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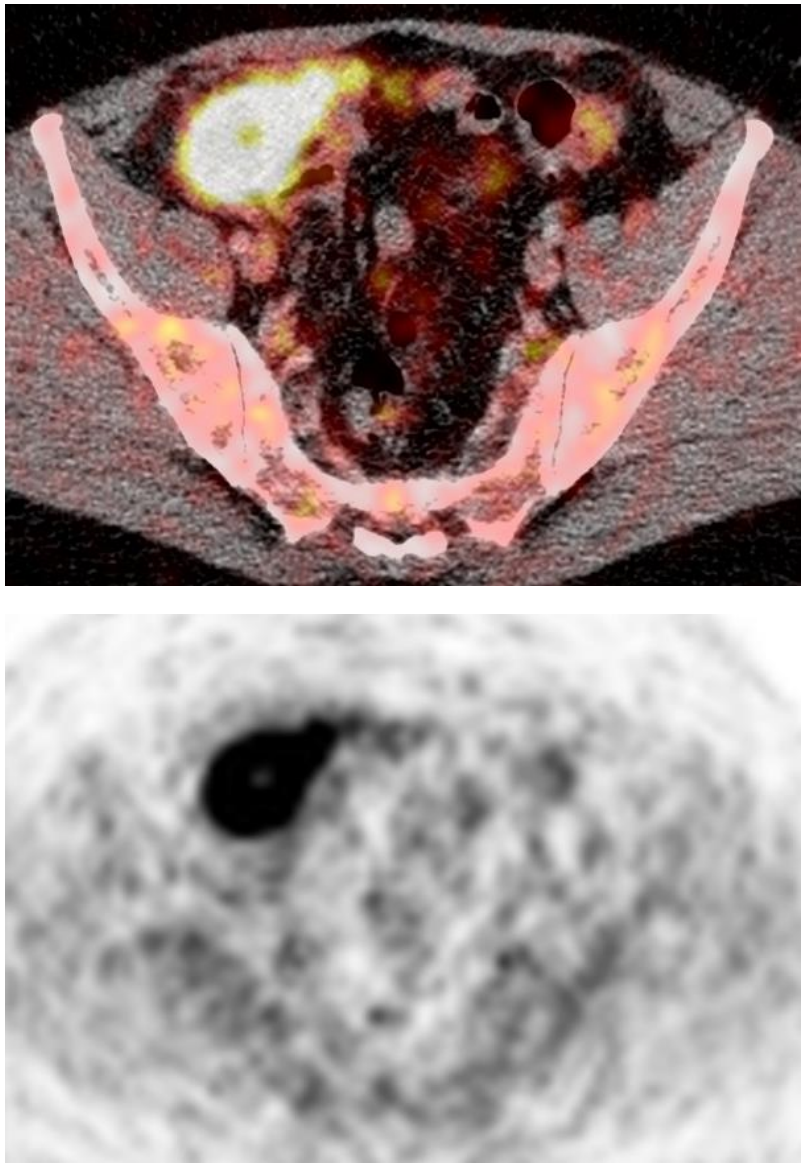
A Rare Case of Non–Hodgkin's Lymphoma of the Appendix

Case Report

- 61 year old woman
- History of gastrointestinal NHL of the stomach 2006, R-CHOP to complete remission
- Eight years later reported 6month history dull RIF pain, flu like symptoms
- Routine blood panel unremarkable
- Raised LDH levels at 236 units/l (120-220 IU/l)
- Elevated beta2 microglobulins were elevated at 5.7mg/l (1.2-2.4 mg/l)
- Anti TTG negative
- Staging CT PET demonstrated increased fluorodeoxyglucose (FDG) uptake at the appendix (SUV max 31) and mild uptake at fundus of stomach

A



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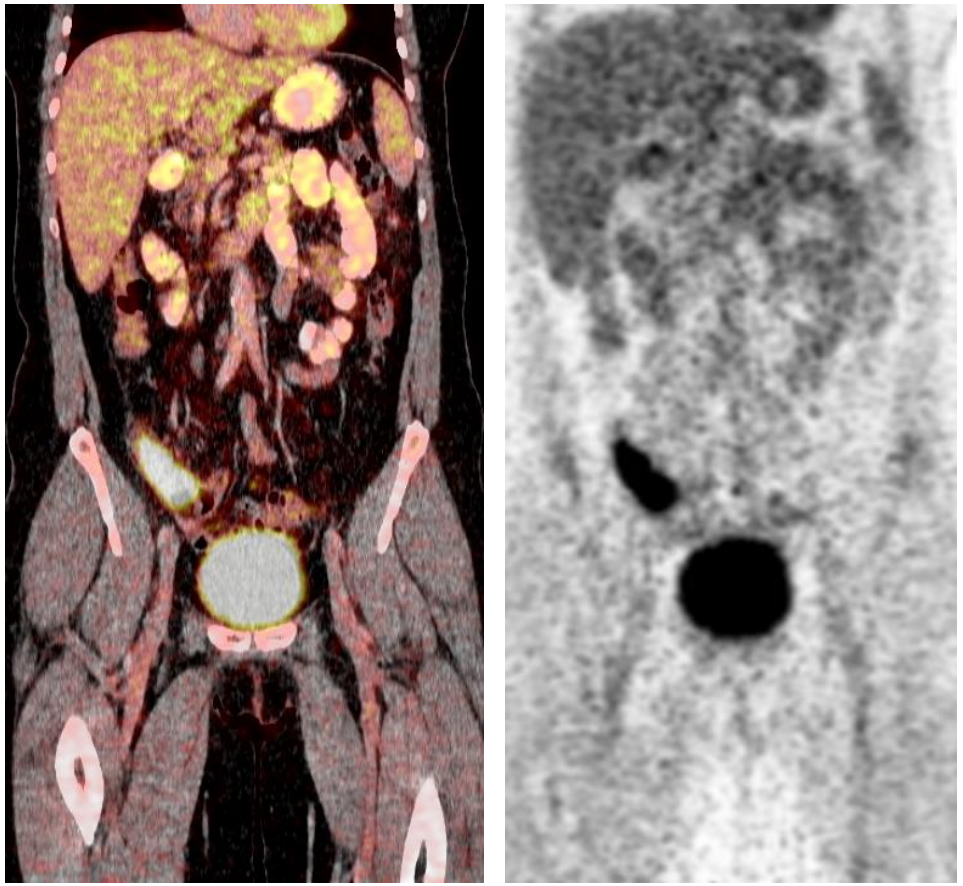
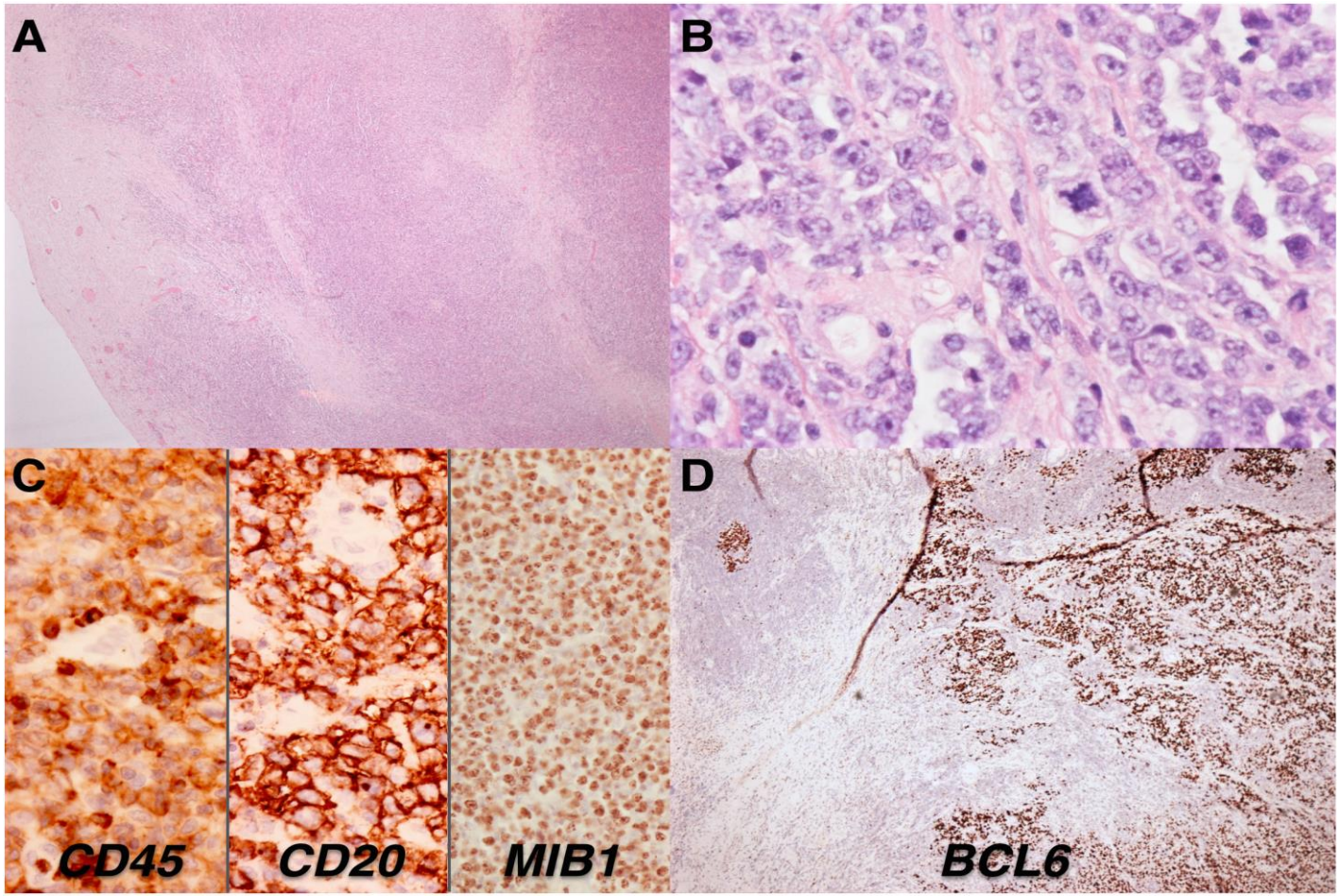


Axial (A) and coronal (B) images from PET CT. Markedly abnormal FDG activity (SUV max 31) within the appendix and caecum.

Investigations

- Gastroscopy normal
- Colonoscopy revealed abnormal swelling of the ileocecal valve
- Biopsies normal
- Discussed at MDM
- Decision made to proceed to appendectomy

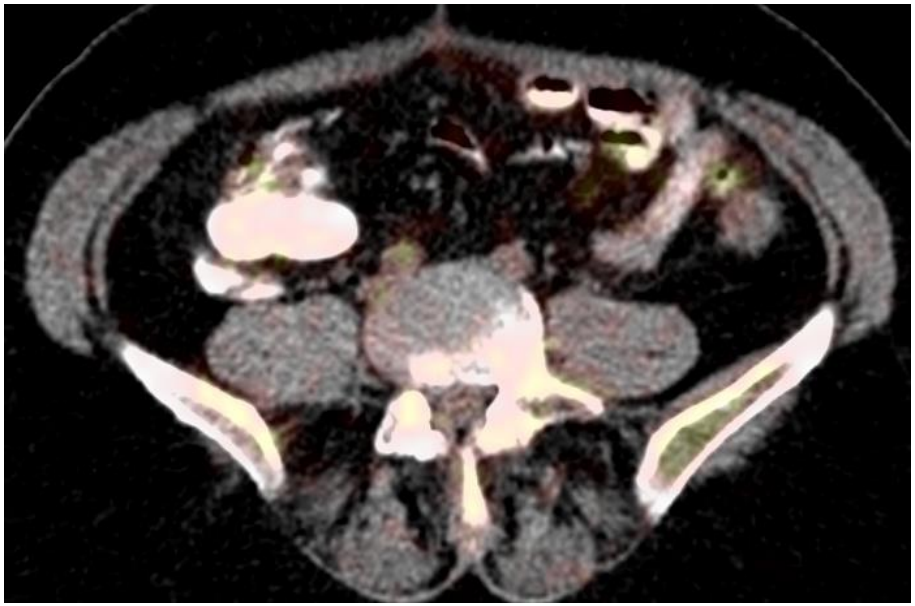


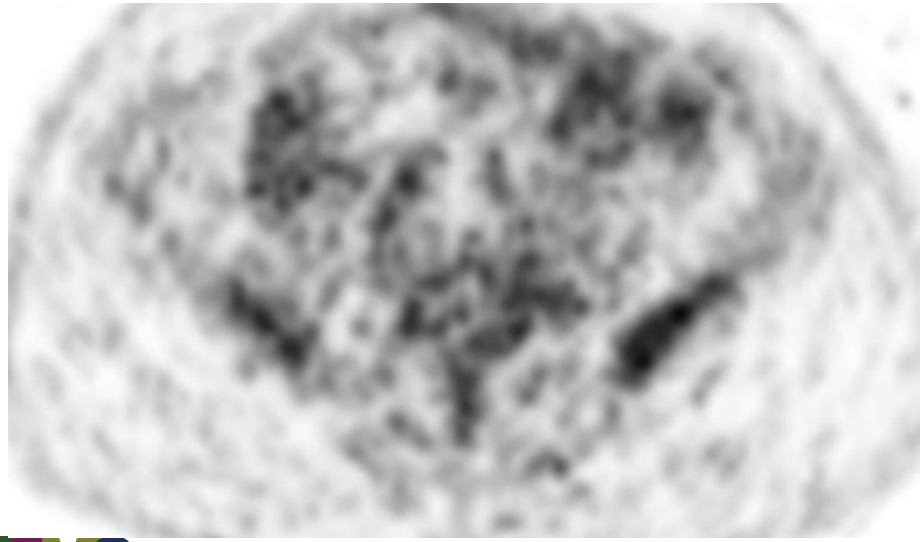
Diffuse Large B Cell of the Appendix. The appendix wall showed a transmural infiltrate (A) of intermediate to large atypical cells (B). The neoplastic cells were positive for CD45 and CD20, while CD3 and CD5 highlighted a background population of T cells, confirming a Diffuse Large B Cell lymphoma (C). The neoplastic cells stained with BCL6, consistent with germinal centre B-cell like cells (D). MIB1 demonstrated a proliferation index of 85% (C).

Diagnosis and Treatment

- Relapsed diffuse large B cell, germinal centre subtype, NHL
- Subsequent bone marrow biopsy showed one intratrabecular lymphoid aggregate with increased reticulin suspicious for involvement
- Treated with salvage RICE (Rituximab, Ifosfamide, Carboplatin, Etoposide) chemotherapy followed by autologous bone marrow transplant
- End of treatment day 100 CT PET showed a complete remission (CR)
- At her most recent follow up 9 months after CR, there was no evidence of recurrence

A





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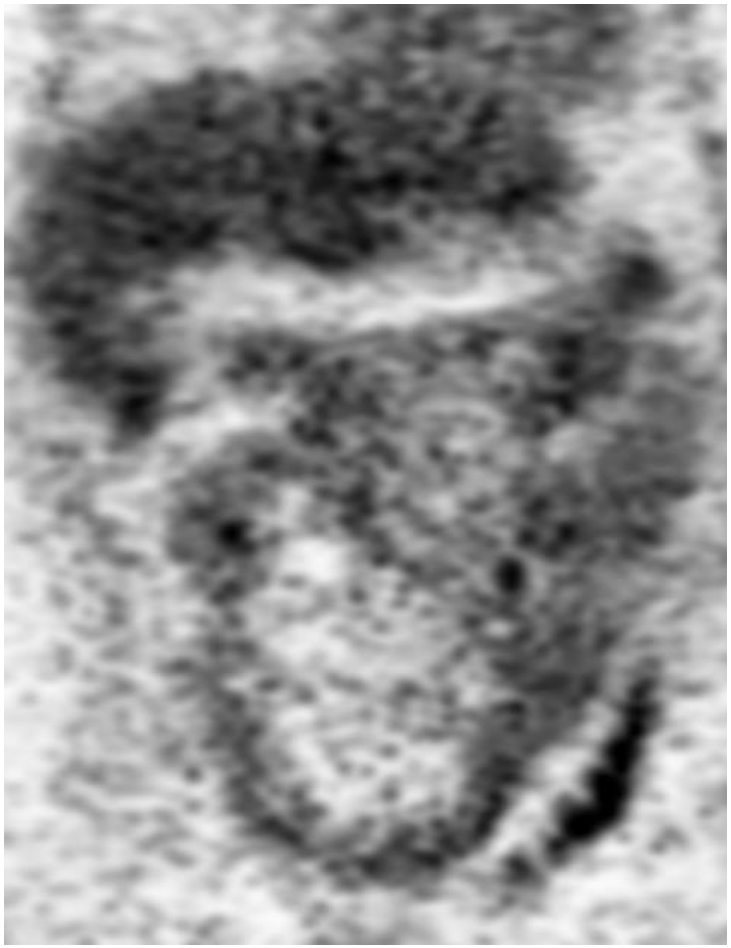

Axial (A) and coronal (B) images from PET CT. No evidence of FDG abnormality in PET CT post treatment

Extranodal NHL

- One third of all NHLs
- GIT most common site (4-20% primary NHLs)
- Stomach and small bowel
- Median age of GIT NHLs 55yrs

- Incidence of primary appendiceal lymphomas estimated at 0.015% (Rao 1991)
- RIF pain, fever, vomiting – suggestive of acute appendicitis
- Can be non-specific – delays diagnosis

- Reaching the diagnosis radiologically prior to surgery is challenging
- Diagnosis often made post op

- Clinical awareness and suspicion is crucial in achieving the correct diagnosis and initiating treatment

Literature Review

References	Year	Number of Cases studied	Number of Appendiceal Lymphomas	Primary or Relapse (Time to relapse)	Age (yrs)	Presenting Symptoms	Primary Lymphoma type (site)
Present Case	2015	1	1	Relapse (8 years)	61	RLQ pain	DLBCL
Tsujimara et al. (4)	2000	1	1	Relapse (8 months)	20	RLQ pain	NK/T-cell Lymphoma (Nasal)
Katz et al. (5)	2002	1	1	Relapse (9 years)	66	Rectal bleeding	Large cell, B cell type
Pickhardt et al. (6)	2001	5	5	Relapse x 1* (6 years)	Mean 54 years	RLQ pain/fever x 3 Lower GI bleeding x 1, Fever/rigors x 1	Mantle cell lymphoma x 2 DLBCL x 1 *Non-Hodgkin's Lymphoma consistent with DLBCL x 1 Large cell undifferentiated malignancy consistent with DLBCL x 1
Chae et al. (7)	2015	1	1	Relapse (3 years)	75	RLQ pain	Mantle Cell Lymphoma
Kitamura et al. (8)	2000	1	1	Primary	84	RLQ pain	T-Cell Non-Hodgkin's Lymphoma
Muller et al . (9)	1997	4	3	Primary	24,69,74	RLQ pain	Diffuse Large, B-Cell (Undifferentiated) Anaplastic Large T cell Marginal Zone B cell
Rao and Aydinalp (10)	1991	1	1	Primary	75	RLQ pain/mass, lower GI bleeding	Lymphoblastic Lymphoma
Fu et al. (11)	2004	1	1	Primary	42	RLQ pain, N+V, fever	DLBCL
Carpenter (12)	1991	1	1	Primary	65	PR Bleeding	Diffuse malignant lymphoma, small Cell, cleaved type
Pasquale et al. (13)	1994	47 (Literature Review)	47	Primary	Mean 25.7 years	RLQ pain x 31, Incidental finding x 5, Non specific symptoms (Abdominal pain, fever, nausea, vomiting, anorexia)	Lymphoblastic sarcoma x 25, Giant Follicular Lymphoblastoma x 9, Lymphosarcoma (unclassified) x 3, Well differentiated lymphocytic x 3, Diffuse large cell x 3, Burkitt's x 3, Unknown x 1
Nanji and Anderson (14)	1983	1	1	Primary	22	Epigastric/periunbilical pain	Burkitt's Lymphoma

Management

- No clear guidelines
- Successfully treated with appendectomy, +- limited right hemicolectomy

- Adjunctive therapy depends on the stage of the disease and the histopathology
- Our patient treated with salvage RICE chemotherapy followed by autologous bone marrow transplant due to her suspicious bone marrow biopsy

Summary

- Rare
- Clinical awareness and suspicion is crucial in achieving the correct diagnosis and initiating treatment