

Common And External Iliac Artery Agensis And Absence Of The Infrarenal Vena Cava Combined With Other Congenital Anomalies Incidentally Discovered During MDCT Evaluation Of Pelvic Varicocele.

L Aquiriano, J Balaguera, M Sarró Cañizares¹, M Continente, L Fanjul, T Camaño

Citation

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Abstract

We present a case of an 18 year-old woman with previous history of congenital cardiopathy, surgically treated in the infancy, referred for multidetector-row CT (MDCT) evaluation of chronic pelvic pain. Pelvic congestion syndrome caused by pelvic varicocele was demonstrated and multiple congenital anomalies (agenesis of the left common and external iliac artery, absence of the infrarenal vena cava, septate uterus and congenital spinal malformations including a thoracic butterfly vertebra, coccygeal agenesis and sacral hypoplasia) were incidentally discovered. To our knowledge, such a combination of congenital anomalies has not previously been documented in the literature.

INTRODUCTION

Many progresses have been achieved in the last years in the investigation of the causes of congenital malformations, however most of them (40- 60%), don't have still a satisfactory explanation. The 50-60% of the congenital malformations are produced for genetic factors (chromosomal anomalies or mutant genes), 10% are produced by environmental factors (infectious agents, teratogenic medications, chemical agents, radiation) and 20-25% are the result of the interaction among genetic and environmental factors (multifactorial etiology)¹.

In the last years different multiple congenital anomalies (MCA) syndromes, have been

described thanks to the evolution of the image techniques that has allowed a morphological better analysis and to the readiness of new genetic data.

The advances in the techniques of molecular biology, as the cariotip fluorescen in situ hybridization (FISH), offers now opportunities understand the cause of many genetic disease².

With everything in occasions is not possible to establish the origin of the multiple malformation.

We present an 18 year-old woman with a syndrome of MCA, with pregnancy and childbirth normal. This syndrome was detected incidentally after the study by chronic abdominal pain. We think that it can be the first case published with these characteristics after review the literature until today.

CASE REPORT

An 18-year-old nulliparous female presented with pain on the right side of the pelvis for more than 6 months. She was referred to perform a CT scan of the abdomen and pelvis for the evaluation of pelvic varicocele and to rule out adnexal mass. The pain was of variable intensity and was not exacerbated by movement as running, jumping or bending down. The patient also complained of premenstrual tension. She had no mictional or gastrointestinal symptoms. At the age of 15 months the patient underwent successful surgical intervention for a congenital cardiopathy consistent with double outlet right ventricle, inter-ventricular communication and banding of the pulmonary artery.

On physical examination, pulmonary and neurological exploration was normal. She presented a soft and painful abdomen in the right lower quadrant without signs of

peritoneal irritation and without palpable masses. In the lower extremities, there were no delayed capillary refill, no pallor or varicosities. Arterial pulsations at the femoral, popliteal and dorsalis pedis arteries were normal. Laboratory tests were unremarkable.

Portal phase contrast-enhanced MDCT with multiplanar reformation (MPR) and maximum intensity projection (MIP) images of the abdomen and pelvis showed bilateral dilated ovarian veins and congenital absence of the infrarenal vena cava (Figure 1). Enlarged ascending lumbar veins on both sides were seen continuous with the iliac veins, draining blood flow to the azygos and hemiazygos system (Figure 2). Müllerian duct anomaly consistent with septate uterus and multiple prominent varicose veins surrounding the uterus were also demonstrated (Figure 3). CT scans of the pelvis showed agenesis of the left common iliac artery, absence of the external iliac artery except for a short distal segment and a well-developed left internal iliac artery. Bone window CT images revealed T11 butterfly (sagittal cleft) vertebra associated with absence of T11/12 intervertebral disk, dorsal and lumbar scoliosis, normal development of only three sacral vertebrae and agenesis of coccygeal vertebrae (Figure 4).

{image:1}

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MDCT angiography of the aortoiliac system was performed subsequently and post-processing three-dimensional volume rendering and MIP images were acquired. The left internal iliac artery was normal in caliber with reformation by collateral vessels arising from dilated left fourth and fifth lumbar arteries and from anastomoses between an enlarged middle sacral artery and the left lateral sacral artery (Figure 5a). A vessel interpreted as the distal segment of the external iliac artery was seen running over the superior pubic ramus, continuous with the common left femoral artery, which was normal in caliber. The distal end of the external iliac artery was reconstituted from branches of the left internal iliac artery and left fourth lumbar artery. A normal course of the deep iliac circumflex artery and anastomosis of the superior and inferior epigastric artery were demonstrated, which may provide a collateral pathway for reformation of the external iliac artery (Figure 5b).

{image:5}

The diagnosis was of pelvic congestion syndrome due to pelvic varicocele in the setting of agenesis of infrarenal vena cava, combined with multiple congenital anomalies involving the left iliac arterial system, the uterus and the thoracic and sacrococcygeal spine.

DISCUSSION

Among congenital defects, the most common are congenital heart defects, which constitute a heterogeneous group with a multifactor etiology. There are more than 50 different types of congenital heart lesions. However, frequently there is combination of several lesions in oneself patient, as the case that we present (interventricular communication, double outlet right ventricle and banding of the pulmonary artery). Congenital cardiopathies can be an isolated event or can happen in combination with other malformations being a part of a syndrome. Congenital cardiopathy is associated to anomalies in another organ or system in 54% of the cases and to anomalies of several organs and systems in 46%³. Our patient presented a combination of cardiovascular, uterine and musculoskeletal congenital defects.

Congenital malformations of the iliofemoral system are quite uncommon, with most discovered at autopsy^{4,6} or incidentally, as in our case. Greeb⁷ described only 6 cases in a series of about 8000 patients who underwent angiography of the pelvis. Although congenital absence of external iliac artery can exist as an isolated finding, chromosomal abnormalities or major congenital malformations associated with it have been reported. Seghezzi et al⁸. reported a case of congenital hypoplasia of the right external iliac artery, congenital megacolon, anal atresia and hypospadias.

Congenital malformation of the external iliac artery has been classified into three groups by Tamisier et al.⁹: group 1, anomalies in origin or course of the artery; group 2, hypoplasia or atresia compensated for by persistent sciatic artery; and group 3, isolated hypoplasia or atresia which can occasionally cause chronic ischemia of the lower limbs. Group 1 disorders are unlikely to cause chronic ischemia of the leg. The patient that we present had no episode suggestive of leg ischemia and required no physical restriction for exercise such as climbing or swimming.

Due to the complexity of the development stages, the vena cava may undergo a very large number of congenital

anomalies. Agenesis of the infrarenal vena cava is usually associated with other congenital malformations of diverse nature until in 50% of the cases; therefore, it is very important to discard another anomaly as soon as it is discovered. Among these anomalies, they are: cardiovascular defects until in 51.2% of the cases¹⁰ (dextrocardia, levocardia, inter-atrial communication, atrium-ventricular channel and stenosis of the lung artery), anomalous rotation of abdominal viscera, lung dysgenesis, aplasia of the right kidney and polysplenia syndrome¹¹.

In cases with congenital absence of the infrarenal vena cava, the external and internal iliac veins join to form enlarged ascending lumbar veins, which convey blood return from the lower extremities to the azygos and hemiazygos veins via anterior paravertebral collateral veins. The collateral pathways most often observed in these cases are intervertebral veins, epidural venous plexus, inferior and superficial epigastric veins and gonadal and periuterine veins¹². Symptoms may be related to a slower return circulation due to the derivation forced from the lower extremities to the azygos vein. These symptoms include lower extremity chronic venous insufficiency, abdominal wall collateral veins, deep venous thrombosis, lung thromboembolism, haemoptysis due to the increase of pressure in the azygos vein and pelvic varicose veins, as in our case¹³.

Physical symptoms of pelvic congestion due to pelvic varicocele causing chronic pelvic pain have been well-documented. The etiology is reflux in incompetent dilated ovarian and uterine veins. Abnormally dilated vessels and collateral pathways may be seen as a result of increase of the return pressure associated with congenital absence of the infrarenal vena cava^{14,15}. Affected patients are typically in their late 20s or early 30s. Pelvic congestion with varicosities in the infundibulopelvic and broad ligaments, draining via ovarian and internal iliac vein tributaries, can cause unilateral or bilateral dull pain in the pelvis. Pain can be worsened by walking and postural changes, can be cyclic with dysfunctional bleeding and dysmenorrhea or may be accompanied by dyspareunia or postcoital ache that may last for hours or days¹⁶.

Another congenital defect in this case report was the septate uterus. This anomaly is caused by nonresorption of the sagittal uterine septum¹⁷. Congenital uterine malformations are variable in frequency and are usually estimated to occur in 3 to 4% in the general population, although less than half

manifest clinical symptoms. The frequency of symptomatic malformation is dominated by septate uterus (close to 50%) compared with other malformations¹⁸. Complete uterine and vaginal septum are two müllerian duct anomalies¹⁹. These defects are associated with a higher incidence of other congenital anomaly than in the normal population (10%), including anomaly of the urinary tract (20-25%), gastrointestinal tract (12%), musculoskeletal system (10-12%), heart, eye and ear (6%)²⁰.

Among the several congenital anomalies that the patient presented, the abnormal development of the vertebral bodies in the thoracic and sacrococcygeal spine stand out in the musculoskeletal system. Congenital malformations of the pelvic waist can take place in the setting of a systemic disease of the musculoskeletal system or in the mark of a local or regional malformation. The locoregional disease is part of the group of the dysostosis. Because the dysostosis are frequently associated to visceral, urinary or neurological anomalies, they are known as the caudal regression syndrome. This syndrome comprises from an asymptomatic aplasia/agenesis of the coccyx to a partial or total sacral agenesis, lumbar anomaly and even dorsal defect^{21,22}. The case reported here did not showed visceral, urinary or neurological alteration.

In conclusion, common and external iliac artery agenesis in association with absence of the infrarenal vena cava, congenital cardiopathy, müllerian duct and spine congenital anomalies is exceedingly rare. To the best of our knowledge, a syndrome with such a combination of anomalies has previously not been reported in the literature. Interestingly, clinical symptoms were uniquely related to chronic pelvic pain because of pelvic varicocele and many of the findings described above were incidentally discovered.

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Author Information

LA Albeniz Aquiriano

Physician Assistant Radiology Service, Service of Radiology and Service of General and Digestive Surgery Hospital Central de la Cruz Roja San José y Santa Adela Madrid Spain

J.Carvajal Balaguera

Surgeon Assistant Surgery Service, Service of Radiology and Service of General and Digestive Surgery Hospital Central de la Cruz Roja San José y Santa Adela Madrid Spain

M. Sarró Cañizares¹

Physician Assistant Radiology Service, Service of Radiology and Service of General and Digestive Surgery Hospital Central de la Cruz Roja San José y Santa Adela Madrid Spain

M^a. J. Sanz Continente

Physician Assistant Radiology Service, Service of Radiology and Service of General and Digestive Surgery Hospital Central de la Cruz Roja San José y Santa Adela Madrid Spain

L. Tomas Fanjul

Physician Assistant Radiology Service, Service of Radiology and Service of General and Digestive Surgery Hospital Central de la Cruz Roja San José y Santa Adela Madrid Spain

T. Martín Camaño

Chief Radiology Service, Service of Radiology and Service of General and Digestive Surgery Hospital Central de la Cruz Roja San José y Santa Adela Madrid Spain