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Case Report

The treatment challenges of an exceedingly rare diagnosis of rectal small cell neuroendocrine carcinoma: a case report

Lithira R. Walisinghe[1,2]*, Frana-Katica Batistic[2], Jennifer Borowsky[3], Raphael Varghese[2]

[1]Department of Surgery, Ipswich Hospital, Ipswich, Queensland, Australia
[2]School of Medicine and Dentistry, Griffith University, Southport, Queensland, Australia
[3]Department of Anatomical Pathology, Princess Alexandra Hospital, Woolloongabba, Queensland, Australia

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***Correspondence:**
Dr. Lithira R. Walisinghe,
E-mail: lithira.walisinghe@health.qld.gov.au

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ABSTRACT

Small cell neuroendocrine carcinoma (SCNEC) of the rectum is a rare, aggressive malignancy with limited evidence guiding management. We report a case of T4N1b rectal SCNEC in a female presenting with severe proctalgia. Imaging and endoscopy identified a near-obstructing rectal mass, with histology confirming SCNEC. Staging demonstrated locoregional disease without distant metastases. Initial management with platinum-based chemotherapy, etoposide, and radiotherapy achieved partial response; however, persistent disease abutting the posterior vaginal wall necessitated abdominoperineal resection (APR) with partial vaginectomy. Histology revealed poor treatment response and close margins, indicating high recurrence risk. She underwent close surveillance, ultimately progressing to metastatic disease 8 months later, and underwent further adjuvant chemotherapy and immunotherapy. She was eventually palliated with continued deterioration secondary to febrile neutropaenia and myelodysplastic syndrome, ultimately passing away a further 2 months later. This case highlights the aggressive nature of rectal SCNEC, uncertainty surrounding optimal management, and the need for flexible, multidisciplinary approaches. Further research is required to establish evidence-based disease specific treatment algorithms for a diagnosis that is void of such definitive guidelines and treatment strategies which are largely extrapolated.

Keywords: Rectal cancer, Rectal small cell neuroendocrine carcinoma, Colorectal surgery, Radiotherapy, Chemotherapy

INTRODUCTION

Neuroendocrine tumours (NET) are a heterogeneous group of epithelial tumours arising from neuroendocrine cells distributed throughout the body.[1] Although the gastrointestinal tract is the most common site of occurrence, rectal NETs are rare, accounting for approximately 1-2% of all rectal neoplasms.[2-4]

Poorly-differentiated NET of the rectum, such as small-cell neuroendocrine carcinomas (SCNEC), are particularly rare, comprising <1% of all colorectal cancers.[5] SCNECs are highly aggressive, with most patients having disseminated disease at diagnosis.[1,5-7]

Whilst management of non-small cell rectal NET follows clear guidelines with multi-modal management options (including endoscopic, surgical, hormonal, immunosuppressant, and the chemotherapeutic treatment), SCNEC is considered a separate entity, and the low incidence has limited development of robust evidence-based treatment guidelines.[2,5,7-9]

Current management is derived from small retrospective studies and case reports, with treatment approaches typically extrapolated from that of small cell lung cancer (SCLC), due to shared morphological, biological and clinical features.[4-6,8-10]

Surgery with or without adjuvant treatment can be considered in localised or resectable disease, while platinum-based chemotherapy regimens (such as cisplatin or carboplatin with etoposide) form the backbone of treatment for most SCNECs, mirroring lung protocols.[2,6-11] Given the generally low certainty behind treatment options in SCNEC, multidisciplinary team (MDT) assessment is essential to guide management decisions, and close follow-up is required given the high risk of early progression and recurrence.[7,11]

Outcomes for SCNEC remain poor, with 11-12 months median survival in metastatic disease, and 5-year overall survival (OS) as low as 6%, despite advances in diagnostics and treatment.[3,5,7]

We report a rare case of recurrent T4N1b rectal SCNEC in a 73-year-old female. Initially diagnosed in 2023, she was managed with a diverting loop colostomy and chemoradiotherapy, achieving tumour regression. Recurrence was identified in 2025 with T2N0 disease, with subsequently open APR and partial vaginectomy. Histopathology demonstrated close margins and poor treatment response. Despite remaining at close 3-monthly surveillance, she progressed to metastatic disease 8 months post operatively (post-op). She received a short period of adjuvant chemotherapy and immunotherapy, however was ultimately palliated with continued deterioration following a hospital admission for febrile neutropaenia and myelodysplastic syndrome, passing away 10 months post-op.

CASE REPORT

A 73-year-old female was referred to the emergency department from a private colorectal clinic in September 2023, with 2 months of severe proctalgia precluding examination, but no bowel changes or rectal bleeding. Her history included neuropathic pain, open cholecystectomy, caesarean section, tubal ligation, and dyslipidaemia, with no family history of colorectal cancer. Investigations were unremarkable aside from a C-reactive protein of 19. Computed tomography of the abdomen and pelvis (CTAP) demonstrated a circumferential enhancing rectal wall thickening over a 51 mm segment with locoregional lymphadenopathy, suggestive of primary rectal malignancy (Figure 1).

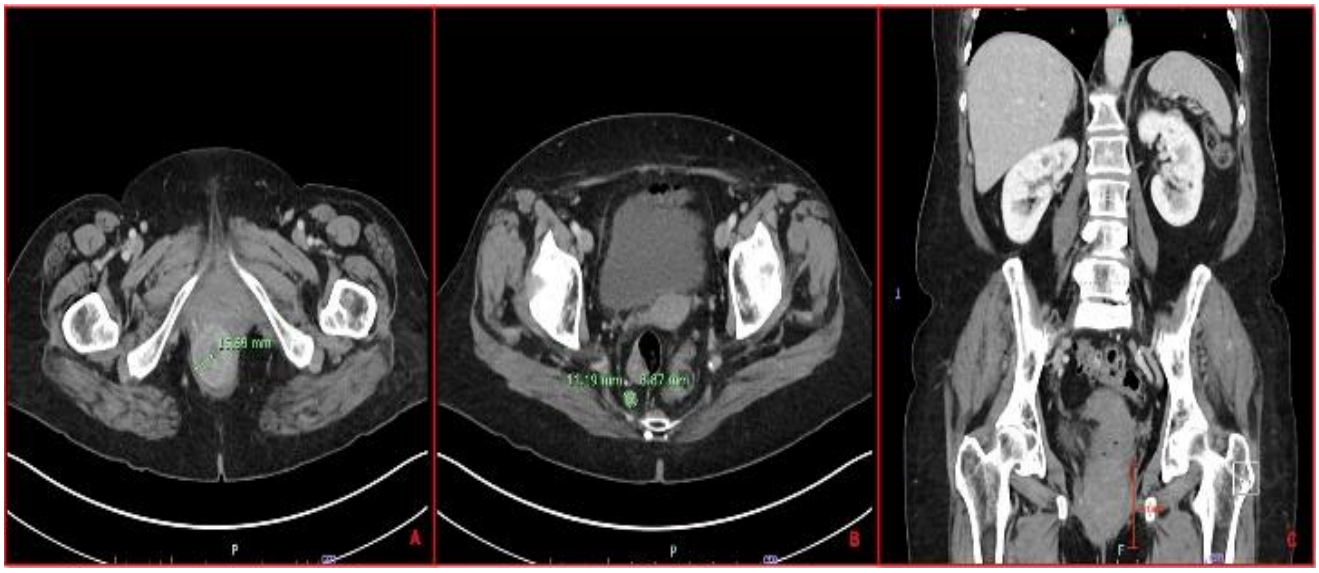


Figure 1 (A-C): CTAP; A) axial sections of circumferential mural thickening of the lower rectum, slightly more pronounced at right distal aspect (green arrow); B) Axial section demonstrating a pathologically enlarged pre-sacral lymph node (green arrows); C) Coronal section emphasising the cranio-caudal length of circumferential thickening as 51 mm (red arrow).

Emergent flexible sigmoidoscopy revealed a large, near-obstructing ulcerated mass involving the rectum and anal canal, occupying two thirds of the lumen circumference (Figure 2). Notably, colonoscopy two years prior was normal. Histology confirmed SCNEC, either primary or metastatic (Figure 3).

Staging CT excluded distant metastases, while pelvic MRI demonstrated T4N1b disease with a predominantly right-sided lesion invading the internal anal sphincter, with suspected involvement of the external sphincter and possibly the right puborectalis (Figure 4). A diverting loop colostomy was performed soon after due to impending obstruction. A following MDT consensus in October recommended dual Dotatate and fluorodeoxyglucose (FDG) positron emission tomography (PET), followed by treatment analogous to SCLC. Both Dotatate and FDG PET showed intense uptake at the site of the rectal lesion and a right inferior mesolectal node concordant with suspected locoregional disease (LRD) and lymph node metastasis. This also suggested potential heterogenous biology of a poorly and well differentiated neuroendocrine tumour (Figure 5).

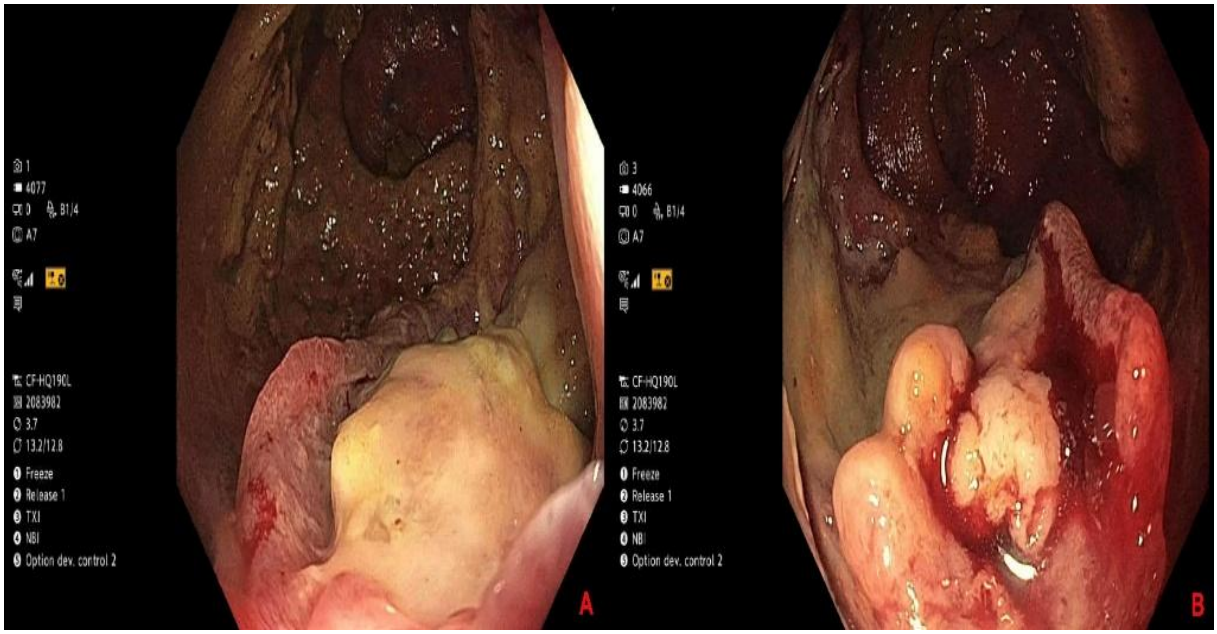


Figure 2 (A and B): A-Flexible sigmoidoscopy images showing an ulcerated mass located at the lower extremity of the rectum and upper anal canal, partially circumferential, involving two-thirds of the lumen circumference. B- also highlights easily bleeding vascular lesion post biopsy.

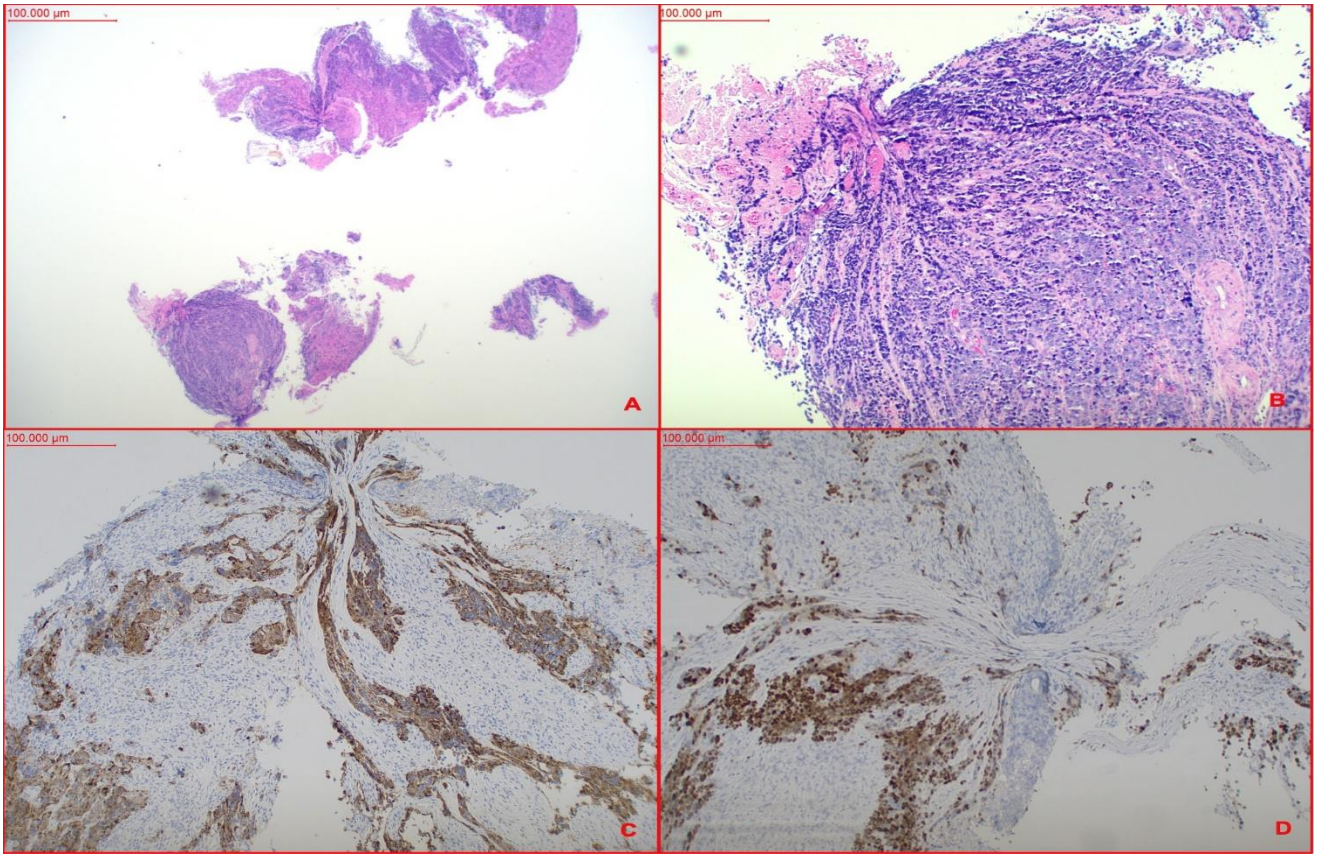


Figure 3: Histopathology sections; A and B show haematoxylin and eosin-stained slides of low and high power respectively showing multiple pieces of tumour composed of small cells arranged in sheets and nests with high nuclear to cytoplasmic ratio, scant cytoplasm, and speckled chromatin. C and D show synaptophysin (SYN) and insulinoma associated protein 1 (INSM1) immunohistochemistry respectively, both positive indicating a SCC.

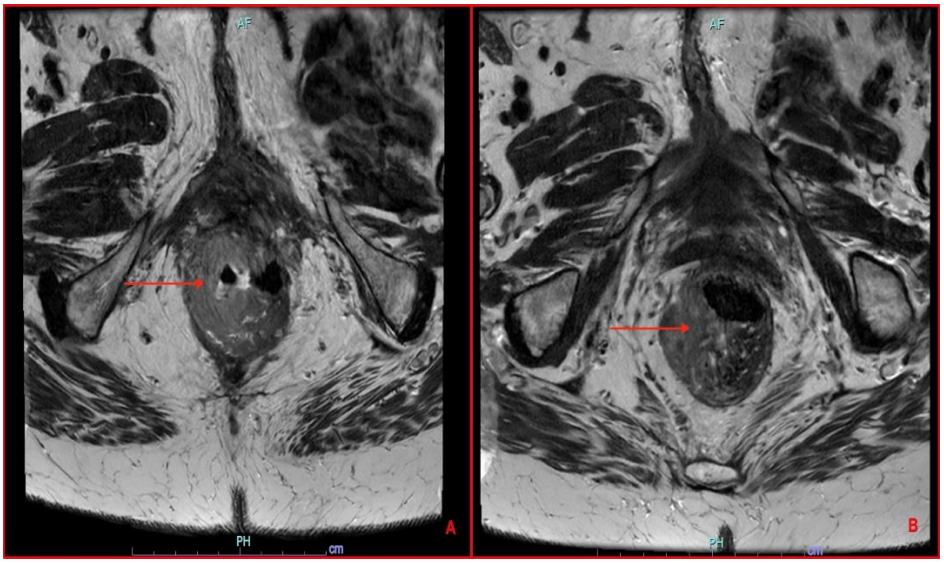


Figure 4 (A and B): A and B showing MRI axial sections from initial diagnosis highlighting a bulky predominantly right sided, near circumferential very low rectal carcinoma (red arrows).

*There is invasion of the right aspect of the internal sphincter with possible involvement of the external sphincter. There is equivocal involvement of the right puborectalis and no involvement of the right levator ani. It is deemed T4N1 disease (suspicious lymph node not visualised in above Figure).

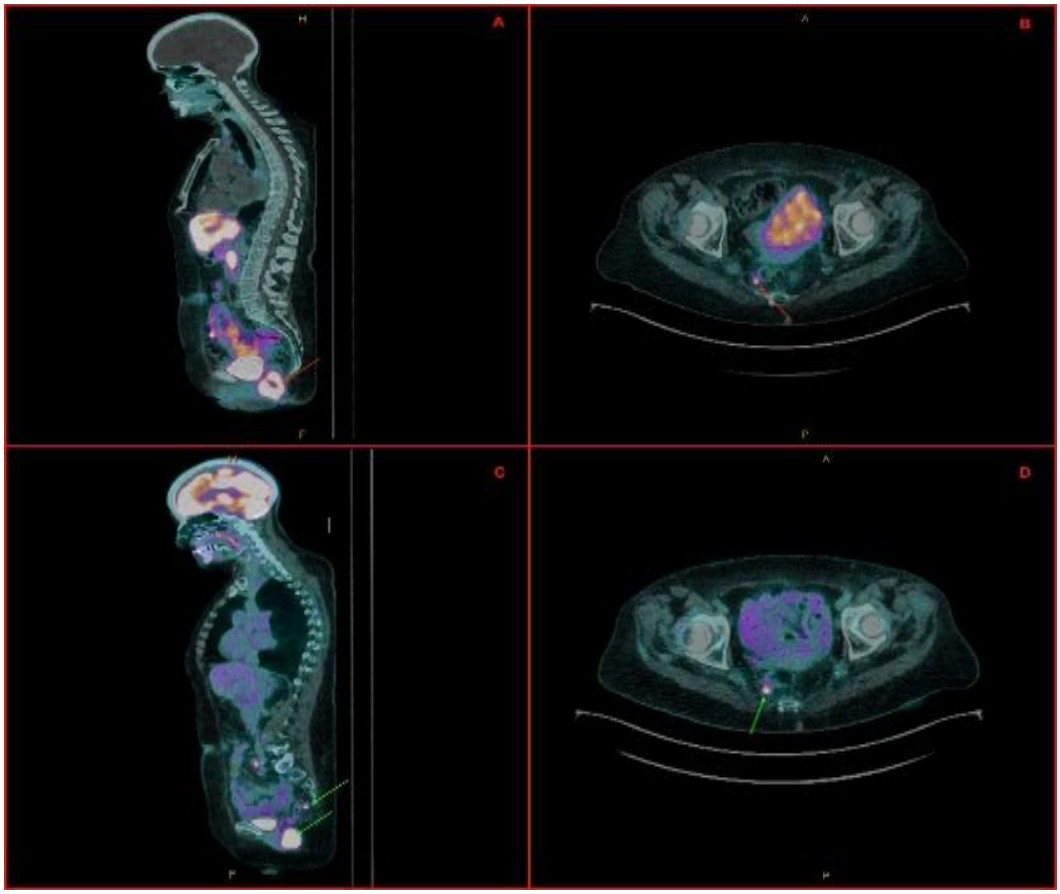


Figure 5 (A-D): A and B showing Dotatate PET sagittal and axial, C and D showing FDG PET sagittal and axial images respectively. Both displaying intense avidity in the location of known low rectal lesion (arrow in A and C inferior arrow), and also highlight an avid presacral lymph node (arrows in B and C-superior arrow D).

*This is in keeping with locoregional disease with regional lymph node metastasis.

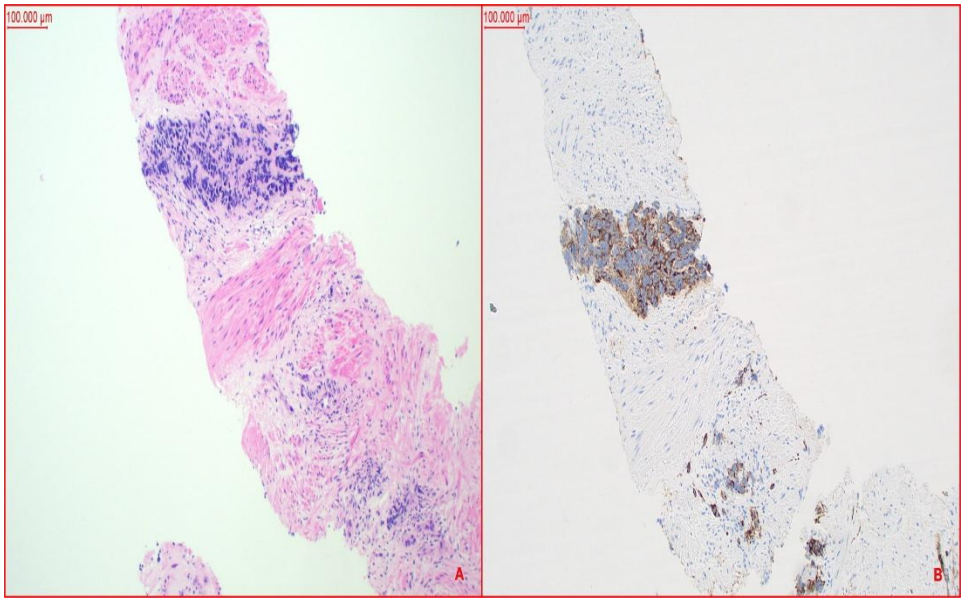


Figure 6 (A and B): A-H and E-stained slide of the rectal biopsy obtained from EUA showing skeletal muscle and fibrous tissue containing residual SCC characterised by nests and sheets of tumour cells with pleomorphic oval nuclei, fine chromatin and scant cytoplasm. B-shows positive SYN immunohistochemistry staining indicating SCC.

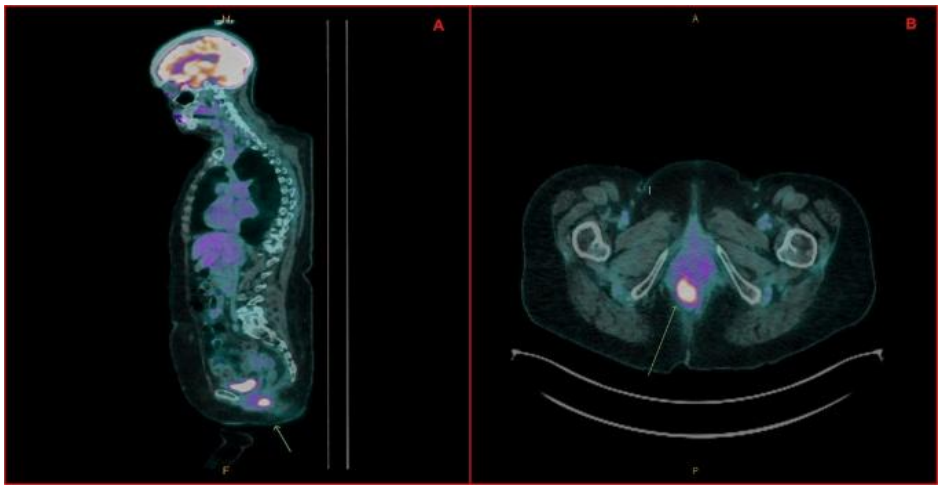


Figure 7 (A and B): Sagittal and axial sections of FDG PET imaging respectively, with persistent intense uptake in right anorectal junction (green arrows). The previously visualised FDG avid nodes are no longer visible here.

Initial treatment comprised three cycles of cisplatin and etoposide, radiotherapy, then three cycles of carboplatin and etoposide, completed over a period of 12 months. There was a short-lived apparent treatment response, with partial volume reduction on MRI and FDG PET with moderate avidity and no nodal disease. However, repeat examination under anaesthesia (EUA) biopsy in the February 2025 confirmed residual tumour at the depth (Figure 6).

After four additional cycles, PET imaging showed persistent intense FDG uptake (Figure 7).

MRI rectum emphasised a 27.5 mm lobulated mass involving the upper two-thirds of the anal canal, just reaching the anorectal junction. It was noted to involve the external sphincter, puborectalis sling, and abut and indent the posterior wall of the vagina without direct infiltration (Figure 8).

Ultimately, mutual decision was reached for surgery with emphasis of low likelihood of cure, and the patient proceeded to have an (APR, partial vaginectomy, and end colostomy in October 2025.

Histology revealed a 33 mm SCNEC with poor treatment response, invading the internal anal sphincter, with the closest margin 0.5 mm. The tumour was adherent to but did not invade the vaginal wall and lymph nodes were negative (0/16), giving a pathological staging of T2N0 (Figure 9).

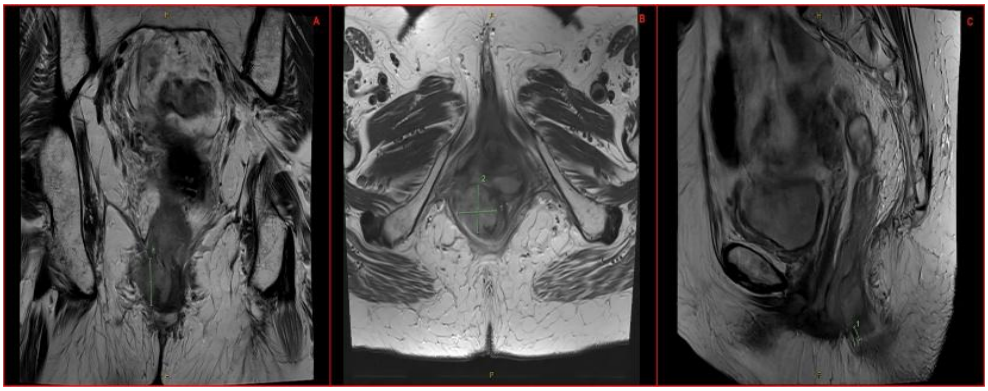


Figure 8 (A-C): Pre op MRI rectum T2 weighted coronal, axial and sagittal sections respectively. A and B show a predominantly ventral and right sided mass in anal canal between 8 to 12 o'clock. Craniocaudal dimension (A green arrow) is 26 mm and axial dimensions are 27.5×26 mm (B green arrows). These images highlight infiltration beyond submucosa to internal sphincter and intersphincteric plane, also involving right sided external sphincter and puborectalis. Lower margin is 8 mm from the anal verge (C green arrow indicates distance to anal verge). B and C also highlight the mass abutting the posterior vaginal wall without direct invasion.

*No adenopathy is identified, hence radiological T2N0.

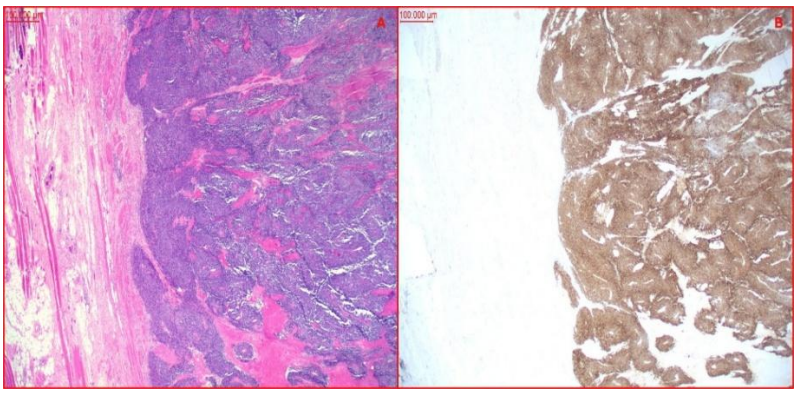


Figure 9 (A and B): Histopathology high powered H and E section and SYN immunohistochemistry respectively.

*H and E shows SCNEC with invasion into but not through the sphincter muscle. SYN again positively indicating SCC.

MDT in November 2025 recommended close surveillance with initially 3 monthly PET scans. Persistent avidity in the locus of her primary tumour raised the question of persistent disease vs. inflammatory change. She ultimately progressed to metastatic disease with biopsy confirmed SCNEC to her T11 paravertebral musculature in early June 2026.

She had subsequently been treated with adjuvant Carboplatin, Etoposide and Nivolumab. In July 2026 she had a prolonged admission under oncology with febrile neutropoenia, and was also later diagnosed with myelodysplastic syndrome. She continued to deteriorate and was eventually palliated in accordance with her wishes. She ultimately passed away in the late August 2026.

DISCUSSION

Colorectal SCNEC, although increasing in incidence in recent years, remains exceedingly rare, forming less than 1% of all colorectal malignancies, with neuroendocrine carcinomas of the anus and rectum comprising an even smaller number.[12,13] Thus, consensus guidelines for treatment of rectal SCNEC are limited; treatment algorithms are largely based on extrapolation from studies of SCLC.[12] The limited literature highlights substantial tumour heterogeneity, contributing to variation in presentation, treatment response and prognosis. These tumours are highly aggressive, frequently presenting with metastases and having poor prognosis even in LRD.[13]

The benefit of surgery and primary resection is uncertain with LRD, with recent retrospective studies by the Mayo clinic showing no significantly longer OS, and some studies showing an almost 50% recurrence rate by 18 months.[6] Despite the survival benefit shown by chemotherapy, the role of adjuvant and neoadjuvant therapy remains uncertain with neither showing definitive evidence of improved disease-free interval or OS.[6,12] Platinum-based chemotherapy regimes in combination with Etoposide remain fundamental in the treatment of small cell carcinomas (SCC), mirroring lung cancer

protocols.[12] Furthermore, addition of radiotherapy to surgery and chemotherapy indicates prolonged survival, however radiotherapy timing and pathological typing have shown no impact on OS.[14]

In our case a T4N1b rectal SCNEC was first treated with platinum-based chemotherapy and Etoposide in addition to radiotherapy, with no initial plans for surgical resection given uncertain curative benefit. Despite some success in treating local nodal metastases and decreasing tumour staging with extended chemotherapy, poor treatment response was determined on subsequent PET imaging highlighting persistent FDG uptake. This was proven on histology, following the patient progressing to an APR. Furthermore, despite LRD, again treatment failure post resection was also highly likely and even greater given close margins and extramural invasion. Eight months post-op she developed metastatic disease with likely early disease recurrence evidenced by persistently avid PET scans around the initial primary tumour site as early as 3 months post-op. The efficacy of her adjuvant therapy could not be fully assessed given her early demise shortly after initiation.

CONCLUSION

Rectal SCNEC is a rare, aggressive malignancy with limited evidence to guide management, and poor outcomes despite aggressive treatment. This case highlights need for an individualised, multidisciplinary approach with adaptability in management according to treatment response, alongside close surveillance for early identification of treatment failure. Further large-scale prospective studies are required to establish clearer treatment pathways and improve outcomes in this uncommon but high-risk disease.

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Ethical approval: Not required

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