Malignant Transformation in Heterotopic Pancreatic Tissue in a Patient with BRCA2 Mutation

Nicholette Goh[1], Danson Xue Wei Yeo[1], Khoon Leong Chuah[2], Sanghvi Kaushal Amitbhai[1], Aung Myint Oo[1], Abdul Kareem Saleem Ahmed[1], Koura Aaryan Nath[1]

[1]*Department of General Surgery, Tan Tock Seng Hospital*
[2]*Department of Pathology, Tan Tock Seng Hospital*

Background

Malignant transformation of heterotropic pancreatic tissue is a rare entity with only several case reports published in scientific literature. Adjuvant chemotherapy following oncological resection for lesions with nodal metastasis has not been well described and there are no guidelines available to guide the management of these patients.

Case Presentation

We present a case of a 33-year-old Chinese male initially presenting with dyspepsia for which an oesophagoduodenoscopy was performed. This revealed a submucosal bulge with central umbilication at the antrum (Image 1) and a subsequent computed tomographic (CT) scan of the abdomen and pelvis showed a 2.3cm enhancing nodule over the anterior aspect of the pylorus suggestive of a GIST (Images 2 and 3).

The patient underwent laparoscopic wedge resection of the tumour with on-table gastroscopy for visualisation. Histopathological assessment (Images 4) demonstrated the presence of heterotopic pancreatic tissue displaying cellular proliferation resembling acinar cells and ducts of the pancreas.

An adjacent invasive adenocarcinoma comprising malignant glands was present and the tumour extended to the radial and deep margins. On immunohistochemistry, the tumour showed findings consistent with a pancreatic ductal adenocarcinoma arising from heterotopic pancreas within the stomach.

Given the involvement of the wedge resection margins, the patient underwent a distal gastrectomy with oncological lymph nodal clearance. 1 out of the 33 harvested lymph nodes harboured micrometastasis while the main gastrectomy specimen did not have any residual malignancy.

Post-operatively, the patient underwent adjuvant chemotherapy with FOLFOXIRI (oxaliplatin, irinotecan, folinic acid, fluorouracil). The chemotherapy regime was guided by the standard of care for pancreatic cancer after discussion at our multi-disciplinary tumour board meeting.

Post-surgery, the patient underwent genetic testing and was found to be positive for BRCA2 mutation. Of note, he does not have a significant family history of malignancy and genetic screening was offered to his first degree family members. The patient will continue to be closely followed up for other BRCA2 related malignancies like prostatic cancer.

Conclusion

Pre-operative diagnostic confirmation is challenging and endoscopic procedures pose a significant false negatives. Reports of nodal metastasis following oncological resection are limited and there are no guidelines regarding adjuvant therapies. We would recommend a chemotherapy regimen similar to that for primary locally advanced pancreatic carcinoma in patients found to have nodal metastasis.



Image 1
Endoscopic photograph of submucosal buldge with central umbilication

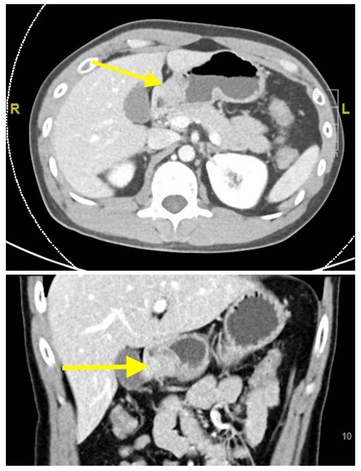


Image 2 and 3
Computed tomographic scan images demonstrating the tumour over the anterior aspect of the gastric pylorus (Arrow)

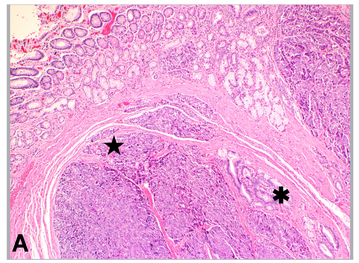


Image 4
Low power view revealing the presence of heterotopic pancreatic tissue with acinar cells (star) and pancreatic ducts (asterisk) in the submucosa of the stomach. (H&E x40)