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Case Report

Case report: Ballotable abdominal mass in a child – Definitely renal in origin?

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ABSTRACT

Introduction and importance: The differential diagnosis of a paediatric abdominal mass can be extensive, as it potentially involves multiple organs including gastrointestinal, genitourinary, endocrine, and gynaecological systems. Hence, a systematic approach to history taking and physical examination is needed to clinch the diagnosis. Specifically, the approach for assessing, investigating, and managing a ballotable left hypochondrial mass in a child can be challenging.

Case presentation: We report a 10-year-old Dusun girl presenting with left hypochondrial pain and noted a left hypochondrial mass on examination. This report highlights the role of clinical imaging during the pre-operative and post-operative phases.

Clinical discussion: Ultrasound and CT imaging was useful in determining that the tumor originated from the tail of the pancreas. The presence of a definite capsule with internal solid-cystic components helped narrowed the differential diagnosis to solid pseudopapillary neoplasm (SPN) of the pancreas. MR liver was useful to rule out liver metastasis in this child.

Intervention and outcome: The patient was scheduled for laparotomy and tumour excision at a regional paediatric centre. Successful excision of the tumor en-mass was performed and the child's subsequent recovery was uneventful.

Conclusion: Clinical imaging plays a critical role in the diagnosis and management of paediatric solid organ tumours. Other than renal origin, suspicion of pancreatic tail origin should be considered by clinicians when encountering a ballotable left abdominal mass.

1. Introduction

An unusual abdominal mass in a young child always raises the clinical suspicion of tumour – benign or malignant. The differential diagnosis of a paediatric abdominal mass can be extensive, as it potentially involves multiple organs including the gastrointestinal, genitourinary, endocrine, and gynaecological systems. Hence, a systematic approach of history taking and physical examinations is needed to clinch the diagnosis.

Bimanual palpation is a useful examination technique to assess abdominal masses. It gives the clinician information regarding the surface characteristics of the mass, its consistency, and mobility. The ability of the abdominal mass to move with gentle pressure on bimanual palpation is described as ballotable [1]. Typically, renal masses are described as ballotable. Less commonly, gastric or pancreatic tail masses can also be ballotable [2–4]. However, it is important to note that the normal kidney can be ballotable in slim individuals.

Pancreatic tail tumours are rare in children. These masses are often misdiagnosed as neoplasms arising from the kidney or the spleen at the initial work-up stage. Oncologic and surgical treatment of various solid organ tumours in children differs greatly, depending on the origin, histology, and extent of the disease. Hence, a comprehensive approach incorporating further biochemical, radiological, and histopathological examinations is utilised to obtain a definite diagnosis with disease staging.

We present a case of a solid pseudopapillary neoplasm (SPN) arising from the pancreatic tail presenting as a ballotable left hypochondrial mass in a child that highlights the importance of clinical imaging in the

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diagnosis and further management of this rare tumour. We report this case in line with the SCARE 2020 criteria [5].

2. Case presentation

A 10-year-old Dusun girl with no known medical illness and surgical history, presented with dull-aching left hypochondrial pain for a one-week duration. There was associated recent loss of appetite but no significant loss of weight. There was no history of trauma nor was there any haematuria in the past. No relevant family history of medical illnesses was elicited. There was no significant drug history or allergies. The pain became progressively more severe and the child was brought by the parent to the hospital. On admission, physical examination revealed a non-tender, ballotable mass in the left hypochondrium which extended to the left lumbar region, with a smooth surface. This raised the clinical suspicion of left renal mass.

Initial blood investigation noted lactate dehydrogenase (LDH) was slightly elevated. Renal function was found to be normal. The rest of the blood parameters, including amylase and tumour markers (alpha feto-protein and beta human chorionic gonadotropin) were unremarkable.

Ultrasound abdomen also revealed a large heterogenous solid-cystic left retroperitoneal mass and a solitary segment VI hypoechoic liver lesion. Both kidneys were normal in size and appearance with no suprarenal mass. Subsequent contrast-enhanced thoracic, abdominal, and pelvic computed tomography (CT) examination was performed to further evaluate the origin, nature, and extent of the mass. Multiplanar CT scan showed a large enhancing solid-cystic mass with distinct capsule arising from the pancreatic tail and a solitary liver lesion (Fig. 1 & Fig. 2). A radiological diagnosis of pancreatic tail SPN was given. A differential of pancreatoblastoma was also considered, as it may have a similar radiological presentation. Ultrasound-guided percutaneous biopsy was performed and the histology was reported as SPN.

The patient's clinical imaging and histopathological findings were reviewed by a multidisciplinary paediatric oncology team. Given the large size of the mass and worsening symptoms, the patient was scheduled for early laparotomy and tumour excision. However, the operation was delayed for 2 weeks as the patient was treated for COVID-19 infection after undergoing routine pre-operative screening.

The surgery was performed under general anaesthesia at a regional oncology center. The operation was conducted by a senior paediatric surgeon with more than 5 years of post-registration experience. Intra-operatively there was a large well encapsulated, lobulated mass arising from retroperitoneum, occupying the left upper abdomen and crossing midline. It measures 15.0 × 10.0 × 8.0cm in size. The tumour is closely adhered to the pancreatic tail, sharing the outer layer (Fig. 3). It abuts the spleen laterally and pushing the spleen superiorly, stretching the splenic vein. Tumour weighing 450 grams was removed en-mass along with a cuff of pancreatic tail tissue (Fig. 4).

Histopathological examination of the surgical sample confirmed the diagnosis of SPN of the pancreas. Immunohistochemistry shows that the tumour cells are diffusely positive for Beta-catenin and CD56, focally positive for synaptophysin, and negative for chromogranin.

By day three post-operation, the patient was able to tolerate clear fluid orally. Day three wound inspection was clean. The patient was discharged on the fifth-day post-operation without complications.

Upon follow-up in the clinic two weeks later, the patient was well and tolerating a normal diet. Physical examination was unremarkable with good progress of wound healing. Postoperative magnetic resonance imaging (MRI) abdomen revealed that solitary liver lesion is suggestive of a liver haemangioma with no evidence of local recurrence (Fig. 2).

3. Discussion

SPN is a rare pancreatic neoplasm with low malignant potential. This rare tumour accounts for not more than 2% of all exocrine pancreatic tumour in most of the reported literature. This tumour was first identified by Virginia Frantz as a heterogeneous group of solid pancreatic neoplasms with benign or malignant features in 1959 [6]. Later in 1996, the tumour was defined by the World Health Organization (WHO) as "solid pseudopapillary tumours" and reclassified them as SPNs in 2010 [7].

Approximately 90% of cases involve young Asian and African-American women with the mean age of presentation at 22 years old [8,9]. A rare occurrence of cases in children and men have been reported as well. There has been postulation regarding the role of sex hormone in etiopathology of this neoplasm in view of high predilection of cases in female [9]. However, the origin of these neoplasms is still yet to be clarified. Over years, it has been recognised with increasing frequency owing to widespread use and improvement of cross-sectional imaging.

Patients with early SPN experience no symptoms. It is usually diagnosed incidentally during the consultation of other medical problems or even during a routine medical check-up. The tumour can reach a considerable size before starting to cause symptoms. Abdominal pain is the most common presenting symptom followed by the presence of a felt upper abdominal mass. SPNs most commonly arise from the pancreatic head (34–40%) or the pancreatic tail (24–36%) [4].

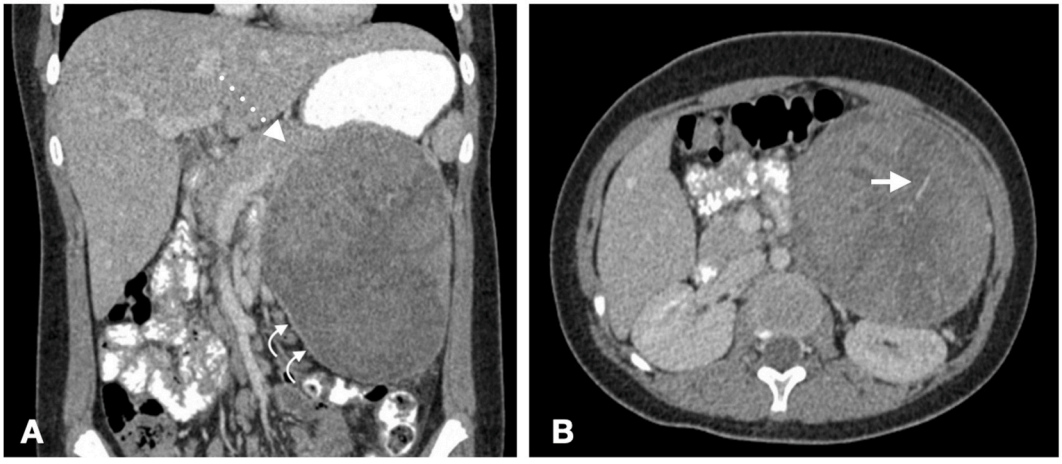

Fig. 1. (A) Coronal section of a contrast enhanced CT abdomen showing a large well-defined, heterogeneously enhancing mass arising from the pancreatic tail, demonstrating a 'claw-sign' suggesting its origin (dotted white arrow). The mass has internal solid-cystic components and is circumscribed by a distinct capsule (curved arrows). No internal calcification within. (B) Axial section of a contrast enhanced CT abdomen showing opacified blood vessels seen within the mass (solid white arrow).

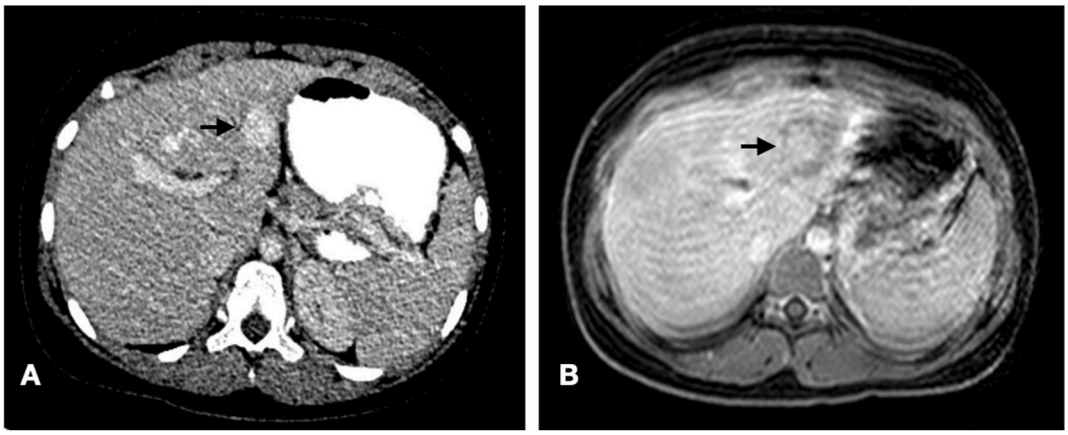

Fig. 2. (A) Enhancing segment III liver lesion noted in CT scan (black arrow). (B) MRI liver dynamic post-gadolinium sequence revealed avid contrast uptake and persistent central enhancement in delayed images (black arrow). Motion artefacts were present in the delayed phase images as the child could not tolerate the scan much longer.

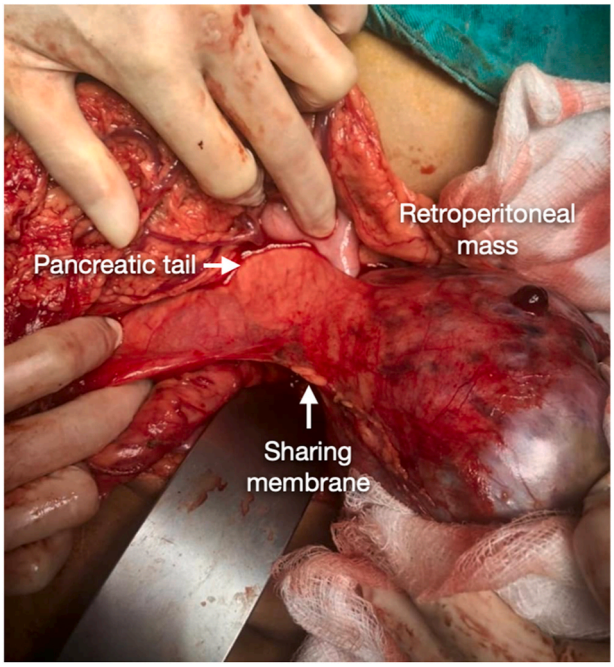

Fig. 3. Intraoperatively noted a retroperitoneal mass sharing outer membrane with pancreatic tail.

Apart from the abdominal mass, the rest of the clinical examination is often normal. A mass becomes evident and easily palpable when the average size of the tumour reaching 8–10 cm in diameter [8]. Consequently, the physical examination is invariably negative until the tumour enlarges to a size significant enough for palpation. To date no tumour marker that is specific to SPNs [8].

During the initial work-up for this particular patient, ultrasonography revealed a retroperitoneal mass with heterogeneous echogenicity with a solitary hyperechoic liver lesion. However, it was not possible to ascertain the origin of the tumour from ultrasound imaging. Cross-sectional imaging helped clarify the origin of the mass from the pancreatic tail. Intra-operatively, this connection between the mass and the pancreatic tail was identified to facilitate complete resection of the tumour. Axial CT images demonstrate the position of the pancreatic tumour which overlies the kidney, which transmits the posterior palpating force from the flank to the anterior skin surface.

Although most of SPNs exhibit indolent clinical course and demonstrate low malignant transformation potential, approximately 10%–15% of cases exhibit aggressive clinical and pathologic features in keeping with metastases [10]. These SPNs with malignant transformation, often affecting older men, are classified as solid pseudopapillary carcinomas (SPC). The liver is most commonly involved in SPN metastases, followed by the neurovascular sheath and lymph node metastases [9]. In our patient, the decision to proceed with laparotomy was based on the worsening pain and discomfort. Post-operatively, MR liver was useful to further assess the liver lesion and to rule out metastasis. The dynamic imaging findings of delayed enhancement is characteristic of liver haemangioma.

Differentiating benign SPN from malignant SPC remains a challenging diagnostic problem. Radiologically, SPNs are typically reported as well-circumscribed pancreatic masses that exhibit variable degrees of internal haemorrhage and cystic degeneration, with or without associated calcifications [11]. Specifically, in our patient, these gross pathological findings were well demonstrated in the pre-operative CT scan. Identification of a distinct capsule and cystic degeneration secondary to fragile vascular network of neoplasm is the imaging hallmark of SPN [10].

Evidence of local infiltration, main pancreatic duct obstruction, large tumour size (>6.0 cm), and pancreatic tail location may favour the diagnosis of SPC [10]. Misdiagnosis is not uncommon for cases of SPN as imaging features of SPN may mimic other pancreatic neoplasms. Echo-endosonography provides a pathway for fine-needle puncture biopsy and yielding a definite preoperative pathologic diagnosis of tumour before proceeding to tumour excision has also been shown to be useful.

The primary treatment for both SPNs and SPCs is complete surgical excision, with very good post-surgical prognostication. Studies show that more than 95% of patients with SPN limited to the pancreas are cured by complete surgical excision with a five-year survival rate is as high as 95%–97%, and an estimated 10-year survival rate of approximately 93% [11].

To date, the role of chemoradiotherapy in treating SPNs and SPCs is yet to be analysed. Few suggest radiotherapy in cases of inoperable SPNs, as these tumours appear to be radiosensitive [8].

4. Conclusion

Other than renal origin, suspicion of pancreatic tail origin should be considered by clinicians when encountering a ballotable left abdominal mass. With the availability of cross-sectional imaging, the advancement of histopathology, immunohistology, and molecular markers, an increasing number of cases of SPNs have been diagnosed in the last decade, providing new insight into this pathology [11].

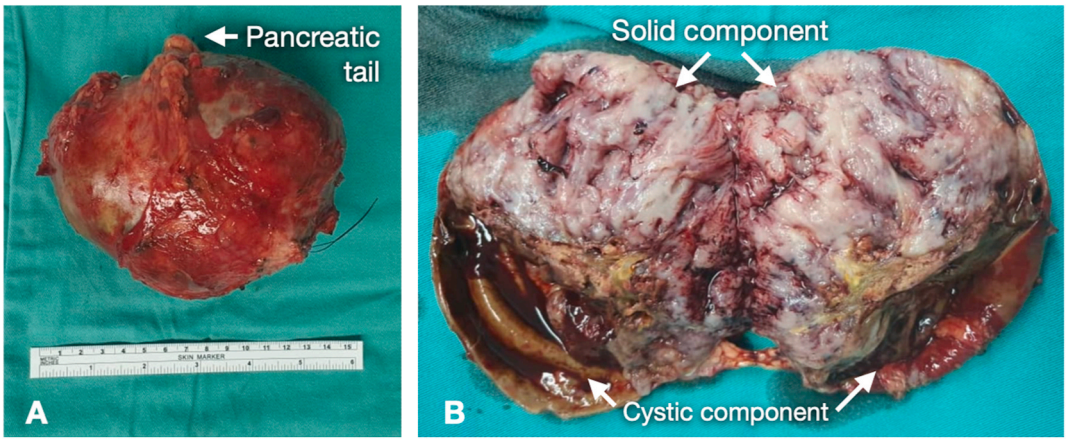

Fig. 4. (A) Gross examination of a large well-encapsulated mass along with a cuff on pancreatic tail tissue measures 15.0cm in length. (B) Gross cross-section of tumor noted cystic (hemorrhagic fluid) and solid tissue components.

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Ethical approval

N/a.

Consent

Written informed consent was obtained from the patient's father for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Guan Tatt Lim and Yong Guang Teh conceptualized the paper and conducted the literature review. Chiak Yot Ng, Hazlina Mohd Khalid and Firdaus Hayati compiled and validated the patient's clincial history, investigations and operative findings. All authors were involved in reviewing the submitted manuscript.

Research registration (for case reports detailing a new surgical technique or new equipment/technology)

N/a.

Guarantor

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Declaration of competing interest

None declared.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.01.003.

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