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Case Report

Imaging Diagnosis of Situs Inversus, Persistence of the Left Superior Vena Cava, Renal Agenesis and Anomaly of the Retroperitoneal Vessels in Niamey: A Case Report

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Abstract: Introduction: Situs inversus is a rare congenital malformation. It can be associated with other malformations. It is often discovered incidentally by medical imaging. The latter also makes it possible to take stock of all associated malformations. Observation: The authors report a particular association in a 4-year-old male patient. Following a transthoracic cardiac Doppler ultrasound indicated for dyspnea at the slightest effort. The examination revealed dextrocardia, minimal membranous septum interventricular communication with left-right shunt, ostium primum inter-atrial communication measured at 6 mm with moderate impact on the left atrium, good systolic function of both ventricles, thin heart valves, dry pericardium. The chest x-ray requested systematically highlighted dextrocardia, enlargement of the upper mediastinum, and a gastric air pocket visible under the right diaphragmatic dome. The thoraco-abdomino-pelvic scan carried out made it possible to demonstrate that the situs inversus was complete and associated with other anomalies. It was a type 3b superior cava venous return anomaly with a single left kidney and an anomaly of the retroperitoneal vessels. Conclusion: Situs inversus is a rare and particular congenital anomaly because it can be associated with several types of malformations, sometimes very complex. Hence the interest in carrying out a complete imaging assessment in cases of situs inversus.

Keywords: Situs Inversus, Congenital Heart Disease, PVCSG, Renal Agenesis, Retroperitoneal Vessel Anomaly, Left IVC, Niamey

1. Introduction

Situs inversus also called “situs transversus” or “oppositus” is a congenital condition in which the major visceral organs are inverted or mirrored from their normal anatomical positions. In humans, it designates an inversion of the position of the viscera in relation to the sagittal plane. It is the mirror image of the situs solitus which is the usual anatomical configuration of the thoracoabdominal viscera [1]. This congenital anomaly affects the thoracic and/or abdominal viscera; it’s called “situs inversus totalis” when it concerns the thoracic and abdominal organs [2]. In addition, other anomalies can be observed associated with situs inversus, whether total or partial, which can often be indicative of this particular anatomy. the reported case is about a 4-year-old patient admitted for additional assessment of congenital heart disease in whom the diagnosis established thanks to chest x-ray and thoraco-abdomino-pelvic scan is an association of situs inversus with others. rare malformations.

2. Patient and Observation

This is a male patient, aged 4 years at the time of diagnosis. He had undergone a transthoracic cardiac Doppler ultrasound indicated for dyspnea at the slightest effort. The cardiac Doppler ultrasound showed:

1) Dextrocardia,
2) An interventricular communication of the minimal membranous septum with left-right shunt,
3) An ostium primum inter-atrial communication measured at 6 mm with moderate impact on the left atrium,
4) Good systolic function of both ventricles,
5) Fine heart valves,
6) Dry pericardium,
7) No visible vegetation.

In view of these results he was sent to the medical imaging department for further malformation assessment. The medical imaging examinations carried out were: chest x-ray and TAP CT.

The routinely requested frontal chest x-ray showed dextrocardia, widening of the upper mediastinum, and a gastric air pocket visible under the right diaphragmatic dome (Figure 1).

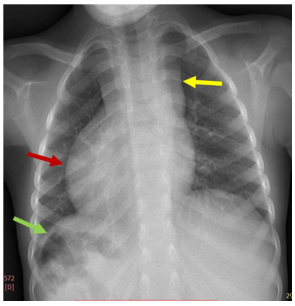

Figure 1. *Lung x-ray performed standing upright in a 4-year-old patient showing dextrocardia (red arrow), widening of the upper mediastinum (yellow arrow), a gastric air pocket visible under the right diaphragmatic dome (green arrow).*

The TAP CT was carried out with a Hitachi® Supria brand scanner put into service on October 1, 2017. The constants used were 80KV and 30mA and the cuts made were 3 mm contiguous. The examination was carried out in a fasting patient, in the supine position, head first. Two series of acquisitions were carried out, one of which without injection of contrast product and the other with injection of contrast product showed:

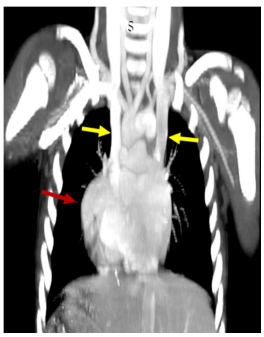

Figure 2. *Thoracic CT in coronal section, mediastinal window, with PDC injection showing two right and left superior vena cavae (yellow arrow), heart on the right (red arrow).*

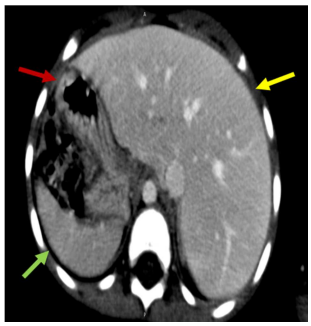

Figure 3. *Abdominal CT in axial section, at the height of the hypochondria, with injection of the PDC, in the portal phase showing the liver on the left (yellow arrow), the spleen (green arrow) and the stomach (red arrow) on the right.*

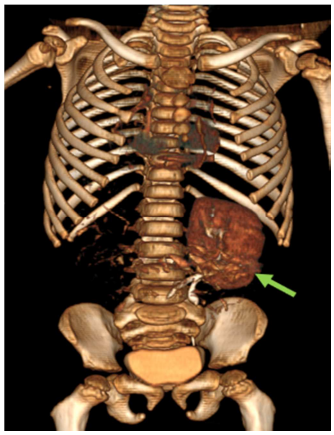

Figure 4. *3D reconstruction showing a single left kidney (green arrow).*

At the thoracic level: three pulmonary lobes on the left and two pulmonary lobes on the right for the lungs (figures 2 and 3), the heart on the right, the aorta on the right, an anomaly of the superior cava venous return type IIIb (figure 4).

In the abdomen: liver on the left, spleen and stomach on the right (figure 3); malformation of the urinary tract with a single left kidney (figure 4); anomaly of the retroperitoneal vessels (the abdominal aorta and the inferior vena cava cross the midline at the level of L2) (figure 5).

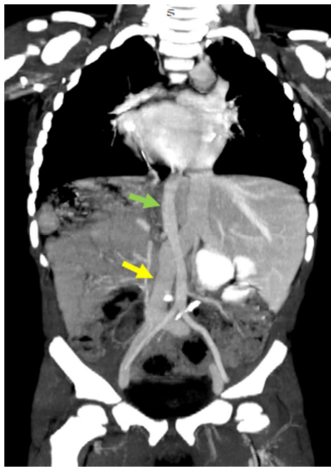

Figure 5. *Thoraco-abdominal CT in coronal MPR section, with PDC injection showing the crossing of the abdominal aorta and the inferior vena cava on the midline, at the level of L2.*

3. Discussion

Situs inversus is a rare congenital malformation, found in approximately 0.01% of the population with a male/female ratio of 3:2 [3, 4]. The mode of inheritance would be autosomal recessive, the genetic mechanism not yet being precise [1, 4]. It is often a chance discovery. Patients with this anomaly usually lead a normal life [5]. Many people with situs inversus totalis are unaware of their unusual anatomy until they seek medical care for a reason unrelated to that anatomy. In certain cases it can be revealed by signs related to malformations that may be associated with it.

In Western countries and the United States, several studies have been devoted to situs inversus, which has not been the case in Africa [5-10]. Cases have been reported associated with other malformations [6, 7] but no study has reported such an association as seen in our patient.

As described by several authors, the chest x-ray confirmed in our observation dextrocardia, the presence of the gastric air pocket under the right diaphragmatic dome thus stipulate a situs inversus totalis; but a widening of the superior mediastinum was also observed. This clinical picture led us to indicate exploration by a thoraco-abdominopelvic scanner.

On CT scan, the presence of two superior vena cavas on the right and left was observed at the thoracic level. Indeed, some authors were already divided on the relationship between the presence of an enlargement of the superior mediastinum and the presence of a persistence of the left superior vena cava on the chest x-ray: according to certain authors an enlargement of the superior mediastinum (enlargement of the aortic shadow and an overhang of the left superior mediastinum) can be observed in relation to the presence of the left superior vena cava [11, 12]; on the other hand, for others the diagnosis is difficult using chest radiography, because in certain situations certain normal images do not exclude the diagnosis [13]. All this underlines the interest of CT which allowed us to make the anatomical assessment of this picture with the presence of two superior vena cavae without any connection between them corresponding to a PVCSG type III B [14] thus constituting the first interest of our study. PVCSG is a malformation due to the persistence of the left anterior cardinal vein. This malformation is the most common abnormality of systemic venous return [15]. They are detected in approximately 3% of autopsies [16] and in 1.3 to 4.5% [14, 17] of patients with congenital heart disease. The incidence in the general population is estimated at approximately 0.3 to 0.5% [18-21].

The second interest of our study lies in the association of situs inversus with an anomaly of the retroperitoneal vessels; the abdominal aorta and the inferior vena cava crossing in the midline at the level of L2. Anomalies of the retroperitoneal vessels and especially the inferior vena cava are rare. During the period of early embryogenesis, venous drainage of the left and right sides of the body occurs independently of each other. After the regression of the majority of the left supracardinal veins and the veins of the interconnection between the sacrocardinal veins, the entire venous drainage of the left lower limb is towards the right side, thus forming the IVC. Disturbance during this process of venous development can lead to anatomical variations in position, such as the left IVC [22]. It is a rare anatomical variant with an incidence of 0.2 to 0.5% in the population [22]. They can be discovered by chance [22]. In our patient, the IVC initially on the right side of the spine crosses the posterior surface of the abdominal aorta at L2 to form the 'normal' portion of the suprarenal IVC but on the left side of the spine; corresponding to a 'left IVC'. In the majority of cases, the left IVC crosses the anterior surface of the abdominal aorta at the level of the left renal vein to form the normal portion of the suprarenal IVC [23-24]. the same observations were made with the patient but in reverse mode because of the situs inversus. IVC anomaly is found in 0.6% of patients with cardiac malformations [25]. This was the case in our patient with a complex heart malformation.

The third interest of our study is the association of situs inversus with an abnormal position of the IVC and right renal agenesis. Malformation of the IVC has been found by certain authors to be associated with kidney hypotrophy [26]; but in our patient renal agenesis was noted. Congenital anomalies of the kidneys and urinary tract being a public health issue, they represent 1/200 live births, they account for 20 to 30% of all malformations and are responsible for 30 to 50% of end-stage renal failure in the child. Not all have a poor prognosis, some even being of the anatomical variant [27]. Renal agenesis, that is to say the complete absence of development of the kidney, can be uni- or bilateral. Unilateral renal agenesis is quite common (1 to 2/1000); it has a good prognosis even if it causes glomerular hyperfiltration in 20 to 30% of cases, likely to cause a certain clinical symptomatology [28, 29]. Despite its more or less high frequency, its relationship with situs inversus has not yet been established.

4. Conclusion

Situs inversus is a rare congenital malformation, often discovered incidentally but can be revealed by other malformations. The particularity of our observation is its association with several malformations, each rarer than the other: unilateral renal agenesis, PVCSG, anomalies of the retroperitoneal vessels. CT remains at the center of all these diagnoses.

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Conflicts of Interest

The authors declare no conflict of interest.

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