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Right sided spleen laying retro-duodenal: A case report and review of the literature

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A R T I C L E   I N F O

Article history:
Received 21 April 2016
Received in revised form 25 April 2016
Accepted 28 April 2016
Available online 6 May 2016

Keywords:
Case report
Spleen
Accessory spleen
Retroperitoneal mass

A B S T R A C T

INTRODUCTION: Unlike left sided accessory spleen that are seen in 10–30% of cases at autopsy, cases of right accessory spleens are extremely rare. This congenital body of healthy splenic tissue simulates tumors from neighboring organs and presents a challenge in formulating a differential diagnosis.

PRESENTATION OF CASE: We present the case of a patient whose CT scan of the abdomen showed a large mass, \(11 \times 8\) cm, arising retro-duodenal and lying just anterior to the right kidney. To the best of our knowledge, this is the only case where the accessory spleen was found retro-duodenal, directly anterior to the kidney and completely separate from the supra-renal gland. The chief complaint of the patient was right upper quadrant pain, radiating to the back, and colicky in nature. The patient was diagnosed with duodenal gastro-intestinal stromal tumor and a retro-peritoneal sarcoma. The mass was removed via a Kocher’s incision and immunohistological examination showed that it was a right sided accessory spleen. The patient’s left sided spleen appeared normal.

DISCUSSION: Efforts to distinguish an accessory spleen from a retroperitoneal tumor with available scans, percutaneous biopsy or biochemical tests are inconclusive. Differential diagnosis between a retroperitoneal tumor and an accessory spleen can only be made after surgical exploration.

CONCLUSION: This case highlights the fact that surgeons should consider the possibility of an accessory spleen when making a differential diagnosis of retroperitoneal tumors.

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1. Introduction

The spleen is the largest lymphoid organ and lies on the left side of the abdomen, between the 9th and 11th ribs, weighing 7 ounces. Accessory spleens are congenital and result from the failure of the primordial splenic buds in the dorsal mesogastrium to fuse during the fifth week of fetal life. Although benign, infarction, trauma and torsion are common complications that may arise from this ectopic tissue [1]. They are relatively common and are seen with 10%–30% frequency at autopsy and in 16% of scans of patients undergoing contrast enhanced computed tomography (CT) [2–4]. This case is that of a patient with a right sided accessory spleen directly anterior to the kidney and completely separate from the supra-renal gland. To the best of our knowledge, this is the first reported right accessory spleen laying retro-duodenal and second largest reported [5–8] (Table 1).

2. Presentation of case

A 44-year-old male, presented at the hospital with a history of right upper quadrant pain, radiating to the back, and colicky in nature (Table 2). All biochemical tests were unremarkable. He was diagnosed as having biliary colic. The patient subsequently had an abdominal ultrasound scan which showed multiple sub-centimeter gallstones, and a mass arising from the liver (Fig. 1). This was followed by a CT scan of the abdomen with IV contrast which showed a large mass, approximately \(11 \times 8\) cm in diameter, arising retro-duodenal, lying just anterior to the right kidney (Fig. 2). The differential diagnoses included a duodenal gastro-intestinal stromal tumor and a retro-peritoneal sarcoma.

The patient was consented for removal of the mass, and a Kocher’s incision was performed. Omental adhesions to the gall-

http://dx.doi.org/10.1016/j.ijscr.2016.04.050
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Table 1
Summary of cases of right-side accessory spleen.

<table>
<thead>
<tr>
<th>Author, year</th>
<th>Age (yrs), gender</th>
<th>Presenting symptom</th>
<th>Size</th>
<th>Location</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kim et al., 2008 [8]</td>
<td>68, male</td>
<td>CT detection of right-sided mass</td>
<td>4 × 3.8 cm</td>
<td>Right retroperitoneal region</td>
</tr>
<tr>
<td>Arra et al., 2013 [6]</td>
<td>24, male</td>
<td>Thalassaemic patient with self detected right-sided abdominal mass</td>
<td>20 cm</td>
<td>Right suprarenal region</td>
</tr>
<tr>
<td>Zhou et al., 2015 [7]</td>
<td>40, female</td>
<td>Ultrasound detection of right-sided mass</td>
<td>3.4 × 2.5 cm</td>
<td>Right retroperitoneal region</td>
</tr>
</tbody>
</table>

Fig. 1. Ultrasound showing mass adjacent to gallbladder, arising from the liver.

Fig. 2. CT scan showing mass separate from right kidney laying retro-duodenal.
bladder and liver were dissected and the duodenum kocherized, to reveal the mass, with no attachment to the duodenum (Fig. 3). There were large veins inserting into the inferior vena cava and the mass was resected en bloc by dissection with the ultrasonic dissector (Fig. 4, Fig. 5). A cholecystectomy was then performed. Immunohistological examination of the specimen revealed a benign spleen (Fig. 6). The patient’s left sided spleen appeared normal on radiological and visual inspection. The patient had an uneventful recovery.

3. Discussion

There are two types of accessory splenic tissue – accessory spleens and spleniculi. Accessory spleens have normal splenic histology compared with splenotic tissues, with histology usually revealing distorted architecture with no hilum, a poorly formed capsule and tissue of any shape or size. These may be due to splenic trauma or rupture and may occur anywhere along the hilum of the
Fig. 5. 11 cm × 8 cm right accessory spleen.

Fig. 6. Microscopic images of accessory spleen. A. H&E. There is an outer fibrous capsule (arrow) surrounding typical-appearing splenic parenchyma. B. H&E. Higher magnification highlights a focus of white pulp (arrow) in a background of red pulp (cords and sinuses). Immunohistochemical stains highlight the native population of CD20-positive B-lymphocytes (C.) and CD3-positive T-lymphocytes.
spleen, the tail of the pancreas, the gastro splenic and splenorenal ligaments, the walls of the stomach or intestines, the greater omentum, the mesentery, the adrenals, and the gonads in the case of splenogonadal fusion [9–13]. They are usually small with diameter between 0.8 and 3.2 cm with a mean of 1.6 cm [3,14]. Accessory spleens have been reported to have compensatory hypertrophy reaching 3–5 cm after a splenectomy or arising after a variety of hematological disorders [2]. Previous cases of right accessory spleens are presented in Table 1. In this case, the enlarged accessory spleen with normal splenic architecture exceeded this range despite the absence of a splenectomy or any clear etiology.

Malignant retroperitoneal tumors are more common than benign tumors and account for approximately ~0.1% of all malignancies. However, differential diagnosis between a retroperitoneal tumor and an accessory spleen can only be made after surgical exploration. Knowing that accessory spleens are benign, the diagnosis should ideally be confirmed prior to surgical exploration. However, distinguishing an accessory spleen from a retroperitoneal tumor with ultrasound. CT or MRI scan is still inconclusive, and percutaneous biopsy of the tissue would be insufficient to differentiate it from a tumor. Singly CT scans with biochemical tests may not be able to correctly identify accessory spleens [15]. Radionuclide imaging with Tc-99m-labeled red blood cells has been proposed to rule out an accessory spleen [16]. For patients who had a splenectomy or in cases with diagnostic uncertainty after the usual imaging modalities, preoperative nuclear medicine scintigraphy could be added to the workup. Multiple case reports have shown the presence and subsequent removal of left sided accessory spleens, being mistaken for retro-peritoneal sarcomas or adrenal masses. [4,17–19].

4. Conclusion

It is exceedingly rare to find a right sided accessory spleen. Most are reported to arise from the right supra-renal gland, however in this case the mass was found retro-duodenal and directly anterior to the kidney and completely separate from the supra-renal gland. In this case the anatomic location and size of the accessory spleen were unusual. The strength of our approach was the reliance on the surgical findings to evaluate the differential diagnosis before any other clinical intervention. This case is a reminder that the possibility of an accessory spleen should always be considered in the formulation of a differential diagnosis when investigating a retroperitoneal mass.

Conflicts of interest

None to declare.

Source of funding

WAW was supported by Washington University School of Medicine, GSAS/CGFP Fund94028C. The contents of this manuscript are solely the responsibility of the authors and do not necessarily represent the views of the affiliating institutions.

Ethical approval

Ethical approval was not required since patient is de-identified.

Consent

Written informed consent was obtained from the patient 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

The operation was carried out by Ravi Maharaj and Wesley Ramcharan. Pathology was reported by Wesley Greaves. Wesley Ramcharan contributed to the clinical management of the patient. The radiological imaging studies were reported and managed by Paramanand Maharaj. Manuscript was written by Wayne A. Warner and Wesley Ramcharan. All authors have read and approved the manuscript.

 Guarantor

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