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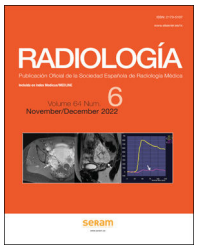

SCIENTIFIC LETTER

Persistent left superior vena cava: clinical and radiological significance

Vena cava superior izquierda persistente: importancia clínica y radiológica

Persistent left superior vena cava (PLSVC) is the most common thoracic venous drainage anomaly and is often associated with other congenital cardiac abnormalities.[1] It is usually found incidentally during central venous catheter insertion and follows an indolent course. However, in a small number of patients in whom the vein drains into the left atrium, a right-to-left shunt occurs, increasing the risk of thromboembolic events[1,2] and other complications, such as cyanosis or heart failure, if the shunt is sufficiently significant.[3,4]

We present the case of a 63-year-old male with no relevant medical history who attended the emergency department with a tension headache with warning signs and fever. A non-contrast brain CT scan was performed and, after administration of intravenous iodinated contrast, a right frontal lesion measuring 2.6 cm in diameter with ring enhancement, expansion effect and associated vasogenic oedema was observed (Fig. 1A). Subsequently, MRI confirmed the lesion as an abscess (Fig. 1B, T1 contrast-enhanced sequence) with signs of ventriculitis (Fig. 1C and D, diffusion and ADC sequences, respectively).

The patient showed no evidence of infection in other areas, including ENT-related ones, nor clinical or echocardiographic signs of endocarditis, immunodeficiencies or other medical or surgical history of interest. In order to identify systemic involvement or possible causes, such as shunts, a thoracoabdominal-pelvic CT scan was performed in the venous-portal phase without cardiac synchronisation, which showed a PLSVC with direct drainage into the left atrium (Fig. 2A–D: sagittal, coronal and volumetric reconstructions, respectively).

The patient required a prolonged stay in the ICU, broad-spectrum antibiotic treatment and surgical intervention in the form of craniotomy to drain the brain abscess. He evolved unfavourably due to the development of hydrocephalus and multiple infectious complications, the most significant of which were respiratory. The patient's poor general condition did not allow correction of the cardiac shunt and he eventually died.

The first reports of PLSVC date back to the 18th century, but the first major description was made in 1850 by Marshal.[1] PLSVC is seen in 0.3–0.5% of the general population and in 12% of patients with congenital heart disease, making it the most common congenital abnormality of the thoracic venous system.[2] The course of PLSVC is usually benign, but if diagnosed, it is recommended that other associated congenital cardiac anomalies be ruled out. The most common coexisting abnormalities with PLSVC are septal defects (ventricular or atrial).[3]

In most cases, PLSVC drains into the right atrium via the coronary sinus and therefore does not produce an intracardiac shunt. Its early diagnosis, however, is important especially in patients undergoing central catheterisation, pacemaker implantation or cardiac surgery.[3]

However, in 10–20% of patients with the condition, PLSVC may drain into the left atrium through a prominent unroofed coronary sinus, either directly into the left atrium or through the left superior pulmonary vein,[3] creating a right-to-left shunt predisposing to the development of thromboembolic episodes and cerebral abscesses,[3,4] as in the case presented here. Transient bacteraemia due to oropharyngeal flora can occur simply by brushing teeth or as a result of simple dental procedures. These microorganisms cannot survive in the oxygen-rich environment of the pulmonary circulation and are eliminated by alveolar macrophages or the lymphatic system.[3] However, a right-to-left shunt causes the loss of this safety mechanism, as venous blood travels to the left heart without passing through the pulmonary circulation, allowing these organisms to access the systemic circuit and potentially reach the brain.

Embolisms or arrhythmias are also frequent in these patients after intravenous punctures in the left upper limb,[2,4] following device implantation or catheterisation. Cases of coronary sinus perforation and even death have been reported, with a higher risk if the right superior vena cava is absent.[4,5]

In addition, when PLSVC drains into the left atrium it requires surgical treatment due to the shunt it causes. Recently, minimally invasive intravascular options with vascular occluders or coils have been described.[1,2]

In conclusion, therefore, it can be said that PLSVC is the most common anomaly of the thoracic venous system, often associated with other congenital defects. Generally, PLSVC drains into the right atrium through the coronary sinus, but in some patients it may drain directly into the left atrium, creating a shunt that predisposes to the development of cerebral abscesses or thromboembolic events.

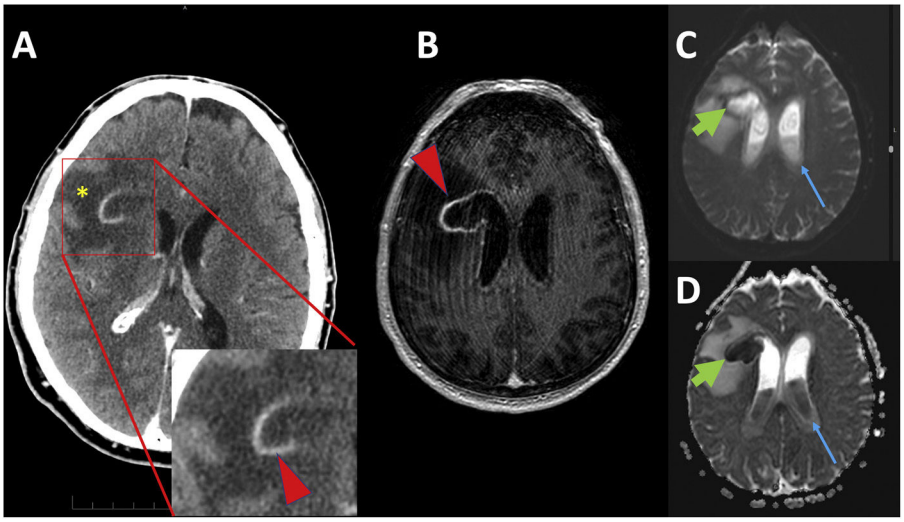

Figure 1 (A) Cranial CT scan: right frontal space-occupying lesion with ring enhancement (arrowhead), in close relation to the frontal horn of the right lateral ventricle, with moderate expansion effect and associated vasogenic oedema (asterisk). (B) 3D T1-weighted, DWI (C) and ADC (D) MRI sequences. The abscess shows markedly restricted diffusion in the diffusion sequence (thick arrows), with signs of ventriculitis (thin arrows).

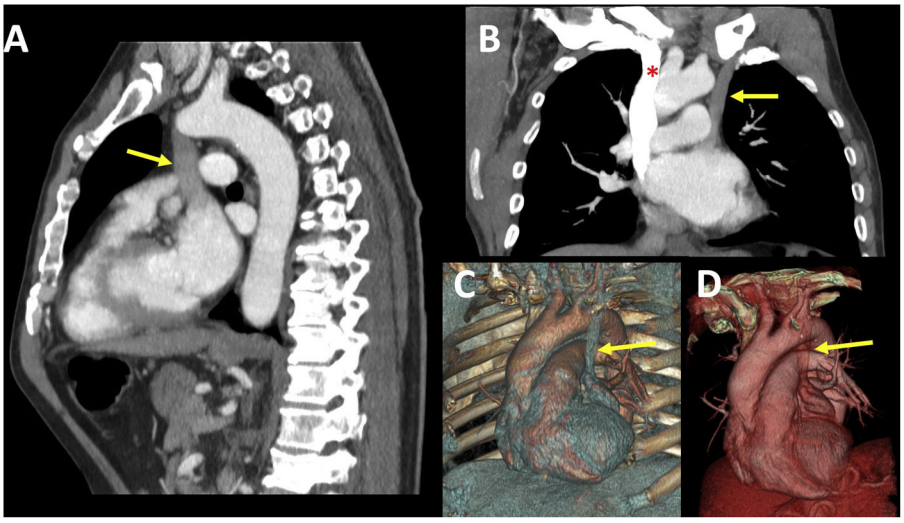

Figure 2 Sagittal (A), coronal (B) and volumetric (C, D) CT reconstructions showing the persistent left superior vena cava (arrows), draining into the left atrium. The right superior vena cava is preserved (asterisk).

Author contributions

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- Development of study concept: María Elena Peña Gómez.
- Study design: Teresa Cobo Ruiz, María Elena Peña Gómez.
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- Statistical analysis: Not applicable.
- Literature search: Teresa Cobo Ruiz, María Elena Peña Gómez.
- Writing of article: Teresa Cobo Ruiz.
- Critical review of the manuscript with intellectually relevant contributions: Teresa Cobo Ruiz, María Elena Peña Gómez.
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Conflicts of interest

The authors declare that they have no conflicts of interest.

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Congenital pyriform apertura stenosis

Estenosis congénita de la apertura piriforme

Upper airway obstruction is a frequent reason neonates are taken to the accident and emergency (A&E) department. When accompanied by certain warning signs, such as respiratory failure or cyanosis, we must investigate whether the cause could be a treatable congenital anomaly.

We present the case of a 15-day-old male neonate who was taken to A&E because of breathing difficulties linked to common cold symptoms. Relevant medical history included two previous similar episodes since birth, which had been less severe and self-limiting. Upon physical examination, the patient was observed to be in poor general condition, with tachypnoea, subcostal retraction and oxygen saturation below 90%. It was also impossible to insert a nasogastric tube through the nasal cavities. Faced with this situation, orotracheal intubation was performed and computed tomography (CT) imaging of the paranasal sinuses was requested.

The CT ruled out choanal pathology, the primary clinical suspicion, but abnormal thickening and medial approximation of both nasal processes of the maxilla was detected (Fig. 1A), resulting in pyriform aperture stenosis, with a maximum diameter of 5.8 (Fig. 1B). An associated abnormality, a solitary median maxillary central incisor (also known as a central megaincisor), was also detected (Figs. 2A and B).

To complete the examination, a cranial magnetic resonance image (MRI) was performed to rule out other associated malformations; this came back normal. On the basis of the various tests conducted, the patient was treated conservatively, with a good response. No surgical treatment was needed. The patient is currently being monitored by the ENT department and is progressing favourably.

Congenital pyriform aperture stenosis is an infrequent but treatable obstruction of the upper respiratory tract in neonates. Incidence is estimated at 1/25,000 live births.[1] Clinically it manifests with episodes of respiratory distress, difficulty feeding, apnoea crises and cyanosis.[2] Choanal atresia, the symptoms of which are very similar, is the primary differential diagnosis.[3]

Diagnosis is made on the basis of clinical suspicion and the results of imaging tests; CT scan of the paranasal sinuses without intravenous contrast is the method of choice. The characteristic finding is an abnormal medial approximation of both nasal processes of the maxilla, which are usually thickened, resulting in a decrease in the transverse diameter between the two spines, whose normal diameter has been established by consensus as greater than 11 mm.[4]

It may appear in isolation or, more frequently, associated with other malformations,[1] hence the importance of early diagnosis and awareness of this entity. Among the anomalies frequently associated with this pathology are the different forms of holoprosencephaly or the presence

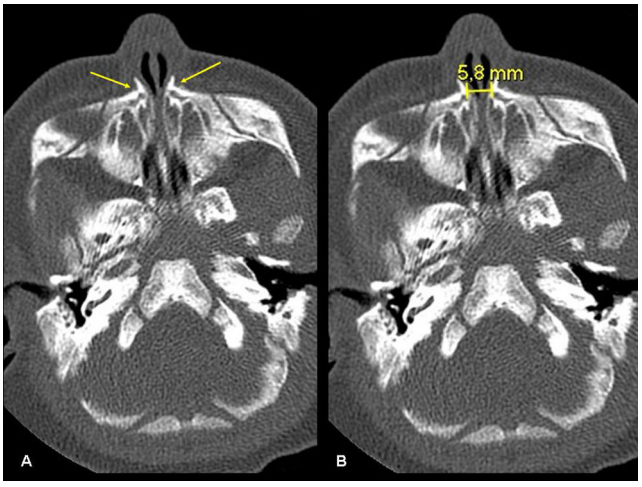

Figure 1 Axial CT section of the paranasal sinuses without intravenous contrast. Abnormal medial approximation and thickening of the nasal processes of the maxilla are observed (A, arrows), representing an abnormally narrow pyriform aperture of 5.8 mm (B).