Congenital Intrahepatic Portal Vein Aneurysm Associated with Left Kidney Hypoplasia as Incidental Imaging Findings in An Adult– A Case Study

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**ABSTRACT**

**Background:** Intrahepatic portal vein aneurysm is a rare condition, considered in less than 3% of all venous aneurysms, and may have a congenital or acquired etiology; another congenital pathology, such as renal hypoplasia occurs in approximately 2.2% of the population, with prenatal or postnatal diagnosis.

**Case Presentation:** We present a rare case to our knowledge, without a correspondent in the literature, which developed both malformations, incidentally diagnosed using Doppler ultrasound and multidetector computed tomography, in an adult female patient with nonspecific abdominal discomfort. The absence of any complications and the stable evolution during three years of follow-up were the arguments for congenital pathology.

**Conclusion:** This case illustrates a couple of rarely associated malformations represented by portal vein aneurysm and renal hypoplasia, the usefulness and limitations of Doppler ultrasound as the first method recommended in the diagnosis and follow-up, and the superior results of multidetector computed tomography as a complementary tool.

**Keywords:** Doppler ultrasonography, multidetector computed tomography, kidney hypoplasia, portal vein aneurysm.

I. BACKGROUND

The embryology of the portal vein occurs between the fourth and 12th week of gestation, related to the vitelline venous system and the umbilical vein [1]. The normal portal vein measures between 10-13 mm in diameter and 6-8 cm in length, and in the cirrhotic liver it can reach up to 19 mm in diameter; portal aneurysm is considered a dilatation of the portal vein above 20 mm [2], and for the intrahepatic branches over 7-8 mm [3]. The anatomical portal vein usually originates from the confluence of the superior mesenteric vein with the splenic vein and divides into the left branch that supplies segments I to IV of the left hepatic lobe and the right, shorter portal branch that forks in the posterior branch that feeds the segments VI and VII, and the anterior branch for segments V and VIII. The typical intrahepatic portal tree occurs in only 65% of cases and many anatomical variants have been described, without functional disorders, but it is important in regulated (controlled) hepatectomy or liver transplantation from a living donor [4]. The rest of 35% cases may present any combination between the main portal vein, left and right branches with the anterior and posterior veins, usually with a bifurcation pattern, but rarely of trifurcation type [5]; in almost all cases, the vessel diameter and the blood flow are within normal limits, without physiological disturbances. However, a certain segment of the extra- or intrahepatic portal vein and of the main branches may have significant congenital dilatations of the aneurysm type, associated or not with abnormal arborizations.

Intrahepatic portal vein aneurysm (PVA) is a rare condition, considered of less 3% from all venous aneurysms [6], but the development of the diagnostic imaging techniques has raised the in vivo diagnoses and the reports in the literature. A large study by Multidetector computed tomography (MDCT) of 4,186 patients found a prevalence of portal venous system aneurysm of 0.43% [7]. The etiology may be congenital or acquired after portal hypertension, cirrhosis, tumor invasion, abdominal trauma, liver biopsy or surgery, necrotizing pancreatitis, incarcerated diaphragmatic hernia, etc.; sometimes the pathogenesis is difficult to specify. However, the stability over time, the absence of any associated disease and the absence of clinically significant manifestations are considered to be arguments for the congenital type. The adaptation of the intrahepatic flow in congenital aneurysms allows a normal hepatic function without supplementary risk of complications whatever the patient’s age, being described five cases in asymptomatic patients of 80-year-old and older [8]. Some cases presented in the literature developed symptoms correlated with the huge size of the PVA of up to 5-7 cm size, of probably congenital origin, such as encephalopathy [9], jaundice, or epigastric pain in an acute thrombose [10]; but almost ¾ cases, usually with small or moderate PVA, showed no symptoms related to its pathology [7]. Intrauterine diagnosis by Doppler ultrasound of the PVA is evidence of congenital etiology [11], but the risk of false negative diagnosis is considered...
increased. PVA may develop rarely complications, such as thrombosis, rupture, and compression of adjacent structures, and a sonographic follow-up is considered the best attitude for a watchful waiting in non-complicated cases [12].

Renal hypoplasia is defined as a congenital small kidney without dysplasia, and may be functional depending on size and vascular supply. The size of an abnormally small kidney is defined below two standard deviations of that of age-matched normal individuals or a combined renal volume less than half that normal. The corticomedullary differentiation is present with a reduced number of nephrons. The reciprocal induction between the metanephric and the ureteric cell lineage occurs after the 5th-6th weeks of gestation and ends in the embryonic stage up to the 10th weeks, with a final number of nephrons; the kidney development continues during the gestation and maturation is completed in the postnatal life.

Due to the difficulty of imaging diagnosis to differentiate congenital renal hypoplasia from renal dysplasia and acquired atrophy, the actual incidence is unknown, but it is considered present in about 2.2% of the population, while some epidemiological studies state an incidence of 1 in 400 births [13 cited by 14]. Many gene mutations were identified, but some intrauterine environmental, metabolic factors and medicine drugs (for seizures, blood hypertension, tranquilizers, painkillers, dexamethasone, etc.) were incriminated, too [14]. in unilateral renal hypoplasia, if the contralateral kidney is healthy, it will develop compensatory hypertrophy and clinical manifestations may be absent. In other cases, urinary tract infections, lithiasis, chronic kidney failure, or high blood pressure may occur.

II. Case Presentation

We present the case of a female patient with a couple of different rare vascular malformations that developed in the first trimester and were incidentally discovered in adulthood.

The patient’s history mentions a single cesarean birth at the age of 31, and a unique congenital right kidney discovered incidentally at routine ultrasonography during pregnancy. Asymptomatic liver with normal biological tests did not require any prior indication for liver imaging, including during pregnancy.

At the presentation in our Service of Ultrasonography at the age of 41, the patient was referred for a follow-up examination of the unique considered kidney and a general abdominal and pelvic evaluation. The initial physical examination was unremarkable: normal body mass index (BMI), non-alcoholic and non-smoker; we highlight a normal blood pressure. The clinical symptoms were related to abdominal discomfort, nausea, bloating, without weight loss or intestinal transit disorders, followed by long intervals of normal condition; in the last year the moderate pale skin was associated with menstrual disorders, represented by menometrorrhagia.

The first imaging test was an abdominal and pelvic Doppler ultrasonography, that illustrated a moderate increase in the liver measurements, with predominance of the left lobe and the caudate lobe, hyperechoic texture with a small increase in the ultrasound attenuation, no biliary duct abnormalities, normal gallbladder, thin suprahepatic veins. The portal tree walls were thickened hyperechoic, with an aneurysmal enlargement of the intrahepatic portal vein up to 22.1mm diameter (Fig.1a), and of its main branches up to 18mm. The diameter of the splenic-mesenteric confluent measured 18mm, too, and the splenic vein measured 10mm, without splenomegaly. The Doppler exam revealed the double sense turbulent flow in the PVA, the so-called yin-yang sign (Fig. 1a-d). No peritoneal abnormal fluid was salient.

Additionally, the ultrasound examination confirmed the absence of the left kidney mass on the left lumbar region and in all spaces susceptible to an ectopy available to ultrasound windows; the right kidney presented compensatory hypertrophy of 285 cm3 (normal range in woman ultrasound 120-140 cm3), with a hypertrophied column of Bertin that was splitting the renal sinus (bifid pelvis, more an anatomic variant than a true renal malformation), thickening of the renal parenchyma up to 2.4 cm (normal media of 1.8 cm), normal vasculature, and normal pyelocaliceal system and upper ureter (Fig. 2).
In the pelvic cavity, the normal bladder presented a unique right unilateral urinary jet, while the enlarged uterus demonstrated changes of adenomyosis with endometrial hypoplasia in both the proliferative and secretory phases (Fig. 3a), associated with the aspect of anovulatory menstrual cycle (Fig. 3b), explaining the menometrorrhagia.

The paraclinical status revealed no markers of hepatitis viruses, but a mild increase in gamma-glutamyl transferase (GGT) of 56.77 U/l (normal range 0–55 U/l), unusual in isolated PVA but higher values in association hepatocellular carcinoma [15]; alanine aminotransferase (ALAT) had increased values, too, up to 50.52 u/l (normal range 0–41 u/l), but normal aspartate aminotransferase (AST); the hepatoprotective treatment resulted in normalizing of these enzymes. The serum urea and creatinine were in normal limits, while a mild microcytic anemia was treatment-resistant: hemoglobin of 10.3–10.8 g/dL (normal 12.5–15.5 g/dL), hematocrit of 31.9%–34.6% (normal 37%–47%), mean corpuscular volume (MCV) of 71.4–75.5 fl (82–98 fl), mean corpuscular hemoglobin (MCH) of 23.0–23.1 pg (26–34 pg). The rest of the biological examinations showed no significant data, except for a small increase in total cholesterol of 229.41 mg/dL (0–200 mg/dL) and HDL cholesterol of 92.07 mg/dL (60–90 mg/dL), which had constant values.

Fig. 2. Color-flow Doppler in longitudinal and axial views of the right kidney allowing the measurement of the increase in renal volume and parenchyma thickness, with a normal vascular mapping (a). The PWD acquisition (b) illustrates a spectral flow of central type with a low resistance index (RI 0.50), an argument of the functional compensatory kidney hypertrophy.

Fig. 3. Transabdominal pelvic ultrasound in sagittal and axial views illustrate a retroverted, enlarged uterus of 490cm3 (normal range 70–80cm3), thickening of the transitional zone (inner myometrium) with hyperechoic punctate endometrial extensions, and hypoplastic intracavitary endometrium with hyperechoic aspect in the secretory (luteal) phase (a). The right ovary with a few infracentimetric follicles (b), without any luteal cyst (the left ovary with similar findings is not shown) and the absence of peritoneal fluid conclusive for an anovulatory cycle.

Fig. 4. MDCT with VR technique in the late arterial phase (a) and in the portal phase (b) illustrate the PVA (green arrows) up to the 23.9mm and the superior mesenteric vein (black arrow); in addition, there is only the right kidney nephrogram with its prominent artery and a short, dilated vein of 19.1mm (white arrow), while the nephrogram of the left kidney is not visible. The VR technique demonstrates a rare trifurcation type of portal vein, without thrombosis.
Further evaluation, represented by a complementary contrast MDCT, confirmed the presence of the intrahepatic PVA, without tumoral changes, portal thromboses, or signs of portal hypertension. The volume rendering (VR) method provided a better illustration and understanding of the portal tree with its rare trifurcation type, and the presence of a single functional right kidney with a short and enlarged renal vein (Fig.4). However, standard contrast multiplanar MDCT showed a small left kidney located in the upper-internal region of the left lumbar retroperitoneal space, which was an ultrasound masked from the ribs, pancreatic tail, spleen and small bowel loops and showed normal morphology, but a filiform renal artery, a weak nephrogram and no obvious excretory phase; yet, the left renal vein was better developed (Fig. 5). In the pelvis, the uterine enlargement had less specific aspect in MDCT, and no salient adnexal changes were visualized.

We did not notice significant hepatic changes during three-year ultrasound follow-ups, but a small increase in the diameter of the PVA, without thrombosis (Fig. 6, Fig.7).
III. DISCUSSION

Two congenital malformations with different vascular pathogenies represent the particularity of this case: a PVA, and a left unilateral renal artery hypoplasia resulting in a hypoplastic unfunctional left kidney. Both malformations issued in the first trimester of pregnancy, and had compensatory mechanisms, with nonspecific clinical symptoms developing lately in adulthood. Biological changes that have shown hepatic dysfunction may be related to inadequate nutrition, ignoring hepatic vascular pathology until adulthood, resulting in mild hepatomegaly with an ultrasonographic aspect suggesting hepatic steatosis, associated with slightly altered lipidogram and with reversible evolution under treatment; the PVA appears asymptomatic in this case. Similarly, no biological alterations of the renal function, and no blood hypertension were developed associated with the severe unilateral kidney hypoplasia. Additionally, the patient had acquired pathologies with clinical expression, such as the constant mild iron deficiency type of microcytic anemia, which was attributed to the menometrorrhagia; the last one was related to the uterine adenomyosis and to the premenopausal menstrual disorders with ultrasound changes suggestive of ovarian follicular apoptosis and anovulatory cycles. The pathogenic of the uterine adenomyosis is generally considered unknown, but recent studies sustain that there is a strong association between endometrial epithelial KRAS-gene mutations (Kirsten rat sarcoma gene; it is the most commonly mutated oncogene in human cancers), causing the alteration of the basal layer of the myometrium and conducting on the development of adenomyosis and endometriosis [16]. This opens the possibility of considering in the case presented above adenomyosis as another congenital alteration associated with the two recognized malformations described and opens the way for new studies.

The limitations of this approach are related to the late diagnosis of PVA at the age of 41, the absence of the previously documented personal imaging history, the absence of predecessor relatives’ examinations, and absence of genetic tests, to analyze a possible inheritance mechanism; the exceptionally rare association of portal and renal malformations was not inherited by the only female child and no information could be obtained on the anatomical condition of the patient’s parents.

This case presentation illustrates the usefulness of ultrasonography in the evaluation of hepatic vascularization by Doppler study, including PVA, in the detection and follow-up of uncomplicated cases, which are recommended for watchful waiting because ultrasound is a harmless technique without the need for a contrast agent; however, MDCT remains the best diagnostic tool, allowing vascular volumetric reconstruction and better global risk assessment. For renal hypoplasia, this case highlights the risk of misdiagnosis by ultrasound as renal agenesis due to the “blind” areas of the retroperitoneal space; it is recommended to keep the method in the control examinations, without risk of irradiation and without a mandatory injection of a contrast agent, sometimes contraindicated. As a rule, because there is an increased risk of multiple malformations, in any pregnant woman with a known/newly detected maternal malformation, even asymptomatic, a complete ultrasound examination of the abdomen is required, supplemented by a more complex imaging examination after childbirth. Because the maternal imaging screening is not a rule, many congenital anomalies may be neglected, and some supplementary risks induced by the pregnancy may be ignored. In the case presented above, the routine ultrasound in pregnancy detected only some kidney abnormality, while the PVA remained unknown. MDCT with standard multiplanar reconstructions provided more accurate information, and VR technique should be considered the only additional tool, because its limits in cases...
of hypo vascular structures. Magnetic resonance imaging is not a routine method and remains a complementary tool reserved for special cases.

IV. CONCLUSIONS

Incidental findings of congenital malformations in adults are becoming more common with the development of diagnostic imaging techniques. The existence of rare malformations, unexpected at routine examinations, poses problems of diagnosis and therapeutic medical conduct, with the option of watchful waiting or therapeutic intervention in the near future. When there is an association of rare malformations, as in the case presented above, each reported experience can contribute to the establishment of optimized behavior, while avoiding diagnostic or treatment errors. Some pathologies that have not yet elucidated their mechanism or are being researched, such as adenomyosis, and imaging examinations can make a valuable contribution.

In this case report, despite two major malformations and an associated uterine adenomyosis that induced microcytic anemia, the lack of significant complications leads to a normal life expectancy, so clinical and imaging surveillance was the preferred attitude.

V. ABBREVIATIONS

ALAT: Alanine aminotransferase
AST: Aspartate aminotransferase
BMI: Body mass index
GGT: Gamma-glutamyl transferase
KRAS: Kirsten rat sarcoma viral oncogene homolog
MCH: Mean corpuscular hemoglobin
MCV: Mean corpuscular volume
MDCT: Multidetector computed tomography
PVA: Portal vein aneurysm
PVD: Pulsed Wave Doppler acquisition
RI: Resistance index
VR: Volume rendering

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CONFLICT OF INTEREST

The author declares that there are no conflicts of interest regarding the publication of this article.

The patient gave her informed consent in writing for the publication of this article as a medical anonymized case report for scientific purposes.

REFERENCES


A. Colan-Georges was born near Craiova, Romania, in 1959. She graduated the Faculty of Medicine of Craiova (1984), and became senior radiologist in 1990, earning all professional degrees in this specialty under the accreditation of the Craiova University of Medicine and Pharmacy. She followed many courses of perfecting in Bucharest, Romania, followed by in-depth studies in Radiology (“Attestation de Formation Spécialisée Approfondie, mention Radiodiagnostic et Imagerie Médicale”) at the Faculty Saint Antoine of the University Pierre and Marie Curie, Paris, France (2001), and a practical stage of breast Ultrasonography and Strain SonoeLastography (“Attestation de stage pratique d’Echographie du Sein”) in the Francophone Center of Formation in Ultrasonography, Aix-en-Provence, accredited by the University of Nîmes, France (2007). In 2006 she earned a PhD in Medical Sciences, with the thesis “Radiological-imaging diagnosis of female infertility of pelvic etiology”.

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