



Idiopathic Arterial Calcification of Infancy

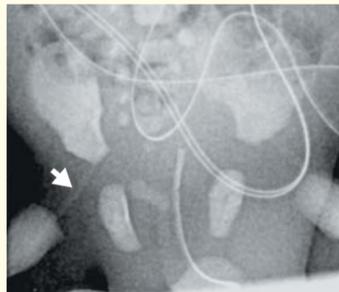
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Case 1

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



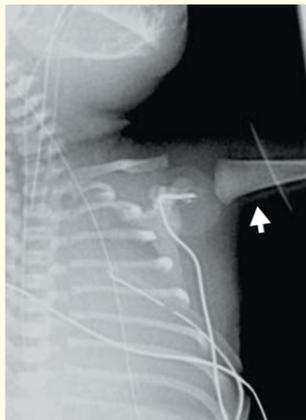
AP view of the abdomen demonstrates linear calcification of the right femoral artery.



AP view of the abdomen demonstrates linear calcification in the abdominal aorta.



Longitudinal ultrasound image of the aorta demonstrating linear hyperechogenicity in the walls of the aorta and superior mesenteric artery.



AP view of the chest demonstrates linear calcification in the axillary and brachial arteries.

Case 2

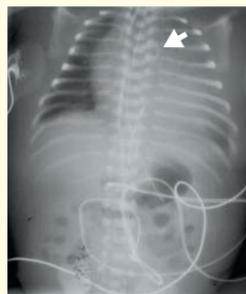
Pre-term newborn delivered by Cesarean Section with cardiomegaly and necrosis of toes.



Longitudinal ultrasound image of the right kidney demonstrates hyperechoic foci in the renal parenchyma consistent with calcification.



Longitudinal ultrasound image demonstrating linear hyperechoic aortic walls.



AP view of the chest and abdomen demonstrates curvilinear calcification of the aortic arch.

Purpose

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 can be administered. 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, arteritis 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 lumen with resultant ischemia. The prognosis is poor with most affected infants dying prenatally or within the first six months of life.

Discussion

Death is usually due to heart failure secondary to hypertension or myocardial infarction.

Imaging: Conventional radiographic findings include calcification of the pulmonary arteries, the coronary arteries, and the aorta with its branches. Intra-articular and periarticular calcifications are also seen. Calcification may also be seen within the parenchyma of solid organs, including abdominal viscera and thyroid. Cardiomegaly is frequently a late finding. Less common findings include hepatomegaly and infarctions of the bowel, liver, and spleen. US is also useful, especially for prenatal evaluation. Prenatal US may demonstrate fetal hydrops, polyhydramnios, and vascular calcification. Postnatal US demonstrates hyperechoic arteries with marked acoustic shadowing. CT is also sensitive for the detection of calcification in vessels. Additionally, CT may reveal unsuspected areas of involvement such as calcification of solid organs or infarctions.

Conclusion

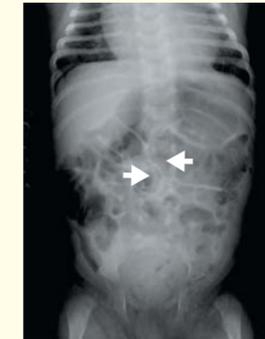
Idiopathic arterial calcification of infancy is a rare disorder with clearly demonstrable radiographic findings. Calcification of vessels, joints, and organs are easily seen by plain film, ultrasound, and computed tomography. The diagnosis can be made with a high index of suspicion after other etiologies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

REFERENCES

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3. Pao D, DeAngelis G, Lovell M, and Hagspiel K. Idiopathic arterial calcification of infancy: sonographic and magnetic resonance findings with pathologic correlation. *Pediatric Radiology* (1998) 28: 256-259.
4. Patel M, Andronikou S, Solomon R, Sinclair P, and McCulloch M. Idiopathic arterial calcification in childhood. *Pediatric Radiology* (2004) 34: 652-655.
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Case 3

Patient presented at two weeks of age with palpable wrist mass.



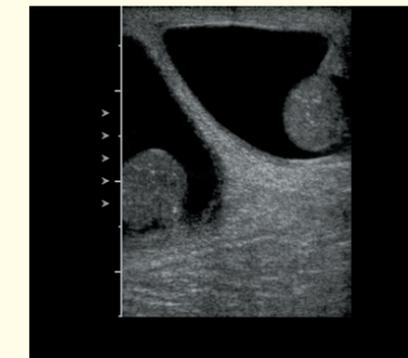
AP view of the abdomen demonstrates calcification of the abdominal aorta.



AP view of the wrist demonstrates calcification in and around the joint.



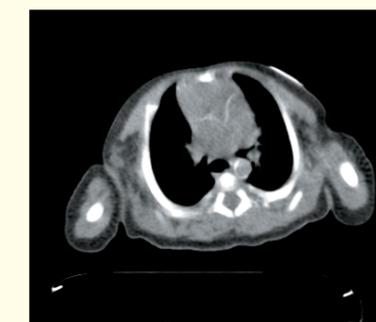
AP view of the humerus demonstrates both calcification of the brachial artery and calcification in and around the elbow joint.



Transverse ultrasound image of the scrotum reveals bilateral hydroceles and punctate hyperechoic densities in the testes.



AP view of the tibia and fibula demonstrates soft tissue calcification in and around the knee and ankle joints.



Axial CT image of the chest demonstrates calcification of the coronary arteries and thoracic aorta.



Axial CT image at the level of the renal arteries demonstrates calcification of the aorta, superior mesenteric artery, and bilateral renal arteries.