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CASE REPORT

IDIOPATIC INFANTILE ARTERIAL CALCIFICATION: ECHOCARDIOGRAPHY DIAGNOSIS AND SUCCESSFUL TREATMENT

Hasan ÖNAL[1] Neval TOPAL[1] Melek YILDIZ[1] Banu AYDIN[1] Aygün DİNDAR[2]

1 Sağlık Bilimler Üniversitesi, İstanbul Kanuni Sultan Süleyman Sağlık Araştırma ve Uygulama Merkezi, Çocuk Endokrinoloji ve Metabolizma Kliniği
2 İstanbul Üniversitesi, İstanbul Tıp Fakültesi, Çocuk Kardiyoloji

ABSTRACT

Idiopathic infantile arterial calcification is rare disease characterized by medial calcification and intimal proliferation of large and medium size arteries (1). Prenatal prezantation is usually with nonimmune hidrops and polyhydamnios (2). Although the clinical characteristics in neonatal period is variable, respiratory distress with cardiac failure is the most common finding. Hypertension due to poor compliance caused by diffuse arterial calcification of the vessels is present in most patients. Clinical experience in treating this rare disorder is limited. We present a neonatal case with Idiopathic infantile arterial calcification who was treated with etidronate disodium, successfully.

Keywords: Calcification, arterial, etidronate disodium

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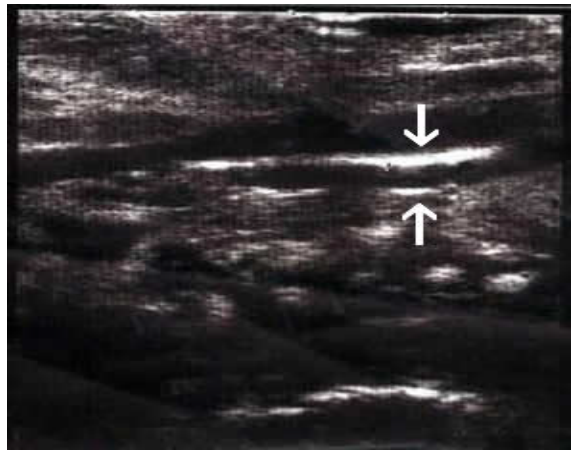
CASE REPORT

A female infant, one of the dizygotic twins was born to a 33 years old woman at 36. weeks of gestational age via caesarean delivery. Her birth weight (2600 gr), height (44 cm) parameters were in the normal range. Her twin brother was healty. The patients was presented with respiratory distress on the first day of life. On physical examination, her respiratory rate was 65 per minute, heart rate was 136 beats per minute and blood pressure was 115 mmhg systolic and 80 mmhg diastolic. Peripheric pulses were palpable and a grade 1/6 systolic murmur was present on the left sternal border.

Echocardiograhy revealed concentric hypertrophy of the left ventricle and interventricular septum, good cardiac contractility and bright echogenic walls of the coronary arteries (Figure 1), aorta and pulmonary artery due to calcification. Cranial ultrasonography was found to be normal. Computerized tomography of the thorax and abdomen showed similar findings including renal arteries. Her biochemical parameters were in the normal range. She was put on prazosin 1 mg/kg/d and furosemid 1 mg/kg/d and her blood pressure was normalised. On follow-up, the urine Ca/creatinin ratio increased. Therefore her anihypertensive treatment was replased with propranolol 1 mg/kg/d. She was given etidronate disodium 5 mg/kg/dto prevent further calcium deposition in arterial vessels.

She was kept on the medical ward with nazogastric feedings. The serum calcium concentration increased to 11.5 mg/dl during etidronate treatment and she developed rickets with high alcaline levels of 1650 U/lt and radiologic findings. She was given high dose of vitamine D. Repeat echocardiograms showed regression of arterial of arterial calcification. She was discharged at 5 months old age. Etidronate disodium was discontinued at 10

Figure 1. Echocardiogram at 18 day of age showing marked hyperechogenicity of the coronary arteries



Correspondence Address: Abdurrahman Nafiz Gürman mahalesi, Kınalıtepe sok, simitaş 7.blok, No:61, Güngören / İstanbul

E-mail:hasanonal@hotmail.com

months of live owing to side effects.

At present she is doing well with normal blood pressure, only an propranolol 1 mg/kg/d. Her weight (8.5 kg) and height (70 cm) parameters are below the third percentile but her head circumference (47 cm) is in the normal range (25p.)

DISCUSSION

Idiopathic arterial calcification in infancy was first reported in 1901 by Bryant and White (3). Since then many case reports and reviews of the disease have been published. Nearly a 100 patients have been reported in the literature so far. Most cases were diagnosed at postmortem (4).

Recently ultrasonography has been a more sensitive investigation to detect the vascular involvement pre and postnatally (5). The diagnosis was made by echocardiography on our patient, that was performed to detect the underlying a etiology of hypertension. Hypertension may be refractory to conventional therapy. Ciana G et al (6) reported a premature baby with severe hypertension associated with idiopathic arterial calcification. They were able to control hypertension with prostaglandin E1 infusion and carried on with a multiple antihypertensive regimen after stabilization.

We could control hypertension in our patient with prazosin and furosemid initially and propranolol later on.

The pathophysiology of this disorder is well documented but the underlying aetiology remains unknown. An autosomal recessive metabolic disorder in the levels of the enzymes responsible for inorganic phosphate balance resulting in abnormal deposition of calcium into the vessels is thought likely (7).

Our patient who has got a healthy twin brother was supporting the autosomal inheritance pattern of the disease. Fetal echocardiographic evaluation of the future pregnancies is recommended for all affected families.

Spontaneous regressions of calcification and clinical improvement have been reported in patients treated with corticosteroids, either alone or in combination with other medications including estrogen and thyroid extract (8).

Meradji et al (9) reported successful therapy with orally administered diphosphonate in two patients, both of whom are doing well into adolescence. Stuart et al (10) have reported two siblings with fetal diagnosis of aetiology who were unresponsive to the treatment with disodium etidronate and both died.

Etidronate disodium is a member of the diphosphanate class of drugs. Its major action is to reduce both normal and abnormal resorption of bone. It can lead to resolution of the arterial calcification, pathologic fractures and rickets (9).

Conclusion:

We gave disodium etidronate during the first 5 months of live to our patient and provided a satisfactory. Outcome with resolution of calcification on repeat echocardiographic examinations. Despite severe side effects we may suggest this agent for the treatment of this rare disorder.

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