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Primary Small Bowel Malignancy in Paediatrics - A Case Report

Case

- 13-year-old boy
- History of 8 weeks cramping abdominal pain, alternating diarrhoea and constipation, intermittent vomiting, 5Kg weight loss.
- No improvement on Movicol.
- Abdominal ultrasound: grossly inflamed bowel, suggestive of Crohn's disease.
- Bloods normal, but faecal calprotectin marginally raised at 98mg/g.
- When he was seen in outpatient clinic a mass was felt in the right iliac fossa.
- Urgent endoscopy assessment: ulcerated, hard, voluminous mass in the caecum causing obstruction of the bowel lumen (**Figure 1**), no other abnormalities seen.
- Histopathology (endoscopic biopsies): granulation tissue reaction, diffuse neovascularisation, acute and chronic inflammatory cells, no evidence of dysplasia, adenocarcinoma or lymphoma.
- MRI: large exophytic annular mass involving the terminal ileum and extending approximately 18cm (**Figure 2**), strongly suggestive of lymphoma, no further lesions identified.
- Successful complete surgical resection of the mass (**Figure 3**).
- Histology confirmed the suspicion of Non-Hodgkin Lymphoma in the terminal ileum.

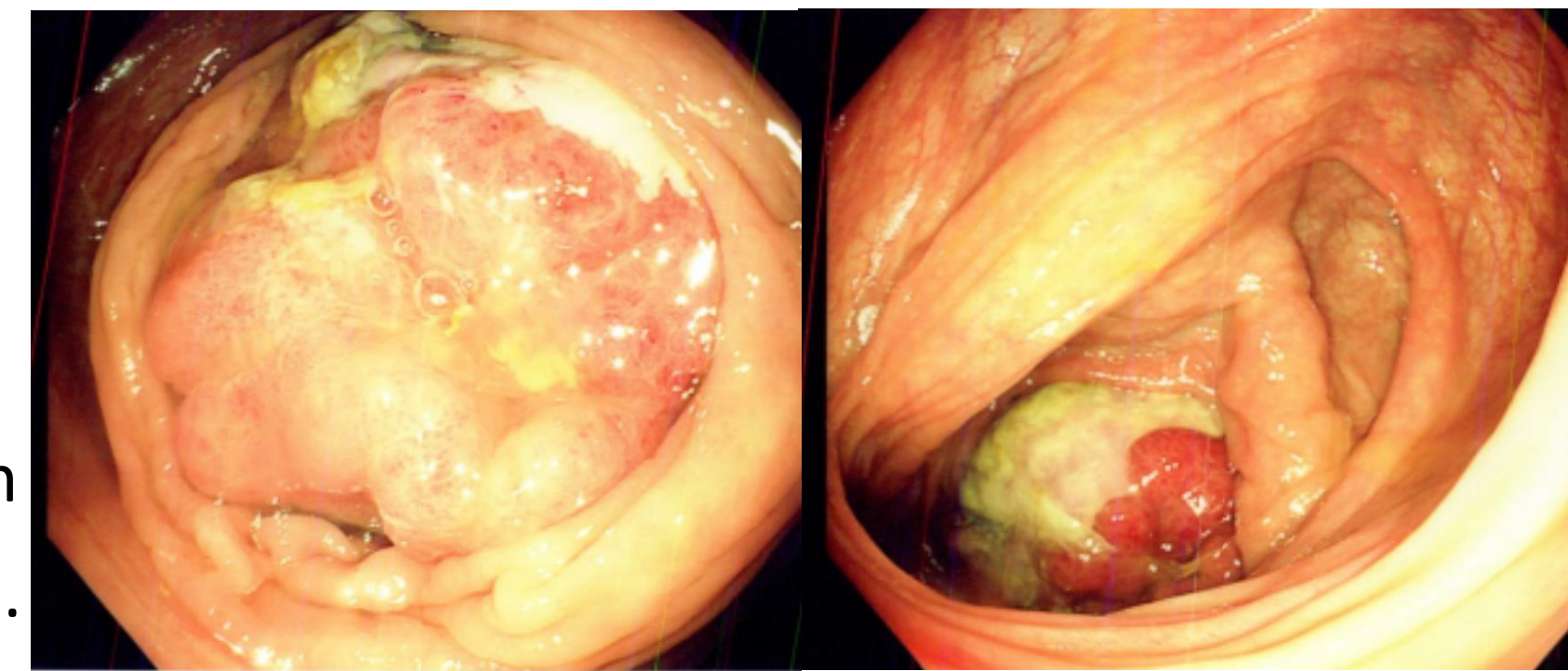


Figure 1: Pictures taken in the caecum during endoscopy showing an ulcerated mass occluding the bowel lumen

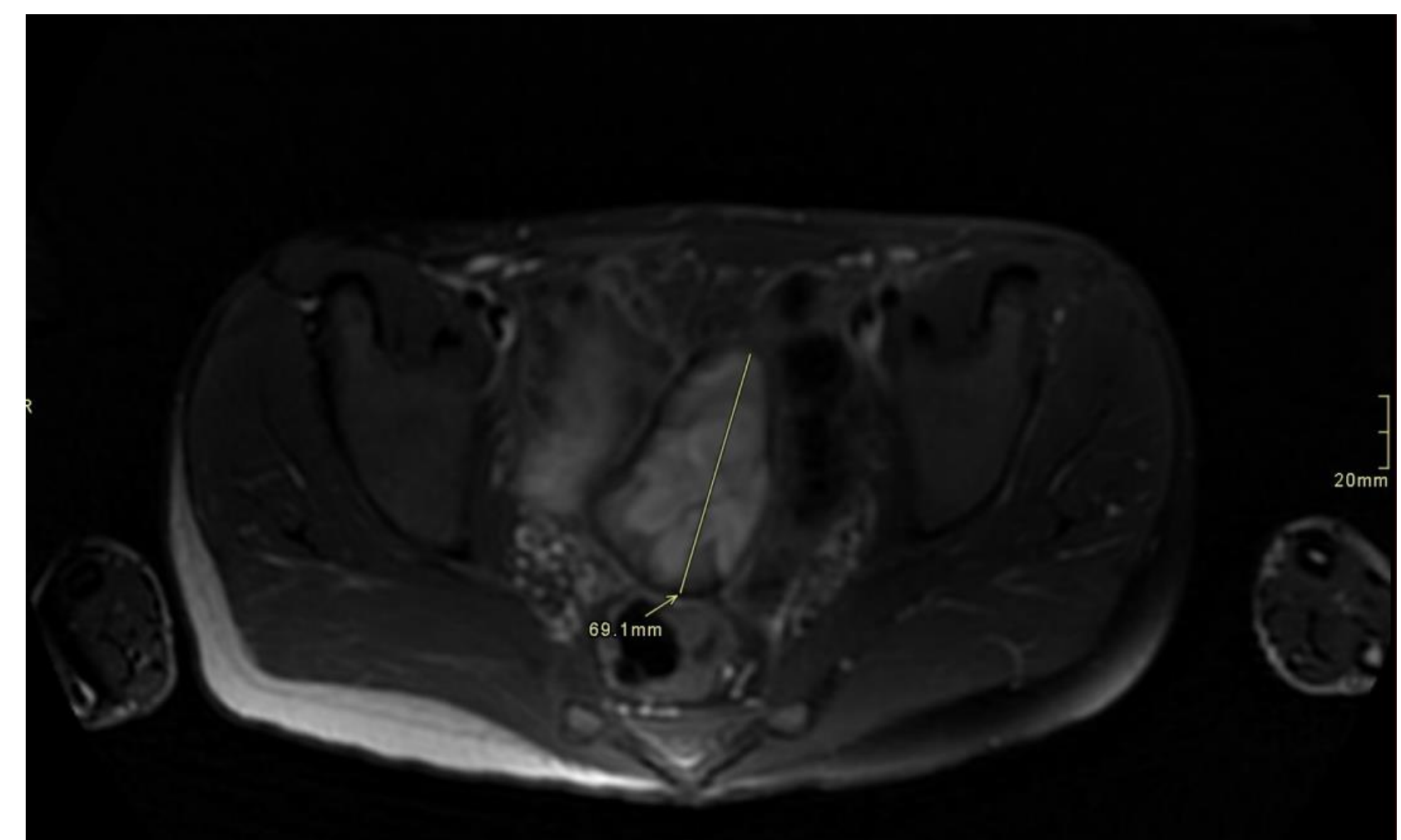


Figure 2: MRI showing a large annular mass involving the terminal ileum

Discussion

Small bowel cancer is extremely rare in children.

Primary gastrointestinal malignancy accounts for < 5% of all paediatric tumours and could be as low as 1.2%^{1,2}.

Malignancies include lymphoma (54-74%), carcinoid (3.4-16%), carcinoma (5.5-10%) and sarcoma (3.6%)^{2,3}.

Benign differentials include inflammatory masses, polyps, haemangiomas and neurofibromas.

Non-Hodgkin's lymphoma is the commonest small bowel malignancy found in children, with a high percentage of Burkitt's lymphoma.

Lymphomas are most frequently found in the terminal ileum and ileocaecal area in children and are more commonly seen in Caucasian males.

Presentation varies from palpable abdominal masses, symptoms of obstruction or abdominal pain from perforation. The tumour can be the point of ileocaecal intussusception.

Treatment is a combination of surgery, chemotherapy and radiotherapy.

Complete resection improves the outcome significantly.

Survival with primary small bowel Non-Hodgkins lymphoma is estimated at 87% at 5 years and 77% at 30 years⁴.

Conclusion

Small bowel malignancy is a rare but critical diagnosis in children.

Presentation can be insidious and can easily be mistaken as functional in nature or as a symptom of possible inflammatory bowel disease.

Early detection and intervention with surgery improves survival and therefore, although rare, malignancy should remain on the clinician's differential diagnosis list until excluded.

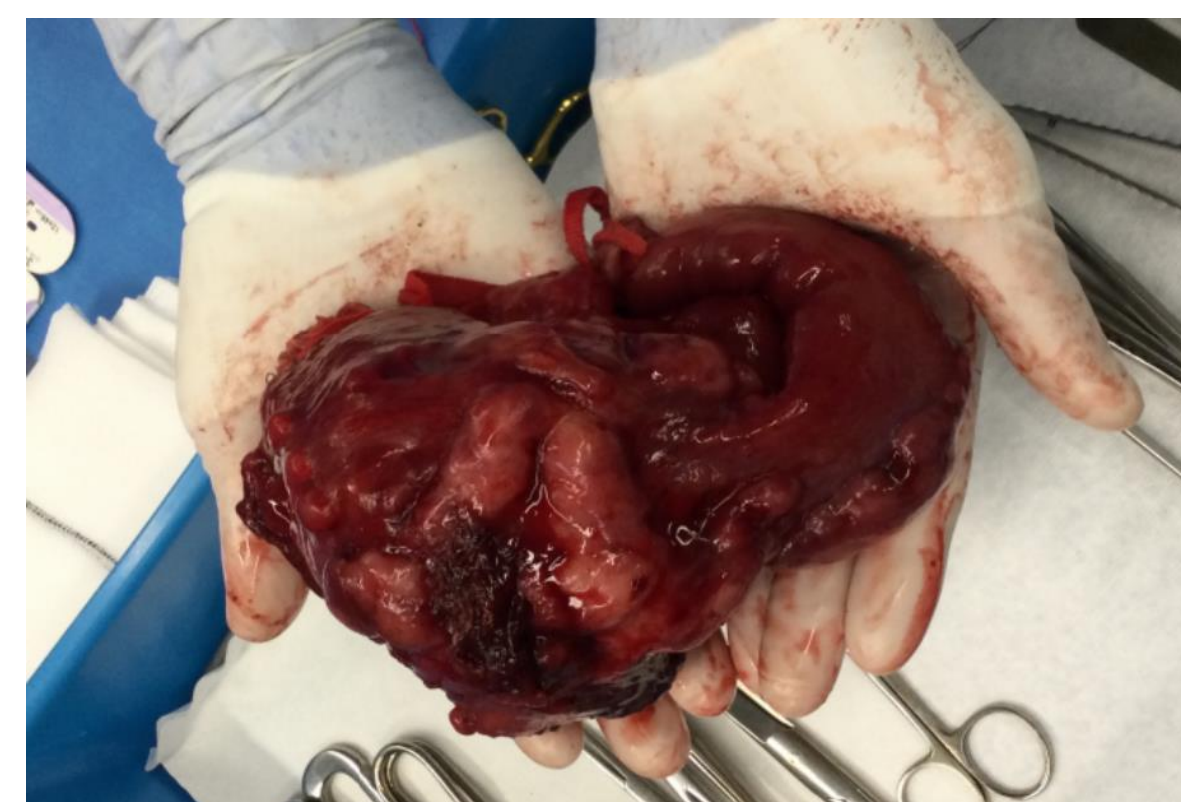


Figure 3: Resected terminal ileal mass

Key Learning Points

- A history of weight loss, as in this case, is vital and should promote early investigation with endoscopy.
- Non-Hodgkin's Lymphoma is the commonest primary small bowel malignancy in children.
- Early detection and treatment has a good prognosis.

References

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