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Case Report
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Giant Accessory Spleen and Rotating Ectopic Kidney: A Rare Association

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Abstract

The accessory spleen is a congenital disability characterized by a separated ectopic splenic parenchyma. Wandering Spleen (WS) is a rare condition where the spleen, free from its ligaments, is allowed to move inside the abdomen, predisposing the patient to life-threatening complications due to torsion of the vascular pedicle; splenic infarction, portal hypertension, bleeding and acute abdomen may occur. WS or Accessory Spleen (AS) is rarely suspected at presentation since symptoms are usually not specific, and definitive diagnosis is often reached only by imaging technologies such as color flow ultrasonography and angio-spiral computed tomography. The size is rarely more than 4 cm. Patients rarely present symptoms, and the diagnosis can be accidental when faced with ectopia in radiological or intraoperative investigations. It is an essential clinical entity since abdomen-pelvic tumors must always be considered as differential diagnoses, requiring detailed evaluation. Surgical treatment is indicated for symptomatic patients or those with complications. On the other hand, the ectopic kidney with rotation abnormality also consists of a singular entity. Congenital anomalies of the upper urinary tract occur in 3% to 4% of those born, most of which are followed up clinically. The present study discusses a case of symptomatic intrapelvic WS associated with an ectopic kidney with rotation anomaly, a rare union, and still with scarce reports in the literature. This study aimed to present the case of a patient with an isolated oversized accessory spleen and a review of the literature.

Keywords: Spleen; Accessory spleen; Wandering spleen; Displaced spleen; Abdominal pain; Splenectomy

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Introduction

The spleen is localized between the 9th and 11th left ribs in the abdominal cavity between the gastric bottom and the left hemidiaphragm. With a weight of approximately 200 g, it represents the largest lymphoid organ in the body [1-3]. It is fundamental for the hematological and immune system and is an essential reserve of approximately 10% to 20% of the blood volume [4]. During its growth, the spleen can develop anomalies such as complete agenesis, multiple spleens or polysplenia, accessories spleens, and persistent lobulation. The Accessory Spleen (AS) is a congenital alteration resulting from the failure to move the splenic tissue to its definitive position in the left hypochondrium. This is a condition present in 10% of the population, located in regions such as splenic hilum or pancreatic tail, with an intrapelvic location being unusual [5,6]. Accessory Spleen (AS) may develop during the sixth week of embryogenesis following the deposition of spleen cells along the path from the midline, usually occurring on the left side. An AS is commonly located near the spleen's hilum and in the pancreas tail, and is generally asymptomatic and diagnosed incidentally during laparotomy or radiological examination performed for other reasons [7]. Adnexal masses typically originate from the genital tract; however, extrapelvic organs are rarely found in this location [8]. Although extremely rare, this possibility must be considered in the differential diagnosis of pelvic masses. Patients rarely present symptoms, and the diagnosis can be accidental when faced with ectopia in radiological or intraoperative investigations [9-11]. It is an essential clinical entity since abdomen-pelvic tumors must always be considered as differential diagnoses, requiring detailed evaluation [12-14]. Surgical treatment is indicated for symptomatic patients or those with complications. On the other hand, the ectopic kidney with rotation abnormality also consists of a singular entity. Congenital anomalies

of the upper urinary tract occur in 3% to 4% of those born, most of which are followed up clinically [15-17]. The present study discusses a case of symptomatic intrapelvic AS associated with an ectopic kidney with rotation anomaly, a rare union, and still with scarce reports in the literature.

Methods

We searched the PubMed, Cochrane Database, Scopus, Web of Science, Embase, and Google Scholar databases (gray literature) using the keywords: Spleen, accessory spleen, wandering spleen; displaced spleen; abdominal pain; splenectomy. The selection of articles was made by two authors independently. A third author was consulted in case of divergence in the inclusion or not of a particular study. Reports related to accessory spleen were selected based on the chosen keywords, human studies, full-text articles, and published in English, Portuguese, or Spanish. Forty-three studies unrelated to the main objective of the review or outside the inclusion criteria were excluded after peer review. We included 26 articles from a total of 37 scientifically validated and relevant published studies. The Research Ethics Committee of Potiguar University - Natal/Brazil, approved the research after signature by the patient informed consent, according to Resolution 466/2012, of the National Commission of Ethics and Research in Humans - CONEP - Ministry of Health/Brazil. Protocol number 138/2019.

Case Presentation

A 32-year-old male, previously healthy, sought emergency care after opening a picture of sudden low back pain of extreme intensity, radiating to the testicles, in a burning character, with no history of previous pathology or abdominal trauma. He was medicated with antispasmodics and analgesics, progressing with pain improvement. During evolution, an ultrasound examination was requested, revealing a single solid spherical nodule, with circumscribed margins, with similar, retrovesical hypoechogenic texture, with the central arterial flow at Doppler (Figure 1), suggested further investigation by computed tomography. The tomographic examination confirmed the renal ectopia on the left and the presence of a solid mass similar to the splenic parenchyma, showing a relationship with the posterior wall of the bladder and with the left seminal vesicle, an apparent cleavage plane with both, but with no evident cleavage plane with the upper

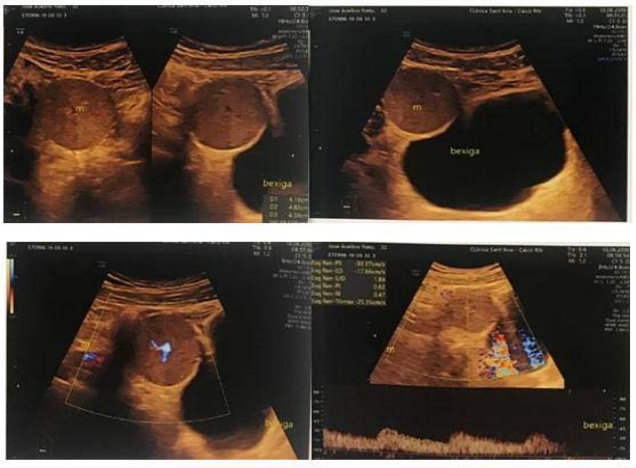

Figure 1: Pelvic ultrasound showing a single solid spherical nodule with circumscribed margins, with similar, retrovesical splenic hypoechogenic texture, with the central arterial flow at doppler.

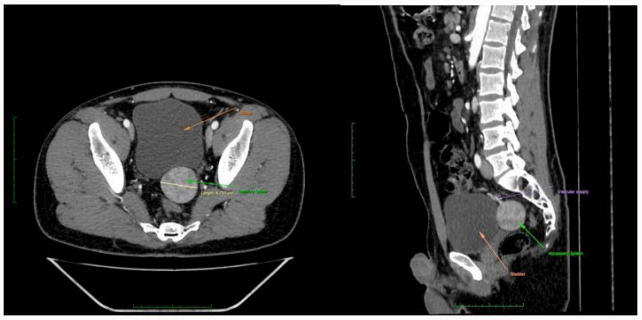

Figure 2: CT scan of the pelvis showing an ectopic spleen with a vascular pedicle.

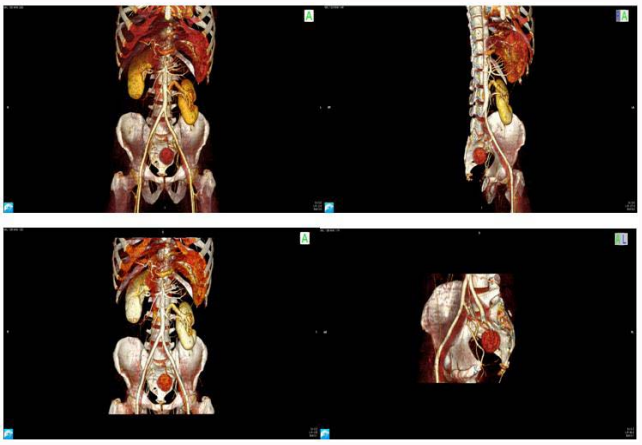

Figure 3: 3D CT scan of the pelvis showing an ectopic pelvic spleen with a vascular pedicle and left kidney rotation in 180°.

rectum and a left ectopic kidney rotating 180º in it longitudinal axis (Figure 2 and 3). After the exams, the patient developed constipation reporting ten days without fecal eliminations, without previous episodes of illness, taking him to the gastroenterology service, where he was examined and submitted to a colonoscopy, which did not reveal any changes. He was treated with symptomatic and laxative phytotherapeutics, improving and oriented to seek general surgery service. The patient was referred and evaluated for a surgical procedure to remove the mass found in the imaging exams. With no comorbidities, no family history of neoplasms, or malformations, he underwent surgery to resolve a pilonidal cyst in childhood. We opted for exploratory laparotomy, with an encapsulated mass with a smooth violet-shiny appearance, measuring about 4.0 cm × 4.0 cm × 4.5 cm (Figure 4), with an ascending vascular pedicle invading the omentum bigger. The mass was resected and sent for anatomopathological study, which characterized tissue compatible with the accessory spleen, congested parenchyma, and atrophy of white pulp (Figure 5). After the procedure, the patient evolved without complications and was discharged in twenty-four hours, being reassessed fifteen days after the surgical intervention, favorable recovery, and no episode of pain in this period.

Discussion

The spleen is an encapsulated organ composed of lymphatic and vascular tissue, located in the left hypochondrium [8]. Some variations in its development have already been observed, including complete agenesis, polysplenia, isolated accessory spleens, and persistent lobulations [9,18]. The organ begins its evolution during the fifth

Figure 4: Exploratory laparotomy, with an encapsulated mass with a smooth violet-shiny appearance, measuring about 4.0 cm × 4.0 cm × 4.5 cm, with an ascending vascular pedicle invading the omentum bigger (Ectopic spleen with a vascular pedicle).

week of embryonic life, from the condensation of mesenchymal cells present between the layers of the dorsal mesogastrium [7,19]. The Accessory Spleen (AS) results from the formation of ectopic splenic tissue or from failure to migrate the organ, from its original site, in the midline, to its final position, in the upper left abdomen. An accessory spleen is defined as ectopic splenic tissue that develops due to the failure of cell fusion during embryonic development while migrating from the midline to the left upper quadrant [15,16]. They can be localized commonly next to the hilus and vascular pedicle, the tail of the pancreas, left ovary or left testis, in the greater omentum, and the mesentery of the small and the large intestine, along the greater curvature of the stomach and in the pouch of Douglas. Macroscopically, a typical accessory spleen usually appears as a solid mass, 1 cm to 2.5 cm in diameter. Masses larger than 4 cm are very rare [2,5] with a smooth, round, ovoid, or minimally lobulated shape. Microscopically, it reproduces a splenic pattern. An accessory spleen commonly has a well-defined fibrotic capsule that separates the surrounding healthy tissue [18]. According to necropsy studies, AS is present in 10% of the population [20]. It's most common locations are the splenic hilum (75%) and the tail of the pancreas (20%), as well as the greater omentum, greater curvature of the stomach, and mesentery [1-3]. Unver Dogan et al. [1] investigated 720 autopsies, and BA was found in 6.7% of cases, 2 of which were pelvic. The patient in the present study presented intrapelvic AS, representing an atypical anatomical variety and with few reports in the scientific literature. In addition to the congenital origin, in rare cases, its source is acquired, resulting from the autotransplantation of splenic tissue during splenectomy or after abdominal trauma, a process defined as splenosis. However, because this patient does not have a history of trauma or splenectomy, it is a congenital intrapelvic AS [21]. WS appears as an incidental finding, without clinical significance, with no preference for sex or race, asymptomatic and accidentally diagnosed during radiological investigations or abdominal surgeries [13-15]. Thus, the report of low back pain with irradiation to testicles is another peculiarity of the present case, since symptoms, as a rule, are uncommon. Due to the clinical presentation, renal lithiasis was suspected, analgesic drugs were prescribed, and an abdominal ultrasound was requested for diagnostic confirmation [18,22]. USG did not show signs of nephrolithiasis but found a solid pelvic nodule with unspecific echographic characteristics. Rectal tumor, bladder tumor, lymphadenomegaly, abscess, retroperitoneal fibrosis, and other masses of intrapelvic location form hypotheses suggested in these circumstances [14,23]. A CT scan was performed, given the demand for more accurate imaging evaluation. The report showed a nodule with behavior similar to the splenic parenchyma during the dynamic contrast phase when BA was then included in the list of differential diagnoses. Preoperative diagnosis of the accessory spleen is difficult, especially in emergencies [17-19,24]. CT scans show a well-margined mass, similar to the splenic parenchyma on the contrast phase. Magnetic imaging can also be used to evaluate tissue aspects and the vascular pedicle of the accessory spleen. Only nuclear medicine imaging can confirm the diagnosis with scintigraphy performed with 99mTc-labelled colloids or Tc-99 m heat-damaged red blood cells because the colloid labeled with Tc-99m is taken from the reticulum-endothelium and makes visible the spleen, liver and bone marrow; however, it is necessary to suspect the diagnosis of accessory spleen for this procedure [22]. Therefore, often only surgical excision can safely confirm the diagnosis [23,25]. For these reasons, many surgical procedures have been done for diagnosis. Laparotomy was performed for resection of the lesion and anatomopathological study, whose report confirmed splenic tissue. The open road was chosen due to the retrovesical location and the team's more significant experience with this technique [14-16]. However, the laparoscopic approach is also an option for treatment. Torsion, spontaneous rupture, hemorrhage, and cystic formations are responsible for the clinical condition of symptomatic patients, but they are rare complications [9,18]. The congestion demonstrated by light microscopy was the probable cause of the symptoms in the patient in question. It is worth mentioning another anatomical variety presented by the patient, detected by imaging exams (USG and CT): Ectopia and 180º rotation of the left kidney, characterizing an exaggerated kidney over-rotation [11,17]. Congenital anomalies of the upper urinary tract involving kidneys and ureters occur in 3% to 4% of those born, with abnormalities in the shape and position of the organs being the most common [7,23]. Most of these renal anomalies are followed clinically, requiring a correct diagnosis of the morphological alteration, as well as the evaluation of possible complications [24-26]. During the review of the literature on the topic, the authors not found reports of an association between an accessory spleen and an ectopic kidney. Despite this, this variety did not affect the patient's symptoms in this case.

Conclusion

In conclusion, the present study demonstrated a rare case of the pelvic accessory spleen associated with a 180° renal rotation that was only definitively elucidated in the intraoperative period. An accessory spleen can be variously located, and the pelvic position is hugely uncommon. Preoperative diagnosis is still tricky, especially in an emergency, and as in our case, the literature shows the difficulty of reaching a diagnosis before surgery. The main misdiagnosis is a neoplastic disease, and for this reason, accessory spleen could be wrongly removed. For this reason, undiagnosed pre or intraoperative pelvic mass, firmly to the spleen, has to be managed carefully. Despite pelvic AS being a rare, usually asymptomatic, pelvic mass condition, it should be considered in the differential diagnosis of symptomatic

pelvic masses.

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