Idiopathic Arterial Calcification of Infancy

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Case 1

Pre-term newborn infant delivered by Cesarean section who presented with non-immune hydrops fetalis, CHF, and ascites.

Our purpose is to demonstrate the characteristic imaging findings of idiopathic arterial calcification of infancy.

Discussion

Clinical Presentation: Idiopathic arterial calcification of infancy is a rare cause of arterial calcification characterized by extensive calcification in systemic and pulmonary arteries. Infants usually present in the neonatal period or later in infancy. Symptoms include lethargy, poor feeding, and respiratory distress with a rapid decline in function ultimately leading to refractory hypertension and cardiac failure. Treatment is controversial, but diphosphonates and steroids may be considered. There have been reports of decreased calcification and improved vascular integrity with treatment. In some cases, radiographic evidence of arterial calcification has completely disappeared with years of therapy.

Pathology: The mechanism for calcification is unknown, but altered iron metabolism, degeneration of elastin fibers, altered prostaglandin metabolism, abnormal response to vascular injury, and disorders of calcium and phosphorus metabolism have been postulated. This disorder is thought to be inherited in an autosomal recessive pattern as familial involvement has been reported. Idiopathic arterial calcification is a diagnosis of exclusion; hyperparathyroidism, hypervitaminosis D during the pregnancy, atherosclerosis secondary to intrauterine infection, and other disorders of calcium-phosphorus metabolism must first be excluded.

The disease is characterized by fibroproliferation of the internal elastic lamina with deposition of calcium hydroxyapatite. This leads to decreased compliance, hypertension, and narrowing of the vessel lumens with resultant ischemia. The prognosis is poor with most affected infants dying prematurely or within the first six months of life.

REFERENCES


Case 2

Pre-term newborn delivered by Cesarean Section with cardiomegaly and necrosis of toes.

Idiopathic arterial calcification of infancy is a rare disorder with clearly demonstrable radiographic findings. Calcification of vessels, joints, and organs are easily seen by plain film, ultrasound, and computed tomography. The diagnosis can be made with a high index of suspicion after other etiologies for abnormal calcification have been excluded.

Conclusion

Idiopathic arterial calcification of infancy is a rare disorder with clearly demonstrable radiographic findings. Calcification of vessels, joints, and organs are easily seen by plain film, ultrasound, and computed tomography. The diagnosis can be made with a high index of suspicion after other etiologies for abnormal calcification have been excluded.

REFERENCES