Metastatic Paediatric Colorectal Carcinoma

Abstract:
R Woods, JO Larkin, C Muldoon, MJ Kennedy, B Mehigan, P McCormick
St James Hospital, James St, Dublin 8

A 16 year old girl presented to our unit with crampy abdominal pain, change in bowel habit, a subjective impression of weight loss and a single episode of haematochezia. She was previously well with no family history of colorectal carcinoma although her paternal aunt and grandmother had Crohn's disease. Physical examination was unremarkable. Haematological and biochemical investigations were normal and Carcinoembryonic Antigen (CEA) was less than 1 ng/ml. Colonoscopy showed an irregular, impassable circumferential lesion, confirmed on biopsy to be at 13cm. Biopsies showed poorly differentiated signet ring adenocarcinoma positive for Cytokeratins 7 and 20. Computed tomography (CT) and magnetic resonance imaging (MRI) showed mural thickening at the rectosigmoid junction staged as T3N0 with a small amount of free fluid in the pelvis and no evidence of metastasis (Figure 1).

Following a multidisciplinary meeting, she underwent laparoscopy, where peritoneal carcinomatosis was noted, with extensive tumour plaques present in the pouch of Douglas. A palliative anterior resection was carried out.

Histopathology showed poorly differentiated signet ring adenocarcinoma of the rectosigmoid junction, with a high degree of lymphovascular invasion, serosal involvement, metastatic involvement of 23 of 43 nodes and confirmed peritoneal metastases leading to a staging of pT4a pN2b pM1 (Figure 2). The appendix was also removed and was histologically normal. Following an uneventful postoperative recovery the patient commenced a course of palliative chemotherapy with Folinic acid, 5-Fluorouracil, Oxaliplatin and Bevacizumab (Folfox and Avastin). She is alive and well ten months postoperatively with no evidence of local disease recurrence or distant metastatic disease on follow-up CT.

Figure 1: axial CT image showing circumferential rectosigmoid mass

Discussion
Recent trends suggest that the incidence of rectal and rectosigmoid cancer is increasing in patients aged under 40 years while the incidence of colon cancer remains largely static. Nonetheless, the disease is increasing in overall incidence and the predominantly aggressive tumour biology equates to an overall poor prognosis. We report the case of a 16 year old girl with signet ring cell rectosigmoid adenocarcinoma and discuss the literature relating to paediatric colorectal cancer.

Case Report
A 16 year old girl was referred to our outpatient clinic with a four week history of crampy abdominal pain, change in bowel habit, a subjective impression of weight loss and a single episode of haematochezia. She was previously well and underwent laparoscopy, where peritoneal carcinomatosis was noted, with extensive tumour plaques present in the pouch of Douglas. A palliative anterior resection was carried out.

Histopathology showed poorly differentiated signet ring adenocarcinoma of the rectosigmoid junction, with a high degree of lymphovascular invasion, serosal involvement, metastatic involvement of 23 of 43 nodes and confirmed peritoneal metastases leading to a staging of pT4a pN2b pM1 (Figure 2). The appendix was also removed and was histologically normal. Following an uneventful postoperative recovery the patient commenced a course of palliative chemotherapy with Folinic acid, 5-Fluorouracil, Oxaliplatin and Bevacizumab (Folfox and Avastin). She is alive and well ten months postoperatively with no evidence of local disease recurrence or distant metastatic disease on follow-up CT.

Figure 2: H&E stain x20 of (a) primary tumour showing poorly differentiated signet ring adenocarcinoma replacing the mucosa, submucosa and muscularis propria, positive for Cytokeratins 7 and 20 and (b) metastatic peritoneal deposit

References