ISSN: 2694-1724
DOI: 10.33552/ARAR.2023.02.000549
Archives of Rheumatology & Arthritis Research
Iris Publishers

Clinical Case

Fibromuscular Dysplasia Vs Kawasaki's Disease

Cespedes Cruz Adriana Ivonne[1]*, Torres Jiménez Alfonso Ragnar[1], Velázquez Cruz Alejandra[1], Zeferino Cruz Maritza[1], Palafox Vargas Martha Leticia[2] and Serrato Auld Roberto Carlos[3]

[1]*High Specialty Medical Unit General Hospital La Raza. Department Paediatric Rheumatology, México*

[2]*High Specialty Medical Unit Specialty Hospital "La Raza". Department Pathology, Mexico*

[3]*High Specialty Medical Unit Pathology Hospital "La Raza". Department Angiology, Mexico*

***Corresponding author:** Cespedes Cruz Adriana Ivonne, Between jacarandas and vallejo Mexico City, Mexico.

Received Date: May 26, 2023

Published Date: June 21, 2023

Abstract

Fibromuscular Dysplasia (FMD) is a non-inflammatory vasculopathy that can present diverse vascular disturbances with the multifocal variant, characterized by arterial segments of varying length and alternating diameters (string of beads), affecting mainly renal, carotid and vertebral arteries and Kawasaki´s disease (KD) is a necrotizing vasculitis occurring in median and small vessels with well-established diagnostic criteria.

Keywords: Dysplasia; Kawasaki

Introduction

Fibromuscular Dysplasia (FMD) is a non-inflammatory vasculopathy that can present diverse vascular disturbances with the multifocal variant, characterized by arterial segments of varying length and alternating diameters (string of beads), affecting mainly renal, carotid and vertebral arteries [1]. It is more frequent in young women and the etiology is unknown. In adults is present in 4% of the population. Systemic arterial hypertension is present in most cases being headache the most frequent symptom. Radiologically is characterized by dilatations and stenoses in different arteries. The diagnosis is a histologic one, being fibroplasia of the middle layer the most frequent finding [2].

Kawasaki´s disease (KD) is a necrotizing vasculitis occurring in median and small vessels. It´s criteria for diagnosis are: fever >5 days, bilateral conjunctivitis (89%), changes in the oropharynx mucosa (96%), palmar or plantar erythema (75%), polymorphous erythema (96%), and cervical lymphadenopathy >1.5 cm (62%). It is the most frequent cause of aneurysms in children, having a more severe course in individuals under 1 year of age [3].

Case Presentation

This 4-month-old female infant, a previously healthy patient starts her disease with spiking fever of 104°F for 3 weeks, non-exudative conjuctival hyperemia on day 5 and ankle edema and soft stools (4/24 hours) for 3 days. Laboratories showed leukocytosis 40,920, platelets 575,000, RCP 24mg/dl. A cardiology evaluation on week 3 revealed a percardial effusion. She was managed with diuretics and antibiotics with abatement of the fever. On week 5, a 1.5cm pulsatile nodule was palpated in left axilla and arterial hypertension is recognized. An angiotomography was performed showing an image compatible with a fusiform aneurysm of the left axillary artery of 2cm in diameter and 3cm in length with a

filling defect in its interior. Similar findings were seen in the right brachial artery as well as coronary and renal arteries (Figures 1, 2). A resection of an aneurysm of the left axillary artery was performed with interposition of a reversed contralateral greater saphenous vein. A biopsy reported hyperplasia of the middle layer secondary to collagen and fibroblasts accumulation without inflammatory infiltration (Figure 3).

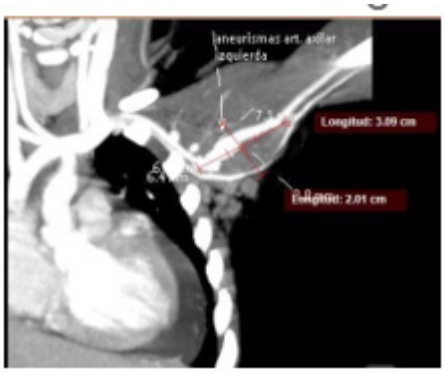

Figure 1: Angio tomography: aneurysm of the left axillary artery and bilateral renal.

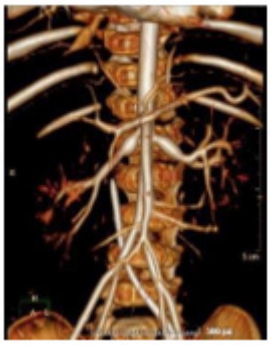

Figure 2: Angio tomography: aneurysm of the left axillary artery and bilateral renal.

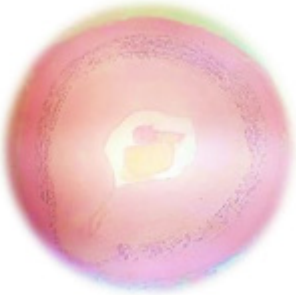

Figure 3: Hyperplasia of the middle layer resulting from fibroblast and collagen fibers accumulation. Stain: elastic fibers.

Currently, the patient is clinically asymptomatic, with a normal psychomotor development. She is on platelet anti-aggregates.

Discussion

This 4-month-old infant who initially presented a picture of a gastrointestinal infectious process with systemic manifestations (fever, conjunctivitis, pericardial effusion, elevated RPC and increased platelets) was thought of having Kawasaki's disease, although it did not meet diagnostic criteria. However, 5 weeks after onset of fever, generalized giant aneurysms appeared, which along with histologic findings made the diagnosis of fibromuscular dysplasia. The latter is a rare disease predominantly of the adult age. Radiologically there were multiple dilatations and stenoses without the characteristic image of "string of beads", yet the

histologic findings in the resected aneurysm are characteristic of this entity.

Conclusion

The presence of multiple aneurysms in the pediatric age should arise the suspicion of fibromuscular dysplasia once the diagnoses of Kawasaki´s disease and polyarteritis nodosa have been ruled out.

Acknowledgement

The collaboration of the doctors of Angiology, Pathology and Pediatric Rheumatology for this work to be carried out.

Conflict of Interest

No Conflict of Interest.

References

1. Heredia Torres N (2020) Dysplasia fibromuscular: una causa rara de hypertensión arterial. An Pediatr (Barc).
2. Adriana Bastos Conforto (2011) Multicenter studies shed light on fibromuscular dysplasia and cervical artery dissection. Arq. Neuro Psiquiatr.
3. Saguil A, Fargo M, Grogan S (2015) Diagnosis and management of kawasaki disease. Am Fam Physician 91(6): 365-371.