Prenatal diagnosis of Persistent Left Superior Vena Cava draining into a cervical hemangioma: Case report and literature report

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Abstract

Introduction Vascular anomalies are alterations in angiogenesis and endothelial proliferation, respectively, that can generate not only aesthetic problems, but also functional disorders in fetuses and newborns. Other alterations in the regression of blood vessels can generate vascular malformations such as persistent left superior vena cava. Case report We present the case of a

INTRODUCTION:

Vascular anomalies can be divided into two large groups of alterations and the International Society for the Study of Vascular Abnormalities (ISSVA) differentiates them according to their clinical course and anatomical, histological and pathophysiological characteristics (1-3). On one hand, vascular malformations, constitute a group of blood vessel abnormalities generated by disorganized angiogenesis secondary to mutations in the vascular endothelial growth factor (VEGF) receptor, are present at birth and progress throughout life (arteriovenous fistulas, spot port wine, nevus flammeus, among others) (3,4). On the other hand, vascular tumors are generated by the abnormal proliferation of the endothelium with aberrant architecture of the blood vessels, generating masses that not only generate aesthetic problems but can also generate functional disorders in fetuses and newborns (2,4).

Some alterations at the vascular level may be the result of the lack of regression of some primitive vessels of the vascular system, as is the case of the persistent left superior vena cava (PLSVC), which constitutes the most common thoracic venous variation and is generated by the persistence of the left anterior cardinal vena cava which by the eighth week of embryogenesis should become the left anterior cardinal ligament (5–7). In isolated cases this alteration is considered a benign condition with a low probability of aneuploidy, associated abnormalities and adverse perinatal outcome, however some studies have identified that its association with other alterations can increase these events (5).

CASE REPORT:

A 22-year-old primiparous woman with a 26.5-week pregnancy was referred to the maternal-fetal medicine department of the Colombia University Clinic for obstetric ultrasound due to the finding of a mass in the cervical region in anatomical detailed ultrasound. The patient had no significant history and normal prenatal control paraclinical findings. An ultrasound was performed in which a predominantly cystic mass was found with multiple septa inside, without color Doppler uptake, with dimensions of 32 x 26 mm in largest diameter,

which goes from the lower cervical and left lateral area to the suprascapular region, suggesting as the first diagnostic possibility of lymphangioma, which is why it was decided to refer to the fetal anomalies board of the department for ultrasound evaluation.

She was evaluated at 28.2 weeks by the fetal anomalies board in which a fetus was found with a septated cervical and left lateral cystic lesion with growth towards the thoracic region with significant uptake on color Doppler, in addition to this finding an enlarged persistent left superior vena cava with effect from a shunt to a lesion described in the cervical region, thus defining that it was a hemangio-lymphangioma, with no secondary obstructive effect (FIGURE 1). The rest of the ultrasound evaluation was normal. It was then decided to call the patient for a follow-up in 3 weeks to assess the behavior of the lesion.

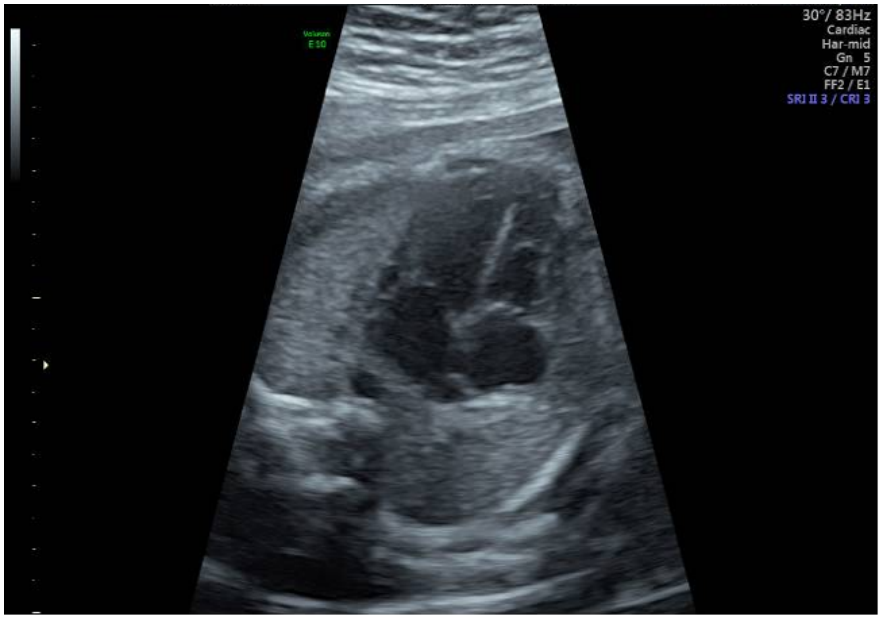

Figure 1. Ultrasound photos performed at the joint of anomalies. Top Left: Image of 4 chambers view with the presence of dilatation of the venous sinus characteristic of PLSVC. Top right: Right ventricular outflow tract with presence of adjoining vessel in the left hemithorax. Bottom left: Abnormal 3VT section with presence of a fourth vessel on the left with dilation and Doppler flow. Bottom right: Connection of the PLSVC to the cervical hemangioma.

She was evaluated again in a meeting at 31.2 weeks. In this evaluation, there were no changes regarding to the size of the lesion, nor a secondary obstructive effect, for which it was decided a follow up with a biophysical profile in 3 weeks and a new assessment by the fetal anomalies board in 5 weeks to define the route and time for delivery.

At 35.3 weeks, the patient attended to the emergency department of the Colombia University Clinic, referred from an outpatient control ultrasound for finding a hydropic fetus without fetal heart rate. Upon admission, fetal death was verified and by the patient's desire it was decided to perform a cesarean section for the end of the pregnancy. A female dead newborn was obtained during the procedure, with effacelation on the thorax, abdomen, upper and lower extremities, overlapping sutures, and a 6 x 4 cm left cervical mass (FIGURE 2). The weight of the gestation product was 1,970 grams, with a height of 42 cm and an APGAR score of 0/0/0. The patient was discharged 24 hours after the procedure due to adequate clinical evolution.

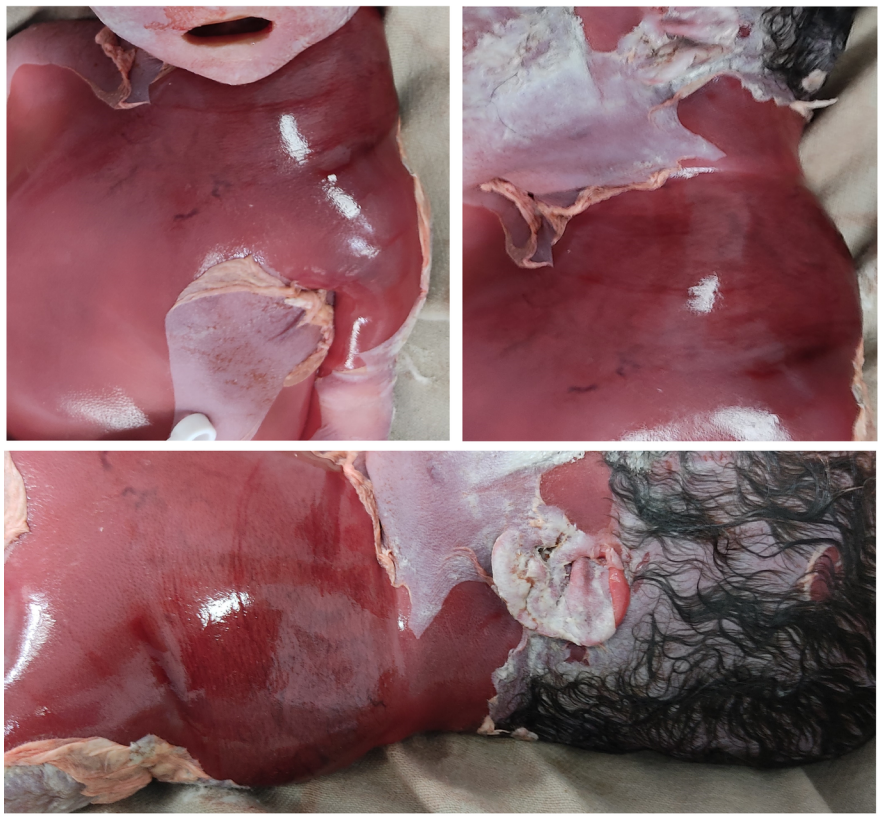

Figure 2: Postnatal record. A left cervical mass of approximately 6x4 cm is observed. Photos taken with permission of the patient.

MATERIALS AND METHODS:

A literature search was performed in the PUBMED database with the terms MESH ("Hemangioma"[Mesh]) AND (Persistent Left Superior Vena Cava[Mesh]), without finding any results available. A new search was then performed with the terms "Persistent Left Superior Vena Cava" AND "hemangioma" finding 6 articles, of which only 1 described a similar pathology, but in an adult patient. To develop the literature report, a new search was performed with the terms ("Hemangioma"[Mesh]) AND ("Neck"[Mesh]) and the search strategy (Persistent Left Superior Vena Cava[Mesh]), applied filters from the years 2018 to 2022 finding a total of 104 articles, of which 88 were eliminated by title and 1 was eliminated by summary. In the end, a total of 14 articles were selected for review.

RESULTS:

Hemangiomas, formed by arteriovenous connections grouped within spaces of flow voids, are the most frequent benign tumors of childhood and within these, at the vascular level, the most common in the head and neck, generating alterations in children's development, both physiological as psychosocial (4,8). Its prevalence reaches up to 4-10% in the child population, without being able to have data regarding its prevalence in relation to its prenatal diagnosis (4,8) Anatomically, when these lesions are located in Superficial dermis are called superficial hemangiomas, manifesting as raised, red lesions; when they are located in the reticular dermis or in the subcutaneous cellular tissue, they appear as raised masses covered with normal or bluish-colored skin, known as deep hemangiomas. When the same superficial and mixed components coexist in the same lesion, hemangiomas are called mixed hemangiomas (4).

Hemangiomas can also be divided into two genetically distinct subtypes, infantile and congenital, and can

also present as an isolated event or in a syndromic association (4). Infantile hemangiomas are red-blue protruding lesions that can appear up to 4 weeks after birth and continue to develop 5 to 6 months after it, in rapid growth that can even lead to doubling in size, all this to later gradually involute from the very beginning from 6 to 12 months of life and up to 4 years (2,4). Its proliferation will also depend on the phase in which the lesion is located, which can be proliferative in which 80% of hemangiomas reach their maximum volume at 5 months (early proliferative phase) or up to 9 months (early proliferative phase). late proliferative), this is followed by the phase of partial regression and total regression (8). This type of hemangiomas can be divided into localized (single, focal) and segmental (plaque-shaped and multifocal) lesions (1,9).

On the other hand, congenital hemangiomas are present at birth and can be rapidly involuting, partially involuting, or non-involuting. The appearance of these lesions occurs in the twelfth week of gestation, which is why its prenatal diagnosis is possible if the ultrasound evaluation is carried out by trained personnel (1). Commonly, the skin manifestations of neonatal hemangiomas are related to liver manifestations, which is why it is always necessary to actively search for similar lesions at the liver in ultrasound assessments (4).

A retrospective study carried out at the University of Chile with 174 patients diagnosed with infantile hemangioma found that 61% of the lesions were located on the head and neck, 21% on the trunk, 10% on the upper limbs, 4% on the lower limbs and 3% in the anogenital region. In these patients, the presence of arteriovenous shunts was also identified in 20% of the lesions and direct afferent branches of the main regional arteries in 15%, which were associated with delayed involution of the lesions in some patients and hindered therapeutic response (8).

The use of ultrasound, especially in the prenatal diagnosis of this type of lesions, seeks to establish the presence of secondary obstructive compromise, especially of the airway that could configure a problem at the time of birth, establishing even the need to perform a cesarean section with the EXIT procedure. (1). Likewise, the evaluation of lesions through the application of Doppler flow helps to classify lesions into low-flow and high-flow lesions, thus predicting their behavior and the response to drug treatment or the appropriate moment for postnatal surgical management (4, 9).

The introduction of fetal echocardiography with its basic cuts and especially the cut of 3 vessels and trachea (3VT) in routine prenatal ultrasound has also made it possible to detect the presence of venous anomalies such as persistent left superior vena cava, this especially when it is related to other congenital heart diseases (CHD), aneuploidies, heterotaxy and extracardiac anomalies (6,10)

PLSVC has a prevalence in the general population of 0.3-0.5%, which increases to 4-8% in patients with congenital heart disease (5,10,11). It has its anatomical origin at the junction of the left jugular and left subclavian veins, running anterior to the aortic arch and the left pulmonary artery to the lateral border of the left atrium to finally drain into the coronary sinus in 90% of cases and in 10% in the left atrium. Dilation of the coronary sinus then becomes one of the main ultrasound signs of PLSVC in the 4-chamber view, as well as the presence of a fourth vessel to the left of the pulmonary trunk and the ductus arteriosus or the appearance of abnormally large vessels ordered (from right to left: aorta, pulmonary artery, PLSVC) in the 3VT section (5,6,12).

A prospective cohort study sought to assess the outcomes of patients with isolated prenatally diagnosed PLSVC compared with those associated with other abnormalities. A total of 256 fetuses were included in this, of which 113 fetuses entered the group with isolated PLSVC; none of these fetuses had adverse neonatal outcomes, however 10 cardiac abnormalities (8.8%) and five extracardiac abnormalities (4.4%) were subsequently diagnosed in the postnatal period. In the group of fetuses with extracardiac anomalies and cardiac anomalies of the 143 analyzed fetuses, 27 of them with septal defects, 23 with conotruncal anomalies, and 17 cases of obstructive disease of the left ventricular outflow tract were detected, setting heart disease as the most common abnormalities. In the study, 11 postnatal deaths were identified, 7 in the neonatal period and 4 after postnatal cardiac surgery; all deaths were attributed to complications from associated pathologies, since the presence of PLSVC could not be related as a cause of perinatal death (7,10)

In relation to aneuploidies, a retrospective study in which 95 cases of patients with PLSVC were analyzed and found a prevalence of 13.5% of aneuploidies in their population. Similarly, it was identified that isolated cases of PLSVC had better obstetric outcomes than non-isolated cases, a group in which the presence of cardiac and extracardiac anomalies worsened neonatal outcomes (11). One of the anomalies that has also been related to PLSVC is the VACTERL anomaly, which has been analyzed in several studies, finding an incidence of association ranging from 3.2% to 25% (13).

Only one of the articles reviewed reported a 57-year-old male who attended the emergency department for chest pain and in whom an irregular mass in the pericardial chamber was documented during echocardiography. During the surgical procedure, it was established that this mass was a cardiac hemangioma measuring 110x65x45 in the wall of the right atrium that seemed to involve part of the interatrial groove; additionally, the lesion presented a communication towards a 3 mm diameter IPVC between the coronary sinus and hemangioma (14).

DISCUSSION:

In the above clinical case, we present a patient initially referred to our unit due to the finding of a hemangioma-type vascular tumor located at the cervical level, which is consistent with what has been reviewed in the literature as the site of greatest clinical presentation of these lesions. On the other hand, we were also able to identify that the vascular component of the hemangioma in the clinical case came from a connection to a persistent left superior vena cava, which constitutes a vascular anomaly never previously described in any case report according to what was reviewed in the literature.

In the only previously reported case of a persistent hemangioma with a connection to the left superior vena cava, it presented as an intracardiac lesion in the right atrial wall that also seemed to involve part of the interatrial groove. This condition is also a very rare presentation since cardiac hemangiomas correspond to only 1-2% of cardiac tumors and there is no report of a similar communication (14).

The fetal death that occurred in the clinical case could be presumed to be associated with the presence of the extracardiac malformation associated with PLSVC, remembering that the presence of isolated PLSVC could not be related to an increase in adverse perinatal outcomes (5). This corresponds to the fact that the observed survival rate in infants with PLSVC alone was significantly higher than that of infants with PLSVC associated with cardiac and extracardiac abnormalities in a prospective cohort study (10).

CONCLUSIONS

Cervical hemangiomas with direct cardiac irrigation are rarely reported in the literature. Prenatal diagnosis becomes an important tool that helps establish the presence of fetal abnormalities that could potentially complicate pregnancy. The detailed evaluation of different structures and the sequential assessment of the fetal heart provide a tool to achieve adequate counseling for parents and to be prepared for the different complications derived from said anomalies. Ultrasound follow-up is essential for the identification of possible complications derived from cervical masses.

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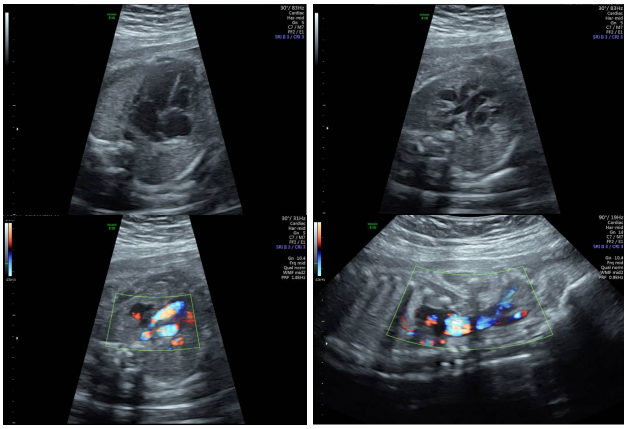

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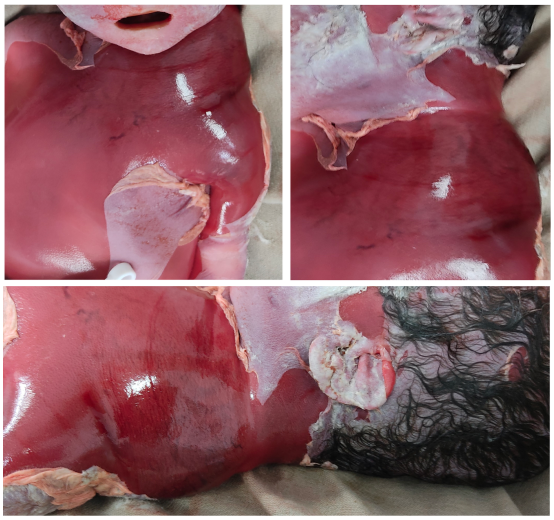

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