

# Anaplastic lymphoma kinase (ALK)-positive large B-cell lymphoma: a rare cause of ileal intussusception

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Accepted 19 December 2022

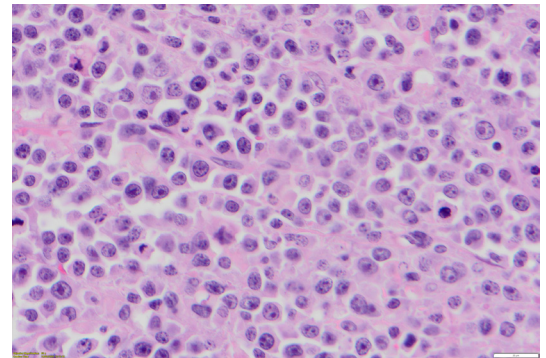
## DESCRIPTION

A man in his 60s, with no medical history of note, presented with persistent vomiting and dehydration on a background of four stone weight loss in the preceding 3-month period. CT scan of the abdomen and pelvis revealed a small bowel obstruction secondary to intussusception ([figure 1](#)). He underwent emergency surgery which revealed an intraluminal ileal mass 100cm from the ileocecal junction which was excised.

Histology of the mass showed an ulcerated, partially polypoidal tumour, the bulk of which appeared situated within the intestinal wall. The neoplasm comprised sheets of medium-to-large blasts, with moderate amounts of cytoplasm, and large rounded nuclei with granular chromatin and variably prominent nucleoli. Many showed plasmablastic differentiation ([figure 2](#)). There were frequent mitoses and apoptotic debris.

Immunohistochemical stains were positive for anaplastic lymphoma kinase (ALK) ([figure 3](#)), CD138, EMA, MUM1, CD45 (focal) and BCL2 (weak), with probable kappa light chain restriction. The cells were negative for CD20, CD79, CD30, immunoglobulin A, CD38, PAX5, CD56 and EBER. The MIB-1 proliferation index approached 100%.

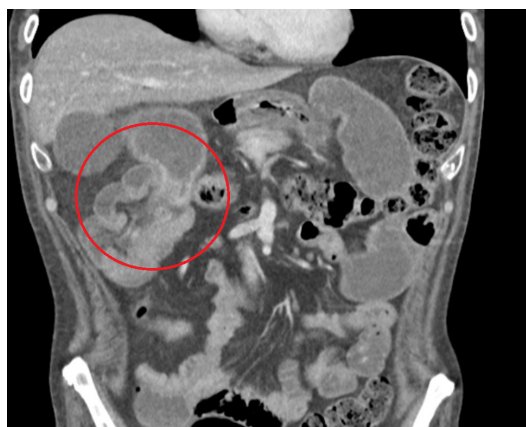
The features were consistent with ALK-positive large B-cell lymphoma (ALK+LBCL) which is a very rare and aggressive neoplasm accounting for less than 1% of all diffuse LBCLs.<sup>1</sup> This tumour most frequently occurs in the lymph nodes, however extranodal site involvement has also been described in the nasopharynx, soft tissue, bone and gastrointestinal tract.<sup>1,2</sup> ALK gene rearrangements,



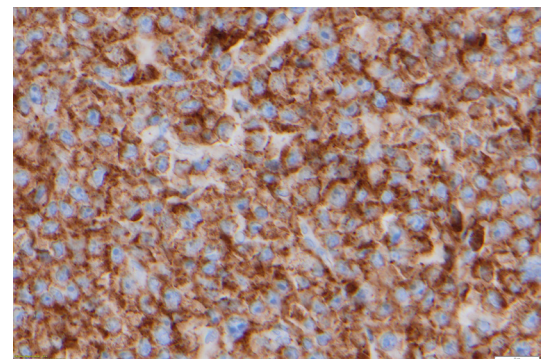
**Figure 2** Histology section with H&E stain at ×600 magnification showing sheets of medium-to-large blasts, with immunoblastic and plasmablastic differentiation.

most commonly t(2; 17)(p23;q23), lead to ALK protein overexpression—a key event in the oncogenesis of this tumour.<sup>1</sup> The vast majority of these lymphomas show immunoblastic and or plasmablastic morphology, with characteristic immunohistochemical staining for ALK, EMA and plasma cell markers, such as CD138 and MUM1.<sup>1</sup> They are negative for T-cell markers, CD30 and B-cell lineage associated antigens such as CD20, meaning they are unlikely to respond to rituximab.<sup>1</sup> ALK+LBCL usually responds poorly to conventional chemotherapeutic regimens, however new drugs, especially ALK inhibitors, show future promise.<sup>2</sup>

Our patient's staging CT scan<sup>3</sup> showed no evidence of distant lymphadenopathy. Although staging bone marrow showed a low population (0.2%) of cells with a possible ALK+LBCL immunophenotype, overall there was a lack of distinctive morphological features to confirm involvement. He was treated with cyclophosphamide, vincristine,



**Figure 1** CT image of the patient's abdomen at presentation (coronal view) showing lead point of intussusception with characteristic 'telescoping' of small bowel (circled).



**Figure 3** Immunohistochemical stain at ×600 magnification showing characteristic diffuse tumour cell positivity for anaplastic lymphoma kinase.



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**To cite:** O'Hare EM, Thekkinedath A, Ngu L, et al. *BMJ Case Rep* 2022;**15**:e253239. doi:10.1136/bcr-2022-253239

## Images in...

doxorubicin and prednisolone (CHOP) chemotherapy, without rituximab.<sup>4</sup> Positron emission tomography scan after completion of six cycles of CHOP chemotherapy showed no residual disease activity, implying complete metabolic remission. At the time of writing, almost 10 months after his initial presentation, our patient has had no episodes of relapse and remains on regular follow-up with the haematology team.

## Learning points

- ▶ Anaplastic lymphoma kinase+large B-cell lymphoma is a very rare and aggressive lymphoma with characteristic morphology and immunophenotype.
- ▶ These tumours are usually negative for CD20 antigen and are thus unlikely to be responsive to rituximab.

**Acknowledgements** The authors would like to thank Mr Samuel J. Radcliffe for his advice on the final draft.

**Contributors** EMO is the lead author of the paper. AT contributed clinical information about the case. LN is the consultant haematologist who managed the care of the patient. EMO and PM are consultant pathologists who reported the original histology and reached the correct diagnosis. Both LN and PM provided critical appraisal of the final article.

**Funding** The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

**Competing interests** None declared.

**Patient consent for publication** Consent obtained directly from patient(s).

**Provenance and peer review** Not commissioned; externally peer reviewed.

Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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