revealed a well-defined soft-tissue mass in the periampullary location with involvement of the extrahepatic CBD measuring approximately 4.6×3.7×8.4 cm in size. The mass was seen to extend till the perihilar region at the level of confluence of the intrahepatic biliary radicles along with invasion of the proximal right intrahepatic biliary radicle. The mass was also involving the second part of the duodenum and the head of the pancreas. CECT also showed a well-defined multiloculated cystic mass with eccentric solid component and nodular peripherally enhancing wall and thin enhancing internal septae in the right adnexa with gross ascites.

Biopsy of adnexal lesion confirmed mucinous cystadenocarcinoma and biopsy of the periampullary tumor showed cholangiocarcinoma. Endometrial biopsy showed hyperplastic cells, ruling out neoplasia.

A rare diagnosis of periampullary distal cholangiocarcinoma (Modified Bismuth Collette classification Type IIIa) with co-existing mucinous cystadenocarcinoma of the ovary (Stage Ic) was made (Figs. 1-3).

Management and outcome

Chemotherapy was initiated for the patient in March 2021. However, despite treatment, the patient’s condition deteriorated, and she passed away in May 2021.

Fig. 1: (a) Non-contrast axial image (b) Arterial phase axial image (c) Venous phase axial image (d) Delayed phase axial image.
The coexistence of periampullary distal cholangiocarcinoma and mucinous cystadenocarcinoma of the ovary is an extremely rare finding. Periampullary cholangiocarcinoma accounts for approximately 6–10% of all cases of biliary tract malignancies and is associated with poor prognosis [3,4]. On the other hand, mucinous cystadenocarcinoma is a subtype of ovarian cancer characterized by the presence of mucin-producing epithelial cells. It represents approximately 10–15% of all ovarian malignancies and is often diagnosed at an advanced stage [5,6]. There are only a few reported cases in the medical literature that document the simultaneous presence of these two distinct malignancies. In a case report published by Lee et al., ovarian masses with metastatic adenocarcinoma originating from carcinoma of the choledochal cyst has been reported [7].

Another case study by Khangura et al. documented a similar rare occurrence. The authors reported a case of a 76-year-old woman with final pathology results yielded a well-differentiated mucinous adenocarcinoma consistent with metastasis from a primary cholangiocarcinoma. The study discussed the diagnostic challenges posed by this combination of malignancies [8].

Both tumors are known to have different anatomical origins and do not commonly metastasize to these respective locations. The etiology and pathogenesis of this concurrent occurrence remain unclear, possibly involving shared risk factors or genetic predisposition. This case underscores the importance of considering rare tumor combinations and highlights the diagnostic challenges and limited treatment options in such cases.

CONCLUSION

This case emphasizes the need for heightened awareness of unusual tumor combinations and the significance of interdisciplin ary collaboration in the management of complex malignancies. Further research is warranted to elucidate the underlying mechanisms and explore potential therapeutic strategies for similar cases.

AUTHORS CONTRIBUTION

Dr. Bhavya Kataria involved in manuscript writing, editing and submitting to the journal.

CONFLICTS OF INTERESTS

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REFERENCES


Fig. 2: (a-c) Venous phase axial images of the adnexa: The right adnexa shows a well-defined multiloculated cystic mass with eccentric solid component within. The mass shows multiple thin enhancing septa within and peripheral nodular wall enhancement. Gross ascites is noted in the pelvis and the peritoneal cavity

Fig. 3: Coronal images (a) non-contrast; (b) Delayed images acquired at 20 min, on delayed images, the mass shows more uniform enhancement as compared to the venous phase images