

An unusual case of a retroperitoneal cyst in a child

Niamh Grayson, Muhammad Younis, Nigel Heaton

CASE REPORT

A previously well 9-year-old boy presented with intermittent left upper quadrant pain, anorexia, and weight loss over six months. His past medical history was significant for being born preterm at 34 weeks' gestation and requiring ventilatory support as a neonate. Computed tomography (CT) scan (Figures 1 and 2) showed a 10 cm encapsulated spherical mass appearing to arise from the pancreatic tail. The working diagnosis was a cystic pancreatic tumour, and he was referred for further management.

At subsequent laparotomy a retroperitoneal mass was found adherent to the left colon and duodenum but free from the pancreas, with associated reactive lymphadenopathy. The mass was biopsied, and surgery was abandoned to allow for formal histological evaluation. Frozen section histology reported a spindle cell lesion comprising of bland spindle cells with elongated cigar-shaped nuclei. Necrosis was present at one end of the core biopsy. Mitotic activity was not seen. Immunohistostaining showed spindle cells expressing smooth muscle actin, desmin, and vimentin. There was no significant expression of S100, CD117, CD34, or AE1/AE3. There was no nuclear or cytoplasmic beta catenin expression. Up to three IgG4 expressing plasma cells were seen per high power field.

Further work-up included a computed tomography positive emission tomography (CT PET) scan which showed a non-avid mass appearing to arise from the tail of the pancreas. Computed tomography-guided biopsy showed degenerate and viable red cells. No other cell type was identified to help determine the nature of the

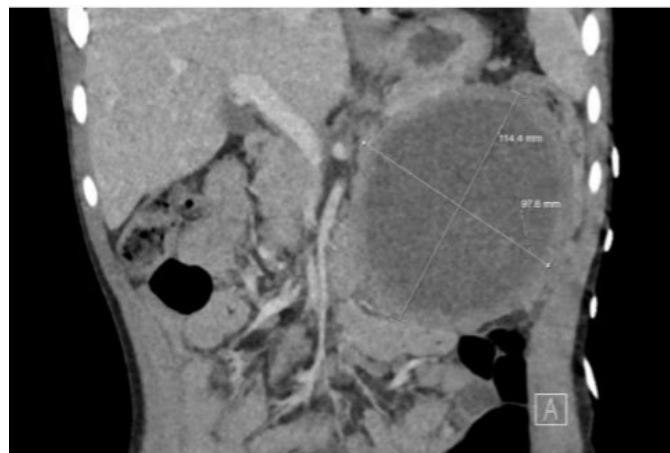


Figure 1: CT coronal view 114 mm × 97 mm retroperitoneal cystic mass displacing the left colon, pancreas, and small bowel.



Figure 2: CT axial view showing the cyst and its relations to the left kidney and vasculature.

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lesion. A decision was made to perform local resection as no malignancy was identified. At surgery the lesion was excised locally without compromising adjacent pancreas, colon, or small bowel. He recovered well post-operatively and was discharged home on day 4. Final histology showed a 130 mm encapsulated thick-walled organized hematoma with no evidence of malignancy.

DISCUSSION

Retroperitoneal cysts are rare with few reports in children. A literature review of PubMed was performed using the search terms retroperitoneal cyst in children and pediatric population. Articles not available in English, pancreatic cysts and patients over age 18 were excluded. The results are summarized in Table 1 [1–10].

The differential diagnosis includes cysts of lymphatic origin and lymphangiomas, benign or malignant mesothelial cysts, enteric cysts, cysts arising from a urogenital origin, dermoid cysts and cysts with traumatic, hemorrhagic or infectious etiology [1].

The reported ages of patients varied from 10 months old to 15 years old with a median age of 9 years old for those included.

Table 1: Previously described cases of retroperitoneal cysts [1–10]

Author	Year	Country	Age in years (if stated)	No. of cases	Imaging modality	Characteristics on imaging	Site	Presentation	Treatment	Histology
Hebra et al. [3]	1959–1990	USA		3				2 Asymptomatic 1 Acute abdominal pain	1 complete excision 2 partial excision	3 Lymphangioma
Nam et al. [6]	1998–2010	South Korea	–	10				5 abdominal mass 1 abdominal pain 3 prenatal imaging 1 incidental finding	7 excision 1 partial excision and OK432 injection 2 biopsy	7 Lymphangioma 3 Pseudocyst
Ferrero et al. [2]	2013	Italy	7	1	Ultrasound MRI	2 non-enhancing cystic lesions	Right abdomen	Palpable mass	Excision	Cystic lymphangioma
Wilson et al. [9]	2006	USA	10	1	Ultrasound CT	Multiseptated mass	Right abdomen	Abdominal pain and distension	Excision	Lymphangioma
Pachl et al. [7]	2011	UK	10 months	1	Ultrasound	Thick-walled cyst	Superior to left kidney	Prenatal imaging	Excision	Enteric duplication cyst
Zhang et al. [10]	2015	China	8	1	Ultrasound	Homogenous mass with mucoid contents	Left retroperitoneum	Incidental finding on imaging	Excision	Bronchogenic cyst
Hyseni et al. [4]	2009	Kosovo	4	1	Ultrasound CT	Thin-walled multilocular cystic mass with internal septations	Right retroperitoneum	Abdominal pain and distension	Excision	Epidermoid cyst
McCrystal et al. [5]	2002	Australia	8	2	Ultrasound	Quadrilateral mass with well-defined capsule	Superior to left kidney	Acute abdomen	Excision	Bronchogenic cyst
			15		CT	Calcified cyst	Superior to left kidney	Left-sided thoracic pain	Excision	Bronchogenic cyst
Deng et al. [1]	2019	China	10	1	Ultrasound CT	Thick walled cyst connected to pancreatic duct	Left upper quadrant	Abdominal pain	Excision	Enteric duplication cyst
Waldhausen et al. [8]	1996	USA	9	2	Ultrasound CT	Large cystic mass	Right upper quadrant	Abdominal pain	Excision	Lymphangioma
			9		Ultrasound CT	Complex mass fluid filled multiple septations Cystic mass	Retroperitoneum	Abdominal mass	Excision	Lymphangioma

The presenting symptoms are non-specific and varied with abdominal pain and distension being the most common. Other presentations included nausea and vomiting, anorexia, acute abdomen, and incidental detection on examination or on imaging.

Investigations varied from case to case. Ultrasound was the most commonly used modality across all of the patients.

Diagnosis can be difficult. The literature review confirmed that the majority of cases were diagnosed as retroperitoneal cysts at operation. On imaging, the cyst

appeared to be associated with the tail of the pancreas but at operation, it was clearly separate and in our case, there was concern that it could malignant. Frozen section revealed spindle cells within the wall and it required formal immunohistostaining and repeat biopsy to exclude malignancy.

Retroperitoneal cysts are treated surgically. Acutely there is a risk of cyst rupture, infection, and hemorrhage. They may cause abdominal pain by compression of surrounding organs [1]. In this case, complete excision was achieved without complication.

CONCLUSION

1. Retroperitoneal cysts are a rare entity with a wide diagnosis. Here we report a non-traumatic hemorrhagic cyst which has not been reported previously and should be included in the differential diagnosis.
2. The presenting symptoms of retroperitoneal cysts vary widely from patient to patient in this literature review.
3. This patient underwent a long diagnostic process which we hope to avoid with documentation of this case.

Keywords: Abdominal pain, Hemorrhagic cyst, Retroperitoneal cyst

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Author Contributions

Niamh Grayson – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Muhammad Younis – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Nigel Heaton – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

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Conflict of Interest

Authors declare no conflict of interest.

Data Availability

All relevant data are within the paper and its Supporting Information files.

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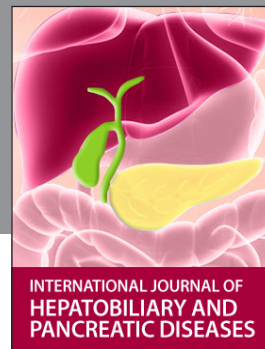
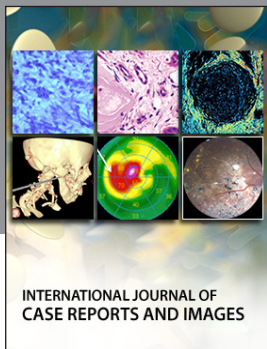
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