



## Case report

## A splenic hamartoma: Adding a new case to the literature: A case report

Tarek Abdelazeem Sabra<sup>a,\*</sup>, Ahmed Maher<sup>a</sup>, Rahf AlRashidi<sup>b</sup>, Hussein Ibrahim<sup>a</sup><sup>a</sup> Pediatric surgery unit, Assiut university children hospital, Assiut, Egypt<sup>b</sup> Kuwait general hospital, Kuwait

## ARTICLE INFO

## Keywords:

Splenic hamartoma  
Child  
Splenectomy

## ABSTRACT

**Introduction:** Splenic tumours are relatively rare and include malignancies such as lymphomas, angiosarcomas, plasmacytomas, primary malignant fibrous histiocytomas, and splenic metastases. Benign tumours of the spleen such as hemangiomas, cysts, and inflammatory pseudotumours are very rare (Kaza et al., 2010, PisaniCeretti et al., 2012) [1,2]. There are fewer than 160 cases of splenic hamartoma or splenomas having been reported in the literature (Basso et al., 2012) [3]. Only 20% of the cases were detected in children (Abramowsky et al., 2004) [4]. Although multi-modality imaging findings were described preoperatively, the final diagnosis was splenic hamartoma based on histology and immunohistochemistry.

**Case report:** Here, we report a case of a 14 year old child left upper quadrant abdominal pain and worsening sickness. Multi-modality imaging detected a solid lesion of the spleen, who required splenectomy and was pathologically diagnosed as a splenic hamartoma. The postoperative course was uneventful.

**Discussion:** Splenic hamartoma is very rare. Only 20% of hamartomas occur in children. They are commonly found incidentally on imaging with no symptoms.

**Conclusion:** Splenic hamartoma is a benign vascular proliferative lesion that requires a multi-modality imaging studies for diagnosis and confirmed by histopathology. It must be included in the differential diagnosis of splenic mass forming lesions.

## 1. Case presentation

A 14-year-old girl was admitted to our department due to a 6-month history of left upper quadrant abdominal pain and worsening sickness for 4 days. No specific drug, family or psychosocial history. The spleen is not palpable on Physical examination. She had no significant past medical history, and the laboratory findings (such as blood routine test, serum chemistry test, and tumour biomarker) were also unremarkable. Abdominal ultrasonography showed a hypoechoic mass with well-defined borders occupying splenic hilum that measured about 6 cm in dimensions. Magnetic Resonance Imaging (MRI) showed mild enlarged spleen with a well-defined capsulated splenic hilar focal mass lesion about 6 cm in diameter. It exhibits intermediate intense signal in T2W and SPIR with small hypointense area inside, no detected significant restriction in DWI with no signs suggesting infiltration or extension to adjacent structures as in Fig. 1. All parts of the manuscript are in line with SCARE criteria [5].

The patient underwent abdominal exploration by the first author through a left subcostal incision. There was a large solid mass occupying

splenic hilum about 5.5\*2.5 cm in dimensions as in Fig. 2. Due to the risk of spontaneous rupture and the fact that malignancy could not be excluded, total splenectomy was performed.

Microscopically, the lesion contained a mixture of unorganized vascular channels and fibrotic cords within the splenic red pulp-like area in Fig. 3. The rest of the spleen showed unremarkable histology of red and white pulp. The platelet count increased to  $1320 \times 10^9/L$  on the 5th postoperative day. Oral aspirin was prescribed and continued for 4 weeks after surgery as anticoagulant prophylaxis. No complications developed during postoperative follow-up for 2 years.

Although splenic hamartomas are very rare tumours, they must be considered in the differential diagnosis of splenic lesions in children. However, a splenectomy may be necessary when malignancy cannot be ruled out preoperatively.

## 2. Discussion

Splenic hamartomas are rare benign tumours. Their reported incidence is about 3 in 200,000 splenectomies [6]. The vast majority of cases

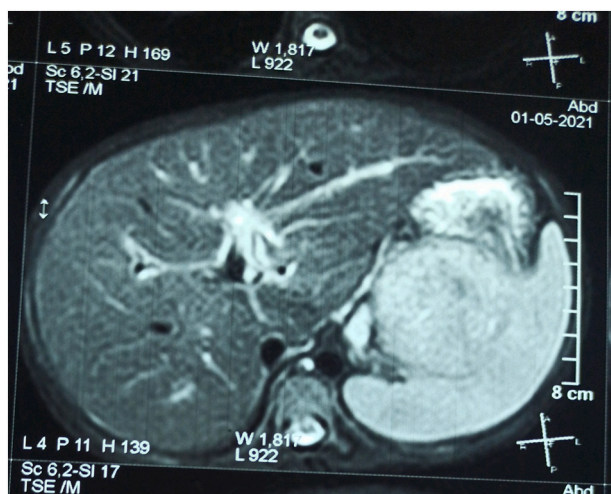
\* Corresponding author.

E-mail address: [tareksabra@aun.edu.eg](mailto:tareksabra@aun.edu.eg) (T.A. Sabra).<https://doi.org/10.1016/j.ijscr.2021.106647>

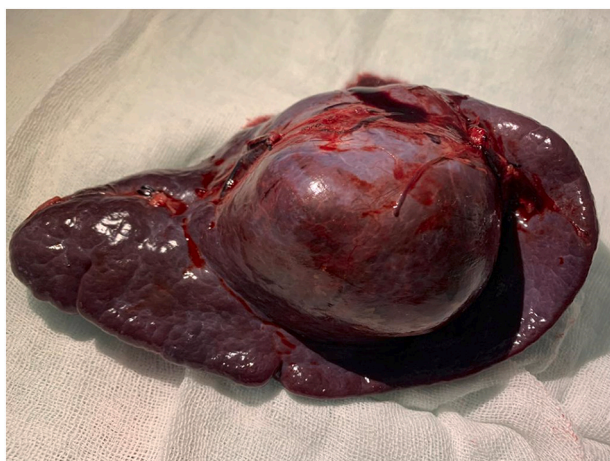
Received 9 October 2021; Received in revised form 21 November 2021; Accepted 28 November 2021

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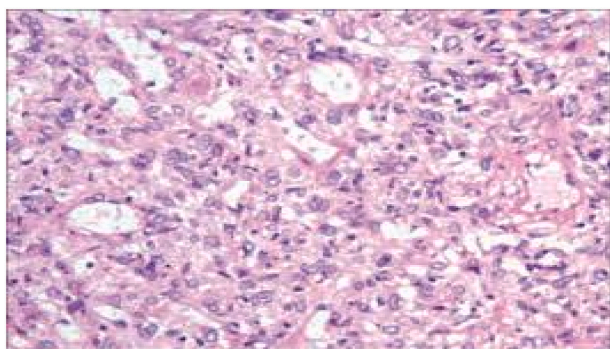
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**Fig. 1.** Mild enlarged spleen with a well-defined capsulated splenic hilar focal mass lesion about 6 cm in diameter.



**Fig. 2.** A large solid mass occupying splenic hilum about 5.5 \*2.5 cm in dimensions for which splenectomy done.



**Fig. 3.** Histopathological examination; lesion containing a mixture of unorganized vascular channels and fibrotic cords of splenic red pulp-like area (hematoxylin-eosin stain). (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

in the literature are of adult population [4,7]. Only 20% of hamartomas occur in children, [4,8–10]. Only 15% of the patients present with symptoms, most commonly abdominal pain, splenomegaly, cytopenia, and incidental spontaneous rupture [3,11]. However, most children

present with systemic manifestations such as fever and malaise [4,8–10].

Despite the fact that the final diagnosis of splenic hamartomas is established by a histopathological evaluation, a preoperative diagnosis using a multi-modality imaging techniques may be possible [8,11–13].

By ultrasound, most hamartomas are hyperechoic relative to the adjacent normal splenic parenchyma [14]. Some splenic hamartomas are homogeneous well-defined solid lesions, with various echogenicity compared to the normal splenic parenchyma, but others may be heterogeneous with cystic changes [11,15]. Not all Splenic hamartomas show hypervascularity because some are hypoechoic and are composed of red pulp, without fibrous trabeculae nor white pulp [15]. Microscopic image of the splenic hamartoma showed the lesion containing a mixture of unorganized vascular channels and fibrotic cords of splenic red pulp-like area (hematoxylin-eosin stain).

Histopathological fibrous splenic hamartomas have a dominant fibrous tissue and MRI shows isointensity or hyperintensity on T1WI images and hypointensity on T2WI images [16]. Non-fibrous splenic hamartomas are more common in the clinic, and MRI reveals an isointense mass on T1WI images and mild hyperintense mass on T2WI images [12].

On delayed images, the density or signal of the lesion is near or slightly higher than that of the splenic parenchyma [11]. Splenic hamartomas should be differentiated from the more common neoplastic disorders of the spleen such as hemangiomas and ominous lesions of the spleen including primary hemangiosarcoma, lymphomas, and metastases [11]. The main pathologic differential diagnosis is with benign vascular tumours or hemangiomas and immunohistochemical staining is required to confirm the diagnosis [4,17]. Hamartomas represent an anomalous cluster of normal splenic red pulp elements. They contain a mixture of unorganized vascular channels lined by endothelial cells and are surrounded by fibrotic cords of predominant splenic red pulp with or without (lymphoid) white pulp [18]. Because of their origin from splenic sinusoids, endothelial cells of hamartomas are CD8+ and CD34– [19]. This staining pattern also differentiates them from splenic hemangiomas, which contain CD8– and CD34+ endothelial cells.

### 3. Conclusion

Although splenic hamartomas are very rare tumours, they must be considered in the differential diagnosis of splenic lesions in children. However, a splenectomy may be necessary when malignancy cannot be ruled out preoperatively.

### Sources of funding

No sources of funding.

### Ethical approval

The case report is exempt from ethical approval in my institution.

### Consent

Written informed consent was obtained from the patient's parents 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.

### Registration of research studies

Not applicable.

### Guarantor

Tarek Abdelazeem sabra.

**Provenance and peer review**

Not commissioned, externally peer reviewed.

**CRedit authorship contribution statement**

Tarek Abdelazeem sabra: main author of the paper and wrote the manuscript. Ahmed Maher: literature review. Rahf Alrashidi: revised the manuscript. Hussein Ibrahim: supervising and editing.

**Declaration of competing interest**

No conflict of interest.

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