CLINICAL CASE - TEST YOURSELF | Paediatric Radiology

Abdominal pain in a 7-year-old male

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PART A

A 7-year-old male presented with abdominal pain of one day duration. An ultrasound (US) diagnosis reported atypical ileocolic intussusception and adjacent hypoechoic nodes, suggesting a possible lead point. Computed Tomography (CT) scan was performed for further investigation (**Figs. 1-2**). Following a failed fluoroscopic reduction, an ileocolic intussusception was surgically reduced without visible or palpable external macroscopic bowel abnormality. Biopsy of a mesenteric node came back as hyperplasia. Patient's postoperative US by an inexperienced operator was unrevealing. Four months later the patient presented with abdominal distention and palpable abdominal masses. A repeat CT scan was performed (**Figs. 3-6**).

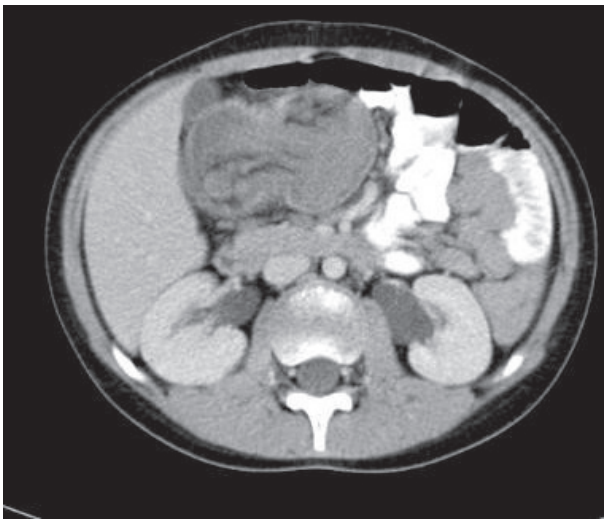

Fig. 1. *Axial CT scan at the mid-abdomen.*

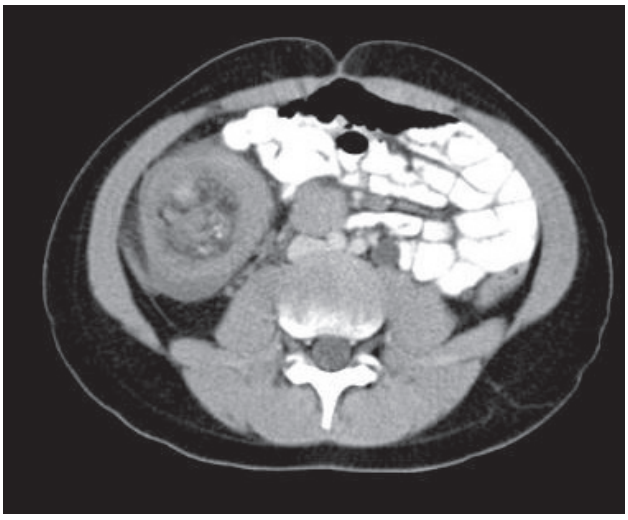

Fig. 2. *Axial CT scan at the upper pelvis.*

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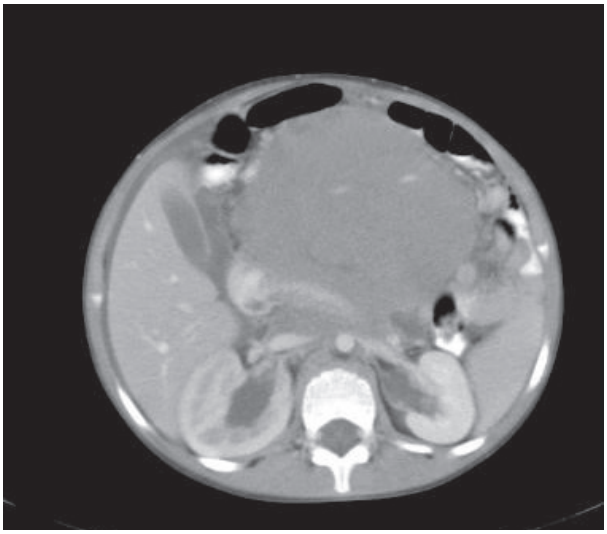

Fig. 3. *Repeat CT scan at the same level as in Fig. 1.*

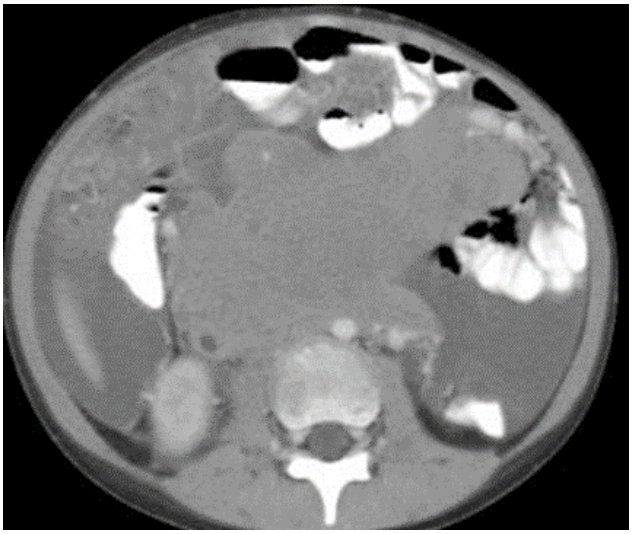

Fig. 4. *Repeat CT scan at the mid abdomen.*

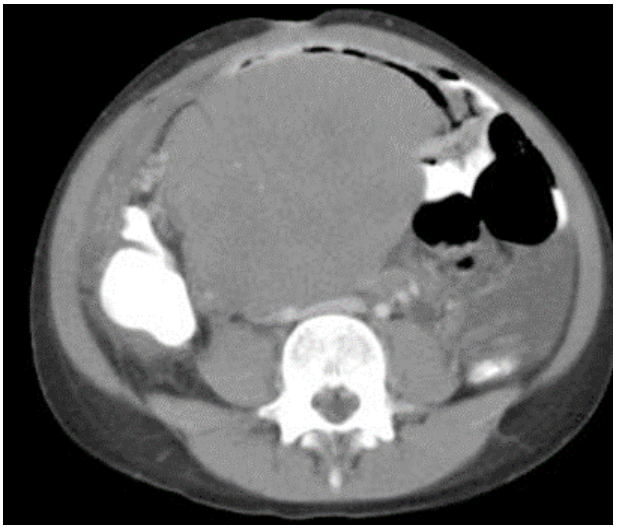

Fig. 5. *Repeat CT scan at approximately the same level as in Fig. 2.*

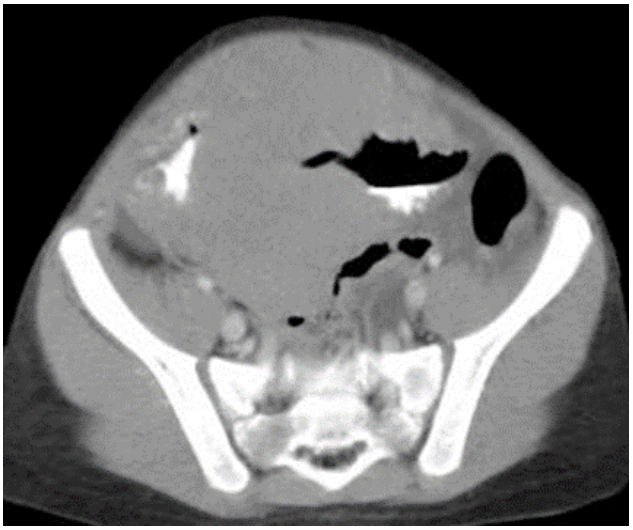

Fig. 6. *Repeat CT scan at the right iliac fossa.*

PART B

DIAGNOSIS: Peritoneal lymphomatosis following reduction of a secondary intussusception

The initial CT disclosed an enteric mass with trapped mesenteric fat, vessels and lymph nodes consistent with intussusception (**Figs. 1, 2**). A definite lead point could not be seen at US or CT, probably because it might have been a flat small lesion. On initial US, large hypoechoic adjacent nodes and age were suggestive of additional pathology. CT also disclosed lymphadenopathy (**Fig. 2**). Intussusception was irreducible and a lead point was not confirmed, neither on radiology or during surgical palpation.

Intussusception in children is usually idiopathic, occurs most commonly in infants aged 5-9 months, with the majority of cases occurring by the age of 1 year [1]. Children younger than 4 months and older than 2-4 years should be investigated for secondary intussusceptions [1]. Commonest lead points include Meckel's diverticulum, duplication cysts, polyps or tumours such as lymphoma [1, 2]. These should be ultrasonographically examined at the base of the intussusception. In our 7-year-old patient, biopsies of all layers of the bowel and of the largest node, which might have led to an earlier diagnosis, were not performed. The postoperative US by an inexperienced operator did not show free fluid in the abdomen. A biopsy and a meticulous US technique with linear transducers might have disclosed hypoechoic thickened ileal wall and enlarging nodes earlier.

Repeat CT scan showed a large intraperitoneal, mesenteric and enteric mass, complicated with ascites and associated with omental thickening. Enhancing peritoneal micronodular deposits, consistent with peritoneal dissemination were visible (**Fig. 3-6**). Biopsy revealed Burkitt's lymphoma.

Post-biopsy course was uncomplicated. The patient received chemotherapy according to the protocol BFM B-NHL 04 and completed this treatment 5 months later. Due to residual tumour (MRI and PET-CT scan positivity), the patient underwent 3 courses of chemotherapy of R-EPOCH, followed by complete surgical removal of small residual tumour. Therapy was intensified by 2 courses R-ICE chemotherapy, high dose BEAM and autologous transplantation from peripheral stem cells. Patient is still followed-up annually, and is disease free 9 years later.

Primary intraperitoneal solid tumours are uncom-

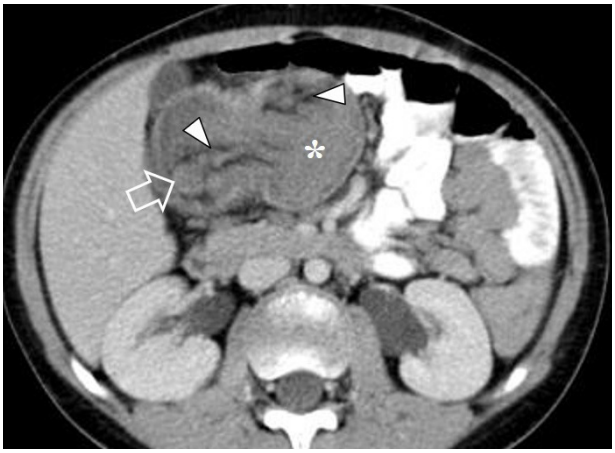

Fig. 1. *CT scan shows an enteric mass with trapped mesenteric fat (arrowheads) and a trapped lymph node (open arrow). Findings are consistent with ileocolic intussusception. The prominent intussusceptum (*) is abundant. This appearance is encountered in both engorged proximal telescoped bowel (intussusceptum) and in small tumours.*

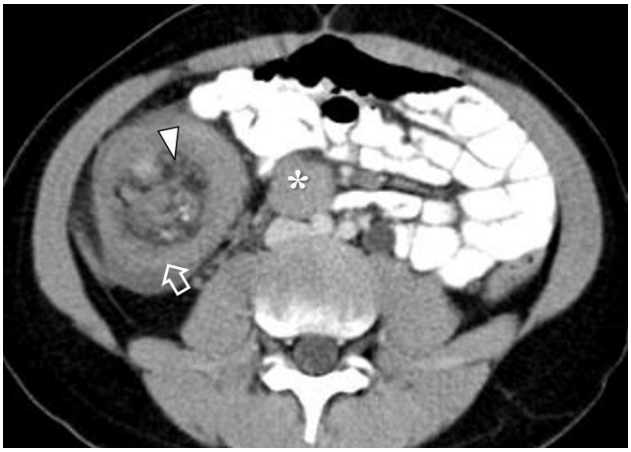

Fig. 2. *CT scan shows a right inferior fossa enteric mass exhibiting the classic "donut" sign with trapped mesenteric fat (arrowhead) and smooth wall thickening (open arrow) due to the apposition of the wall of intussusceptum and intussuscipiens. There is a round node anterior to the bifurcation of the aorta (*).*

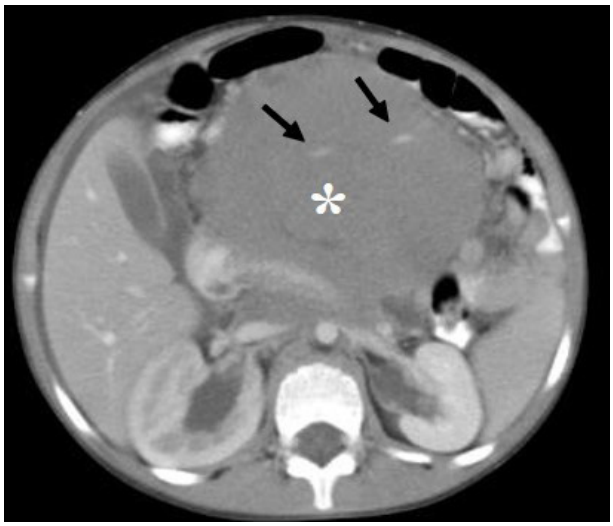

Fig. 3. *Repeat CT scan at the same level as in Fig. 1. There is a large solid mesenteric mass (*) displacing the bowel loops and completely encasing the mesenteric vessels (arrows).*

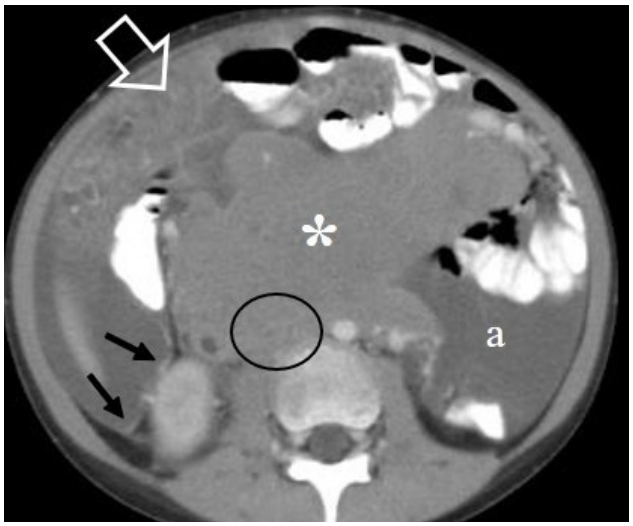

Fig. 4. *A large mesenteric mass (*) is associated with extensive omental thickening (open arrow), ascites (a), and obliteration of the lumen of inferior vena cava (within circle). Note enhancing peritoneal reflections with tiny enhancing focal micronodules (black arrows).*

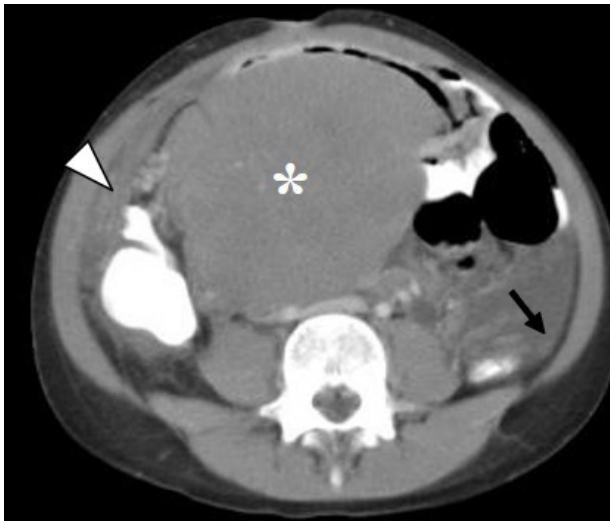

Fig. 5. *Repeat CT scan approximately at the same level as in Fig. 2. The node has evolved into a huge mass at the inferior mesentery (*). Note omental thickening on the right (arrowhead) and an enhancing nodular peritoneal reflection (black arrow).*

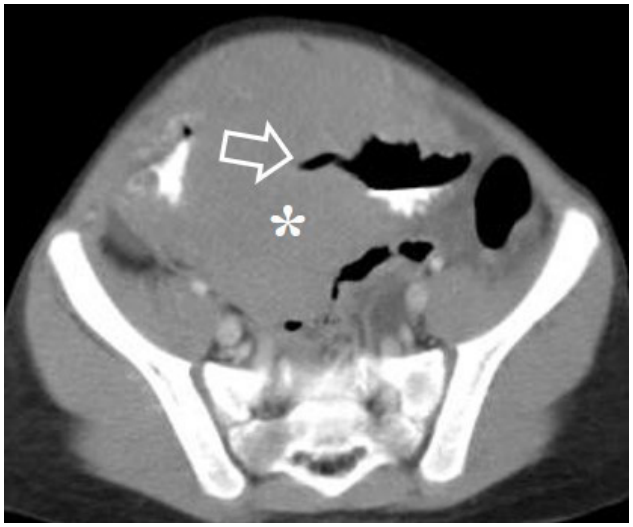

Fig. 6. *CT scan at the pelvis shows the lower aspect of the mass (*) encasing the ileum which shows a massive wall thickening. The mucosal surface of the bowel is ragged (open arrow) and the lumen is dilated without proximal obstruction.*

mon in children compared to adults and carry different histologic diagnoses. Diffuse peritoneal disease may be due to desmoplastic small round cell tumour (DSRCT), rhabdomyosarcoma, or non-Hodgkin lymphoma (NHL) [3]. Peritoneal metastases causing diffuse peritoneal disease in children are due to ovarian germ-cell tumours, lymphoma, intraperitoneal spread of an intracranial tumour via a vetriculo-peritoneal shunt or of a retroperitoneal tumour like neuroblastoma and Wilms' tumour [4-6]. Our patient was a male with no history or evidence of an intracranial or retroperitoneal tumour.

Lymphomas which may spread into the peritoneum are usually NHL. In children with NHLs, extranodal clinical presentation is much more common than in adults, the most common locations being intraabdominal and intrathoracic [4]. These children may frequently present with symptoms mimicking acute appendicitis and with intussusception caused by intraluminal projection of a small tumoural mass. Burkitt's lymphoma is the most frequent subtype of NHL in childhood and accounts for approximately 34% of these cases [7]. Peritoneal lymphomatosis can be seen as an initial presentation of lymphoma, it may develop as the disease progresses or even as a manifestation of transformation of an indolent subtype of lymphoma to a higher grade, most commonly diffuse large B-cell lymphoma [8]. Small bowel lymphoma manifests as circumferential tumoural spread throughout the intestinal submucosa, progressively infiltrating the bowel wall [7]. Multifocal involvement is not uncommon. The lumen may be narrowed because of mucosal infiltration and excavation or may exhibit dilatation, due to replacement of the muscular layers by tumour spread and infiltration of the myoenteric plexus [4]. Nodal involvement and extranodal extension may produce loss of definition of individual nodes within a confluent mass as occurred herein [4].

DSRCT was the main differential diagnostic possibility to peritoneal lymphomatosis in our case. DSRCT is an aggressive neoplasm of the paediatric small round blue cell tumour family, which also includes Wilms tumour, neuroblastoma, Ewing sarcoma, primitive neuroectodermal tumour, and rhabdomyosarcoma [3]. It is a rare childhood multifocal peritoneal malignancy with frequently disseminated abdominal disease at presentation [9]. It predominantly affects boys and young adults. When encountered in females, the tumour can be mistaken for ovarian cancer. In the abdomen, the disease most commonly involves the omentum and peritoneum, followed by the retroperitoneum. Typical findings of DSRCT are multiple peritoneal, omental, and serosal masses without a clear organ of origin ranging in size from 3 to 22 cm, with a single dominant mass situated at the retrovesical/retrouterine space, the peritoneum or omentum [3, 9]. Occasionally there is necrosis or calcification, whereas a substantial number of patients have diaphragmatic involvement [3, 9]. In our patient the dominant mass was mesenteric.

In adolescents and adults, the peritoneal surface may be secondarily affected by epithelial cell lines resulting in carcinomatosis, by mesenchymal cell lines resulting in sarcomatosis and by lymphoid cell lines resulting in lymphomatosis [8]. Although these entities share similar radiologic features, there are some contributory findings that may support the final diagnosis of lymphoma. This is extremely important since lymphomas are primarily treated with chemotherapy and not surgery [8]. Enhancing peritoneal nodules and omental deposits tend to occur in peritoneal carcinomatosis. However, epithelial neoplasms are less common in children compared to adults. Large mesenteric masses are more in favour of lymphoma and sarcoma. In our case, the homogeneity of the mass, the pattern of circumferential excessive wall thickening of the ileum and the history of a recent and possibly secondary intussusception were in favour of peritoneal lymphomatosis.

Over the past decades, the outcome of children with malignant lymphoma has improved, with long-term

KEY WORDS
Secondary intussusception; Peritoneal lymphomatosis; Child; CT

event-free survival rates of >90% in childhood Hodgkin's disease and >80% in childhood NHL [10]. Current treatment for malignant lymphoma aims at maximising the chance of cure, while minimising early and late toxicity such as infertility, premature menopause, cardiac disease, and most importantly, risk of second neoplasms [10]. Our event-free survivor supports these outcomes.

This case emphasises awareness of the possibility of secondary intussusception in older children, the need of communication between subspecialties and the need of performance or supervision of US tests by experienced radiologists. It also illustrates the differential diagnosis of peritoneal spread of tumours in children. **R**

Conflict of interest

The authors declared no conflicts of interest.

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