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Laparoscopic splenectomy for a simultaneous wandering spleen along with an ectopic accessory spleen. Case report and review of the literature

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ABSTRACT

BACKGROUND: Wandering spleen and accessory spleen are uncommon entity occurring during embryonic development. Wandering spleen results in an excessive mobility and migration of the spleen from its normal position in the left hypochondrium while accessory spleen is characterized by ectopic splenic masses or tissue disjointed from the main body of spleen.

Due to the nonspecific and multiple symptoms the clinical diagnosis of both conditions is uncertain even with imaging techniques, such as CT and MRI. The coexistence of both diseases (wandering spleen and accessory spleen) is uncommon.

CASE REPORT: A 17-year old European female with a history of minor beta thalassemia and recurrent attacks of abdominal pain. Pre-operative management consisted of routine laboratory tests, ultrasound, CT scan. An ectopic spleen along with an accessory spleen were diagnosed. After a multidisciplinary board a laparoscopic splenectomy was performed. Post-operative recovery was uneventful, and the patient was discharged on the 6th post-operative day with the indication to continue the therapy with low molecular weight heparin (LMWH) for 30 days

CONCLUSIONS: This case represents a simultaneous condition of wandering splenomegaly along with an ectopic wandering spleen. The coexistence of these two rare conditions is peculiar such as the age of the patient, as literature reports such diseases to affect children or more commonly people in the range of 20–40 years of age. Laparoscopic treatment for this particular condition is also unusual.

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1. Introduction

Wandering spleen and accessory spleen are uncommon entity occurring during embryonic development. Each one of these diseases is rare and the association of both seems extremely rare and uncommon.

In ancient times, the knowledge referring to wandering spleen was based on the humor doctrine, according to which the spleen was believed to be an organ producing black bile. Galen himself called the spleen “organum plenum mysterii” [1].

Over the centuries the condition of wandering spleen was surrounded by an *aura* of mystery, since it was associated to hypocondria, hysteria and even to neurasthenia [2].

In the middle of the 19th century, Carl von Rokitansky, one of the most influential members of the Vienna School of Medicine, described the condition of wandering spleen in the “Lehrbuch der

pathologische Anatomie” [3]. In the years from 1854 to 1863 the Polish physician Józef Dietl was the first to describe patients with wandering spleen and indicated this condition to be life threatening [4].

Twentieth century medicine described wandering spleen or hypermobile spleen as an anatomic condition characterized by excessive mobility and migration of the spleen from its normal position in the left hypochondrium mainly represented in children [5–11] or women of reproductive age [12]. An accessory spleen is present in 10% of the population, with one or more splenic masses, with an average diameter of 1 cm, developed in initial phases of fetal life. The most frequent localizations are the splenic hilum and the pancreatic tail [8–12].

Patients with a wandering spleen may be present a movable mass in the abdomen, with chronic or intermittent abdominal pain caused by partial torsion and spontaneous detorsion of the splenic peduncle [10]. Due to the nonspecific and multiple symptoms the clinical diagnosis of both conditions is uncertain even with imaging techniques, such as CT and MRI [13,14].

The first splenectomy for wandering spleen was performed in 1878 by Martin; Soleimani reported in literature only 238 cases

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of wandering spleen from 1895 to 2005 with an age distribution ranging from new-borns to age 81, with 2 peaks, in childhood and in the 3rd decade of life.

The coexistence of the two rare conditions (wandering massive splenomegaly and ectopic wandering spleen) is peculiar such as the age of the patient and the laparoscopic approach.

This case report was conducted, and is reported in accordance with the SCARE criteria [24].

2. Case report

A 17 year-old-woman presented to our Institute with chronic intermittent abdominal pain without nausea vomiting or genitourinary complaints. She had history of minor beta-thalassemia, no trauma or injury. On admission, the patient presented with general condition, no fever, blood pressure 120/80 mmHg, 80 bpm and BMI 17.58.

Laboratory data showed leucopenia ($4.65 \times 10^3/\mu\text{l}$) and a hemoglobin level of 11.6 g/dl, renal, liver function, amylase and lactate were within normal limits.

Examination of the abdomen reveals the triad of classic physical examination findings reported by Gindrey and Piquard in 1996 [11] unveiled a loose mobile intra abdominal slightly tender mass with well defined margins and parenchymatous consistency in right iliac fossa, right lumbar and umbilical region.

On abdominal ultrasonography an expanded spleen arising to the pelvis from right lumbar region was observed.

The abdominal intravenous contrast-enhanced computed tomography (CT) scan abdomen showed an ectopic spleen displaced in right side and mesogastrium of size $15 \times 6 \times 12$ cm with an abnormal wandering pedicle (Figs. 1 and 2). Dilatation of the collateral veins in left ipocondrium and mesenteric site was also recognized, suggesting chronic splenic congestion. Moreover, accompanying report noted an accessory spleen of size 2 cm rear

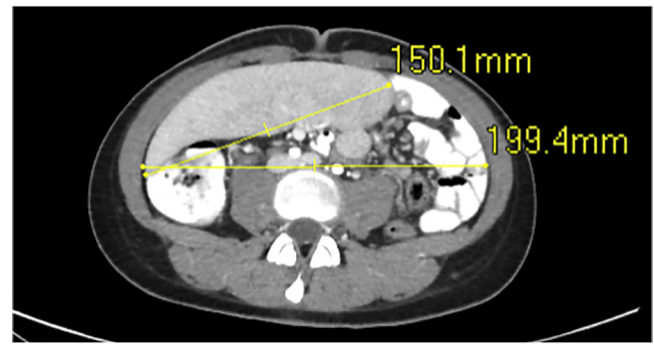


Fig. 1. Preoperative CT scan showed an enormous wandering spleen with dislocation of neighbor organs and an indeed bi-iliac diameter of 19 cm.

left splenic pole with abnormal long pedicle (Fig. 3). The spleen was not visualized in the left upper abdomen.

After a multidisciplinary board (surgeon, radiologist and hematologist) and a careful evaluation of the higher risk for complications (torsion, infarct and rupture) associated to the coexisting wandering and accessory spleen (on the splenic ileus), a laparoscopic splenectomy was performed. Patient's written consent was obtained.

2.1. Surgical technique

The patient was placed supine in Trendelenburg position. A lateral approach was performed: Hasson's-type trocar was inserted on the left anterior axillary line, and pneumoperitoneum was performed to an insufflation pressure of 12 mmHg. The two remaining trocars, 5 and 12 mm, were inserted under laparoscopic control. Laparoscopic exploration showed an enormous spleen with an extremely elongated winding vascular pedicle in mesogastrium, right side and right iliac fossa. Furthermore, accessory spleen on

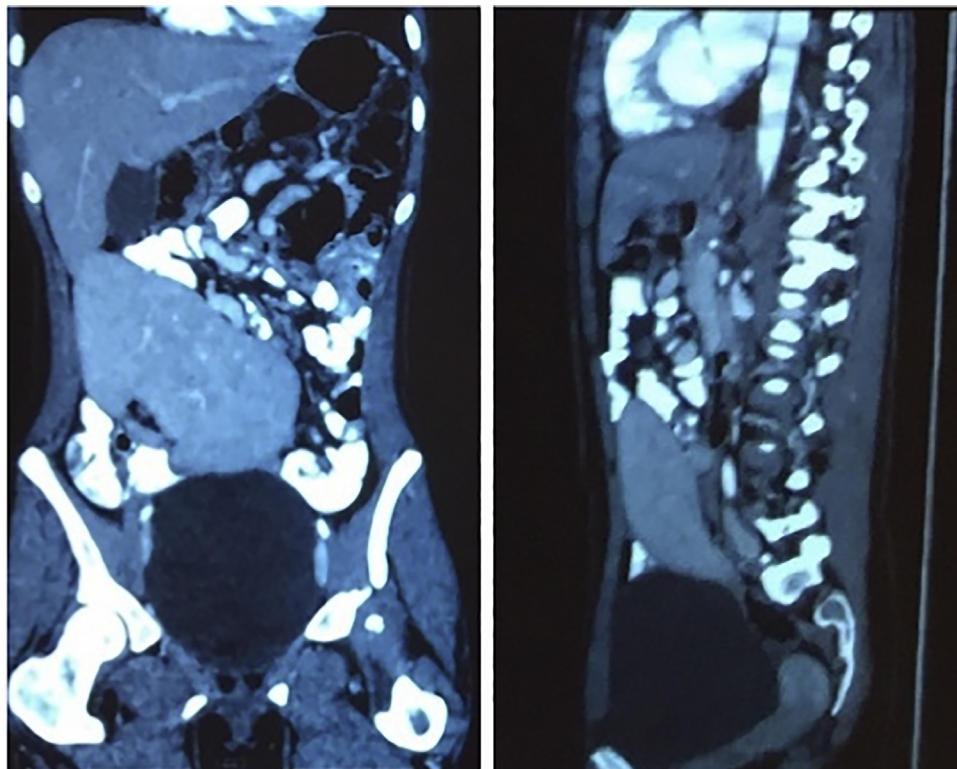


Fig. 2. Preoperative CT scan showed an ectopic spleen displaced in right side and mesogastrium of size $15 \text{ cm} \times 6 \text{ cm} \times 12 \text{ cm}$ with an abnormal wandering pedicle.

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