WANDERING SPLEEN - A POSSIBLE CAUSE OF ADRENAL ”MASS“ - CASE REPORT

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LUTAJUĆA SLEZINA KAO MOGUĆI UZROK ADRENALNE „MASE“ - PRIKAZ SLUČAJA

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ABSTRACT

Wandering spleen is a very rare clinical condition characterized by spleen absence in the normal anatomical location in the upper left quadrant of the abdomen and its presence at another location in the abdomen or pelvis. The ectopic spleen is extremely rare in children, where its increased mobility is the result of a congenital disturbance of the fixation for the anterior wall due to the absence or weakness of the supporting ligaments. Wandering spleen is usually asymptomatic, but its torsion is possible, as well as infarction or rupture which demand an urgent diagnosis and surgical treatment. The diagnosis of wandering spleen can easily be overlooked due to low incidence and insufficient clinical experience, which multiplies patient's risk from life-threatening conditions. We present a case of wandering spleen in an 11-year-old girl with acute abdominal pain, which after ultrasound examination raised suspicion on the right adrenal gland tumor. Additional diagnostics verified an ectopic spleen in the right adrenal box, after which the recommended preventive splenopexy was seriously considered. Due to the fixation of the vital spleen in the new position, but also the negative attitude of the parents towards the surgical intervention, clinical monitoring was selected, with exclusion of intense physical activity that carries the risk of traumatization of the spleen. As the girl has been in good health for over 3 years and without symptoms, we consider that the selection of conservative access although difficult, was correct. We hope that our experience in treating wandering spleen in girls will increase the number of valid facts about this rare condition.

Keywords: *spleen, splenic diseases, splenic torsion, wandering spleen*

SAŽETAK

Lutajuća slezina je vrlo retko kliničko stanje koje odlikuje odsustvo slezine u normalnom anatomskom položaju u gornjem levom kvadrantu abdomena i prisustvo na drugom mestu u abdomenu ili karlici. Ektopična slezina je izuzetno retka kod dece, gde je njena povećana mobilnost posledica kongenitalnog poremećaja fiksacije za prednji trbušni zid usled odsustva ili slabosti potpornih ligamenta. Obično je asimptomatska, ali je moguća njena torzija, infarkt ili ruptura, kada je neophodna urgentna dijagnostika i splenektomija. Dijagnoza lutajuće slezine ne može se lako prevideti zbog niske učestalosti i nedovoljnog kliničkog iskustva, što višestruko povećava rizik pacijenta od stanja opasnih po život. U radu se prikazuje slučaj lutajuće slezine 11-godišnje devojčice sa akutnim abdominalnim bolom, kod koje je nakon ultrazvučnog pregleda posumnjano na tumor desne nadbubrežne žlezde. Dodatnom dijagnostikom je verifikovana ektopična slezina u desnoj nadbubrežnoj loži, nakon čega je pažljivo razmatrana preporučena preventivna splenopeksija. S obzirom na fiksiranost vitalne slezine u novom položaju, ali i negativan stav roditelja prema hirurškoj intervenciji, izabrano je kliničko praćenje, sa poštedom od intenzivne fizičke aktivnosti koja nosi rizik od traumatizacije slezine. Kako je devojčica tokom skoro 3 godine od dijagnoze dobrog zdravstvenog stanja i bez simptoma, smatramo da je izbor konzervativnog pristupa iako težak, bio ispravan. Nadamo se da će naše iskustvo kao i pozitivni ishod u tretmanu lutajuće slezine kod devojčice doprineti povećanju broja važnih činjenica o ovom retkom stanju.

Ključne reči: *slezina, bolesti slezine, torzija slezine, lutajuća slezina*

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INTRODUCTION

"Wandering spleen" is a very rare clinical condition characterized by the absence of spleen outside the normal anatomical position in the upper left quadrant of the abdomen (1, 2) and the presence at another place in the abdomen or small pelvis (3, 4). The etiology is multifactorial and potentially very serious, sometimes even life-threatening. (5-7). This condition is extremely rare in children, where, with the developmental disorder, weakness and disturbance of the fixation of spleen supporting ligaments to the front abdominal wall (4, 7, 8), there may be hypoplasia of the anterior abdominal wall muscle with the appearance of dry plums ("belly syndrome") event/congenital hernia of the diaphragm, volvulus of the stomach, an ectopic kidney, obstructive anomalies of the urinary tract and/or retention of the testis (8, 9).

In adults, wandering spleen is in a state where the force of gravity, trauma, enlargement of the spleen in various illnesses, or hormonal changes in pregnancy can cause increased mobility of the spleen (10, 11). Wandering spleen most often appears as asymptomatic or with non-specific gastrointestinal symptoms, dysuria or dysmenorrhea in women (10-12). Sometimes, there are episodes of acute or recurrent abdominal pain with/without melanoma and hematemesis caused by torsion and spontaneous deterioration of ligaments and/or blood vessels of the spleen and other organs (13-15). Symptoms can be alleviated by stretching and occupying the most suitable position in which the ligaments swell and the spleen returns to normal.

After such episodes, fibrosis and abscess of the spleen, as well as hypersplenism, can develop (5-16). In the most severe cases, there is a clinical picture of the acute abdomen condition (2-15) due to arising ischemia, bleeding, necrosis and acute splenomegaly, which can fatally affect patients and requires rapid diagnosis and immediate surgical treatment in the form of splenectomy (17, 18). In the cases of recurrent serious symptomatology, preventive splenopexy is increasingly being proposed, which permanently reduces symptomatology.

CASE REPORT

We present the case of an 11-year-old girl who was diagnosed with a tumor in the right adrenal gland during an abdominal pain episode, but after the diagnosis, the condition of wandering spleen was verified. After a slight improvement and recurrent episodes of abdominal pain, clinical trials were verified at the age of 4: Situs solitus, Displasio v.mitralis cum regurgitatio trivialis, Reflux oesophagitis, urease negative Gastritis antralis. After H2 blocker therapy, a significant reduction in symptoms was experienced, which were repeated at the age of 7. Then, the echosonographic examination of the abdomen was neat, and the introduced therapy with a proton pump blocker continued to reduce symptoms for more than 3 years.

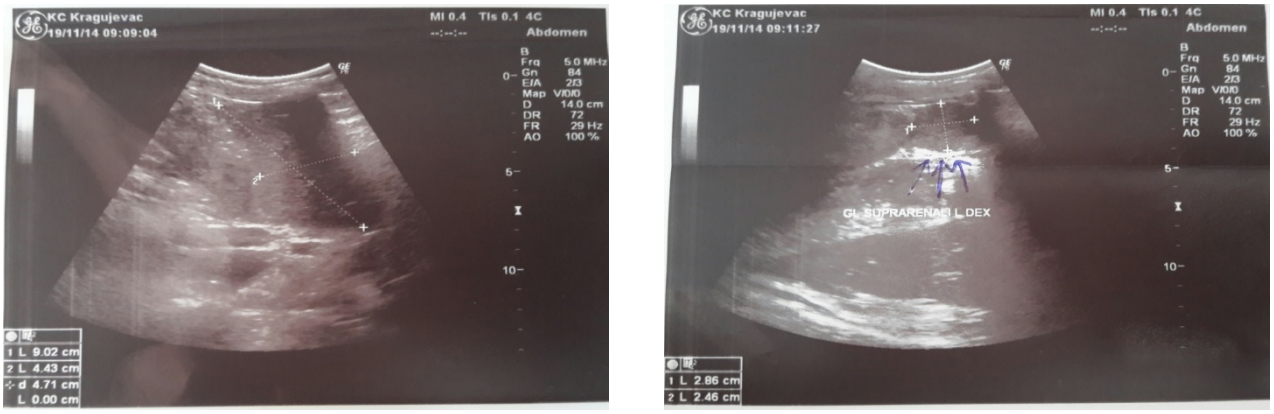
At the age of 11, the girl was hospitalized at the Clinic of Pediatric Surgery, Clinical Centre Kragujevac, for vomiting and severe pain in the right lumbar bed. After the ultrasound examination of the abdomen, a tumor in the right adrenal lodge was suspected. An additional examination at the Pediatric Clinic established that the girl had regular vital functions, growth, development and nutrition: TV + 0.25SD, BMI 16, 43kg/m2 (p50) and initial pubertal signs (B2, PH2). The analysis performed - blood count, biochemical analysis and hormonal status were in reference values.

The ultrasound examination described an elliptical 90x44mm diameter change in the left upper quadrant of the abdomen that could correspond to the tissue of the spleen, left lobe of the liver, and tumor formation in the right adrenal lodge (Figure 1: a, b). In the absence of clinical and biochemical indicators of the right ankle tumor, a contrast-abdominal NMR was performed (Figure 2: a, b) which showed that the left lobe of the liver was present in the left hypochondriac, which did not contain the spleen tissue. As the right retroperitoneal, behind the liver and above the right ankle, two changes of the correct crown shape, diameter 35mm and 33mm, dominantly benign, were seen as an anomalous position of the spleen. A selective spleen scintigraphy (99mTc-denaturated erythrocytes) at a given location showed a spleen of the correct shape that intensely and relatively evenly connected the radionuclide (Figure 3). Thus, the diagnosis of the isolated fixed ectopic spleen of the preserved vitality was established.

After that, the best therapeutic options for the girl were considered - primarily, preventive splenopexy was recommended. Given the occasional symptomatology, the preserved function and the apparently permanently fixed position of the spleen in a new position, as well as the negative attitude of the parents towards a surgical intervention, a more conservative approach was selected, with the restriction of more intense physical activity, which increases the risk of traumatization of the spleen.

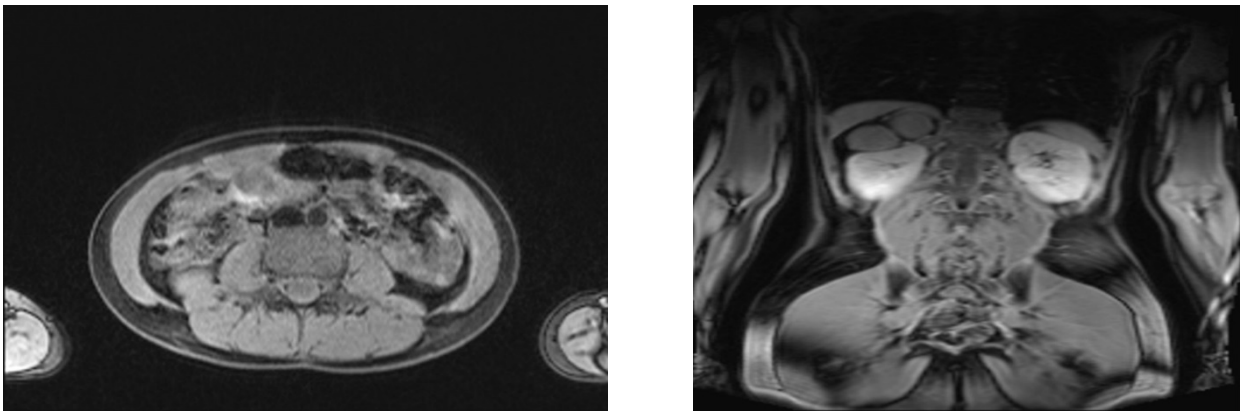
For more than 3 years of monitoring the girl was healthy, without abdominal or hematologic symptomatology.

Figure 1 (a, b): Abdominal ultrasound recordings show that spleen tissue does not clearly differentiate into the left hypochondriac, where normal liver signal is seen (a) while two ovoid formations are visualized in the region of the right adrenal gland (b)



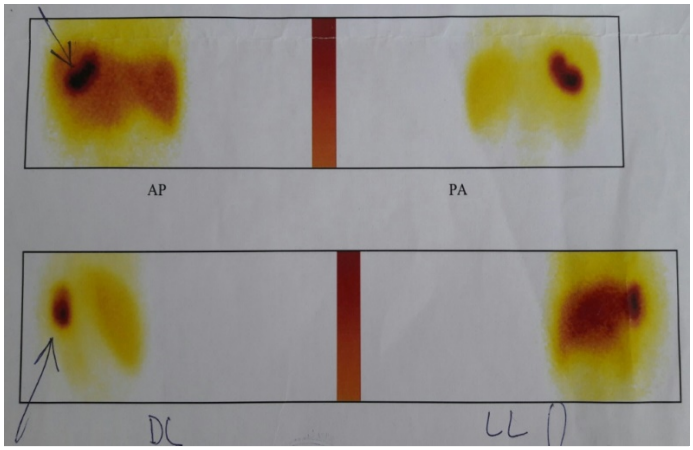
a) b)

Figure 2 (a, b): Transverse and sagittal MR section of the abdomen showing a blank left hypochondriac, partly filled with the left lobe of the liver (a) and retroperitoneal and below the diaphragm, above the upper pole of the right adrenal gland two circular changes diameter 33mm and 35mm, which correspond to ectopic spleen (b)



a) b)

Figure 3. Selective spleen scintigraphy (99mTc-denatured erythrocytes) shows a spleen of regular shape, with intense and relatively evenly bound radionuclide, located in the right hypochondriac, behind the right lobe of the liver and above the right adrenal gland.



DISCUSSION

Wandering spleen in children is a condition that is rarely displayed in the professional literature, (1, 2) where it is most often talked about its congenital origin (4, 13, 20). Given the early onset of symptoms, as is the case in our patient, asymptomatic flow with occasional abdominal symptomatology can be attributed to occasional torsion/detachment of ligaments and/or vascular spleen or spontaneous gastrointestinal obstruction (19, 21, 22). An echosonographic examination revealed the ectopic position of the spleen only after its permanent fixation in the new position (12, 23). Diagnostic procedures have met the criteria for diagnosing an isolated wandering spleen (6, 9), since the situs inversus was excluded in differential diagnosis (5, 9). Analyzing recommendations for the treatment of wandering spleen in children who depend on the severity of symptoms, detailed estimates of its location, size, and functional status (18-20), we did not consider splenectomy as it is recommended in acute cases or in cases of hypersplenism (13-18). Considering the important hematological and immunological functions of the spleen at that age, further treatment after splenectomy must be very careful (4, 6).

Due to the high risk of developing a life-threatening infection with encapsulated strains of bacteria, immunization against these causes (13-15) is mandatory, but also the use of antibiotics at the first signs of infection (4, 17). Leading to a maximally conservative approach, we seriously considered preventive splenopexy (21-23). By this indication, the first classical splenopexy was performed in 1990, and laparoscopic splenopexy in 1998, continues to be technically advanced (21-25). Relying on occasional symptomatology, preserved vitality and apparently permanently fixed new position of the spleen, and looking at the negative attitude of parents towards surgical therapy, we chose a more conservative approach. The fact that the girl has been in good health for almost 3 years after diagnostics, without symptomatology, suggests that our therapeutic choice, although difficult, was correct, so we hope that our experience will contribute to an increase in the number of valid facts about this rare condition.

CONFLICT OF INTERESTS

Not declared.

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