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Infantile hemangiomatosis, a rare cause of high-output heart failure and pulmonary hypertension in the newborn baby: a case report

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Brief Report

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Abstract

Infantile hemangiomatosis is among the most common vascular tumours of childhood that is generally accepted as benign. Some cases may have multiple hemangiomas with organ involvement, especially of the liver. This case report will present the clinical and laboratory findings obtained during the treatment and follow-up of a 36-day-old female baby with hemangiomatosis with diffuse liver involvement, high-output heart failure, and pulmonary hypertension.

Case

Our case was the 36-day-old female baby of a 30-year-old mother, born at 39 weeks and 3 days with gravidity 2 and parity 2. At birth, she was 2900 g, her height was 49 cm, and head circumference was 34 cm. The mother had controlled hypothyroidism and had got a COVID-19 infection during the 4th month of her pregnancy. The patient came to our outpatient clinic with jaundice and murmur. She had icteric skin, mild oedema, tachypnia, and subcostal and intercostal retraction. Her body temperature was 36.5°C, and respiratory rate was 72/min. Her heart rate was 165 bpm, blood pressure was 68/47 mmHg, and O_2 saturation was 93%. Her oscultation results showed 2/6 systolic murmur. She had a distended abdomen and a 4 cm hepatomegaly without splenomegaly. She had non-blanching, isolated hemangiomas on the right occipitotemporal region (approximately 2x2 cm), left subscapular region (approximately 1x1mc), left lateral thigh, left dorsal thumb, right anterior leg, and left shoulder, as well as many milimetric sized, red/purple hemangiomas (Fig. 1).

Laboratory tests showed that haemoglobin 10.0 g/dL, haematocrit 29.1%, leucocyte 7900/mm^3, thrombocyte 529,000/mm^3, thyroid stimulating hormone (TSH) 12,1 mcg/dL, and C-reactive protein negative. Other tests were normal. Cardiomegaly was detected in chest radiography (Fig. 1).

Echocardiography showed dilated right heart chambers, left ventricular diastolic diameter (LVDD) 22.77 mm (z-score + 2.34), right ventricular diastolic diameter (RVDD) 21.4 mm (z-score + 5.69), normal systolic functions (shortening fraction (SF): 35%, ejection fraction (EF): 66%), atrial septal defect (5.6 mm), mild mitral valve regurgitation, and significant tricuspid valve regurgitation. Pulmonary artery systolic pressure was measured as 84 mmHg through tricuspid valve regurgitation. Her cardiac output was 2L/min. Diffuse hypoechoic, spherical lesions were found in the liver in the subcostal echocardiography view (Fig. 2).

Abdominal ultrasound showed hepatomegaly (75 mm) and multiple hypoechoic lesions (largest 15x14 mm) in the parenchyma. No pathologies were seen in transfontanelle cranial ultrasonography.

The patient's abdominal magnetic resonance examination showed diffuse enlargement of the liver, especially the left lobe. There were multiple tumoral lesions, the largest being 21 mm in diameter, in her liver parenchyma, which were hypointense in T1A images, hyperintense in T2A images, and noncontrast enhancing in postcontrast images. Diffusion MRI showed restricted diffusion in the lesions. Hepatic veins were dilated (Fig. 2).

The patient was given treatment for heart failure (captopril, furosemide, and spironolactone), feeding and oxygen support, and her fluid intake was restricted. Oral propranolol (3x1 mg/kg/dose) treatment was administered for diffuse infantile hemangiomatosis.

The patient's heart failure findings, respiratory problems, and large skin lesions regressed during follow-up. Smaller skin lesions became darker in colour and their diameters decreased.

The abdominal ultrasound done one month after the treatment showed a decrease in the patient's liver size (61 mm) and the hypoechoic lesions (largest 11x12 mm). The two following check-ups, done every 3 months, showed a further decrease in liver size (54 mm) and the lesions'

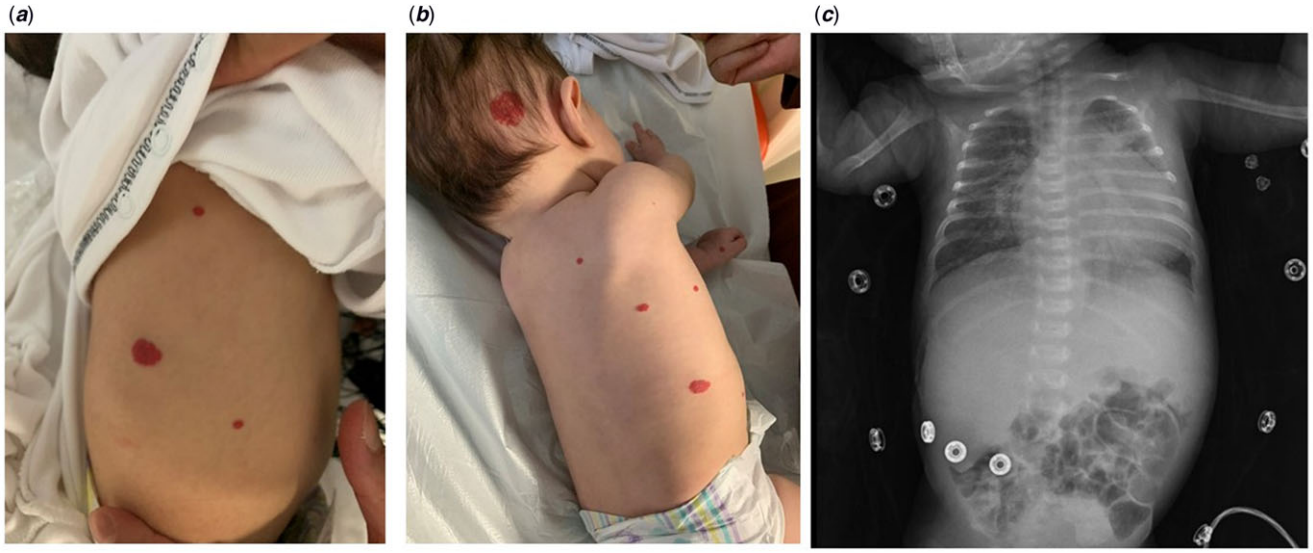

Figure 1. The patient's cutaneous hemangiomas before treatment (A), in the 6th month of treatment (B), and chest and abdomen X-ray (C).

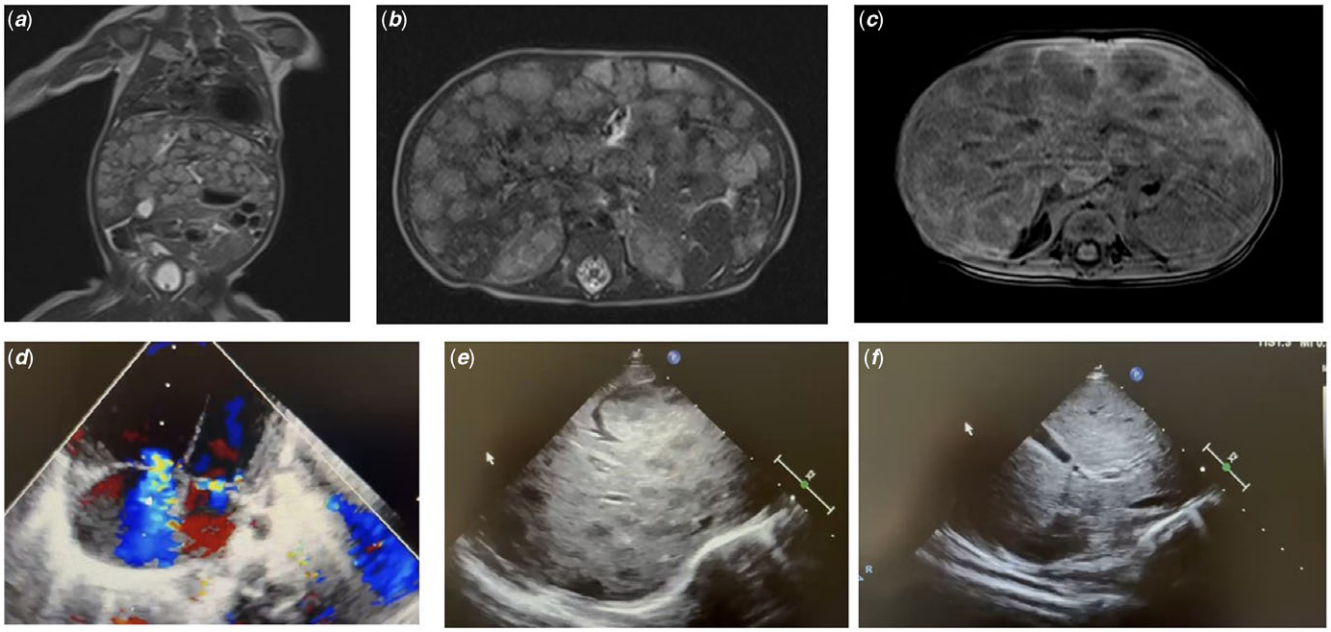

Figure 2. MRI images in the coronal plane (A), T2A image (B), and T1A image (C) in the transverse plane. Echocardipgraphy images of valve regurgitations in 2D view (D), subcostal view of liver hemangiomas before (E) and after (F) treatment.

size (largest 6 mm). Her cardiac output decreased to 0.9 L/min. Her valve regurgitations and dilation in inferior vena cava (IVC) and hepatic veins were also reduced. Her cardiac findings ameliorated during the second month of the follow-up. Her tricuspid valve gradient decreased to 23 mmHg. Subsequently, captopril treatment was ceased. Oral propranolol treatment is still being continued.

Discussion

Infantile hemangiomas are the most prevalent benign vascular tumours of childhood. Its incidence in children can go up to 5%.[1–3] It is seen more frequently in Caucasian people and females.[4,5] Low birth weight, premature birth, multiple gestations, increased maternal gestation age, in vitro fertilisation, preeclampsia, and placental anomalies are among the risk factors for infantile hemangiomas.[4,5] However, their pathogenesis is not fully elucidated. The currently accepted hypothesis states that the endothelial progenitor cells in the circulation migrate to regions with conditions that favour their growth such as hypoxia or developmental disturbances.[1]

The lesions' progression which starts at 1–3 months is usually completed by the fifth month.[4] Later on, most lesions involute. Approximately 40% of the lesions disappear until the age of 5, 75% until 7, and 90% until 9.[6]

Cutaneous hemangiomas manifest as multiple lesions in 10–20% of cases.[6] Multiple cutaneous hemangiomatosis usually

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presents with the risk of organ involvement, which can be fatal.[7] The involvement of the liver, lungs, brain, eyes, and intestines is the most prevalent.[6]

The fact that our patient was diagnosed with infantile hemangiomatosis while being examined for cardiac murmur makes this case intriguing. She was diagnosed based on cardiac findings and the presence of skin lesions with small diameters. When there are more than 5 cutaneous hemangiomas, the risk of hepatic hemangiomatosis significantly increases.[7] Although our patient's skin lesions were relatively small, they were numerous, raising the question of hepatic hemangiomatosis. The fact that multiple and diffuse hepatic hemangiomatosis frequently presents with high-output cardiac failure, hepatomegaly, valve regurgitations, coagulopathy, pulmonary hypertension, and hypothyroidism[3,8] – some of which were significant findings of our case – made us lean towards this potential diagnosis. Our hypothesis was confirmed by abdominal ultrasound, the principal method used for the diagnosis of infantile hemangiomatosis. However, if the lesions appear malignant in the ultrasound, other imaging techniques such as MRI may be required.[8,9] Since the hepatic lesions of our patient were large and diffuse, we also did an MRI scan to rule out any potential malignancies.

After being found to be effective against hemangiomatosis in 2008, propranolol has been accepted as the first-line treatment.[2] A meta-analysis has shown that propranolol treatment was more efficient in infantile hemangiomatosis cases compared to steroids, vincristine, and laser treatment.[10] Hence, we also administered oral propranolol treatment to our case. We did not observe any side effects and did not require additional treatment. After one year of treatment, our patient's hepatic lesions significantly regressed.

In conclusion, infantile hemangiomatosis cases can consult paediatric cardiology clinics, as in our case, with hypovolemic heart failure and pulmonary hypertension due to hepatic involvement; hence, it is crucial for cardiologists to be aware of this pathology in order to make accurate diagnoses and treatments. The significant improvement of the severe clinical picture of our case also underlines the efficacy of the first-line drug propranolol in the treatment of infantile hemangiomatosis with hepatic involvement.

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Competing interest. None.

Ethical standards. Written informed consent was obtained from the patient's guardian.

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