90 CONCURRENT ENTEROPATHY-ASSOCIATED T-CELL LYMPHOMA AND JEJUNAL ADENOCARCINOMA IN PATIENT WITH COELIAC DISEASE: A CASE REPORT

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Introduction Although coeliac disease (CD) is classically associated with malabsorption and the attendant complications arising from it, population studies have shown that patients with celiac disease (CD) are at increased risk of malignancy especially lymphoproliferative malignancy and gastrointestinal cancer. This risk is especially higher in patients with refractory celiac disease (RCD) type II, for enteropathy-associated T-cell lymphoma (EATL), and in patients who are not compliant with gluten-free diet (GFD).

Aims/Background There have been a number of case reports of either EATL or small bowel cancer arising in patients with CD. Review of available literature reveals small number of cases in which both EATL and small bowel cancer occur concurrently in CD.

Method We report a patient with known celiac disease who was diagnosed post-humously with concurrent enteropathy-associated

T-cell lymphoma, jejunal adenocarcinoma and refractory celiac disease (RCD) type II.

Results This is a case of 65-year-old female with osteoporosis who was diagnosed with CD in 2008. She had dietetic input at her regional hospital due to initial difficulties adhering to strict GFD. Two years following her diagnosis, she had a CT-scan of the abdomen to investigate microcytic hypochromic anaemia and this revealed extensive reactive mesenteric lymphadenopathy and minimal circumferential regular wall thickening in the small bowel. She also had a small bowel follow-through and colonoscopy which did not identify any significant lesions. A follow-up CT scan in January 2011 showed resolution of the lymphadenopathy but a mildly thickened small bowel loop.

She then developed right inguinal mass with CT-scan showing solitary focus of presumed lymph node enlargement in the right thigh. Biopsy of this mass indicated a peripheral T-cell lymphoma (CD30+ALK negative) diagnosis and she was treated with six cycles of CHOEP chemotherapy regime with end of treatment in April 2012. Follow-up imaging (CT PET) showed reduction in the size of the inguinal nodal mass but there were FDG-avid involvement of the small bowel. She presented acutely in July 2012 with sepsis and CT of the abdomen performed on admission showed small bowel mass in the left flank with mural thickening and multiple faint hypo-densities scattered throughout the liver. Whilst being investigated for this mass, during the same inpatient stay, she deteriorated (upper gastrointestinal bleeding, coagulopathy, intra-abdominal haemorrhage) which necessitated urgent laparotomy with resection of the mass. She had a difficult course postoperatively. Decision was made to withdraw escalated care as she remained unwell despite maximum supports. Patient then died.

Histology of the resected jejunal mass was reported posthumously. Sections of the jejunal tumour confirmed moderately differentiated adenocarcinoma. The morphology and immune-profile of the surrounding lymph nodes, including review of the right inguinal mass specimen, were consistent with EATL. Immunohistochemistry performed on non-neoplastic adjacent jejuna mucosa showed expression of CD3 lymphocytes and loss of CD8 lymphocytes consistent with RCD type II.

Conclusion This case is one of the few cases reported in literature of concurrent EATL, in the gastrointestinal tract and peripherally, and adenocarcinoma of the small bowel in patients with CD. It highlights the importance of close follow-up of patients with CD who are still symptomatic despite GFD.