G10 Fibromuscular Dysplasia of Pulmonary Arteries: Report of Two Cases

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After attending this presentation, attendees will learn of a case showing complications of fibromuscular dysplasia (FMD) (aneurismatic dissection, arterial obstruction) most likely triggered by chest trauma and by possible cardiotoxicity due to association local/general anesthesia.

FMD is a non-atherosclerotic and non-inflammatory vascular disease, with a familiarity of 10% (inheritance dominante autosomica), and is characterized by fibrous or muscular or both types proliferation subverting normal architecture of the arterial wall. Etiology of FMD is unknown although various hormonal and mechanical factors have been suggested. Fibromuscular dysplasia generally affects women (94%) in their fertile age; it is frequently associated with pregnancy or hyperestrinism, but FMD can occur in any age, infancy included.

Clinical manifestations of FMD depend on involved arterial segment, histological type, and complications (obstructions, aneurysm rupture; embolism; sudden death). FMD commonly affects renal and carotid arteries, and less frequently it's observed in other small and medium arteries; pulmonary localization is rare. Prevalence of symptomatic renal FMD is about 4/1000 cases, twice as to that observed in carotid arteries. Histologically, FMD has been classified into three distinct types: intimal fibroplasia; fibromuscular medial dysplasia (medial hyperplasia, perimedial fibroplasia), and periarterial (adventitial) fibroplasia. Angiographic classification includes multifocale type, related with histological variant "medial fibromuscular dysplasia"; tubular and focal types, both no related with specific histological type.

In this study, two cases of unknown FDM involving pulmonary arteries are described. Clinical manifestation occurred in one case following a road accident related trauma and, in another case, following an anaesthetic induction and local anesthesia before surgical procedure.

Case 1: A 52-year-old obese man while driving a car got into an accident and suffered severe multiple trauma. He was taken to the Emergency Room where he presented coherent and breathing (SpO2 92%), with SBP/DBP 150/90 and CF 92b/m.

Chest x-ray showed several rib fractures on the right side, associated with bilateral hydrothorax, upper pneumomediastinum; mild right pneumothorax. After the first day the patient refused hospital care and discharged himself but a few hours later, he went to another hospital due to persistent pain. When he arrived was mildly dyspnoic and had bilateral basal pleural effusion. During his second hospitalization, he received antibiotics, anti-thromboembolic, anti-hypertensive, and gastroprotective therapies, and had progressive improvement of his clinical conditions. Six days after release, the patient suffered cardio- respiratory arrest and was not responsive to rescue procedures. Autoptic histological finding were mainly in the lungs that showed conspicuous bilateral pulmonary hemorrhage associated with perimedial fibroplasia variant of FDM, with aneurismatic and dissecant patterns, in lack of pulmonary embolism.

Case 2: A 31-year-old female patient was scheduled for rhinoplasty. Presurgical hematochemical and cardiovascular examinations were normal and anesthesiological risk class was ASA1. Twelve minutes after general anaesthesia induction and immediately after infiltration of nasal mucosa with mepivacaina and adrenalina, a rapid decrease of both oxygen saturation and cardiac frequency occurred until there was irreversible cardiocirculatory arrest, and no response to rescue procedures. Autoptic histological finding were mainly in the lungs that showed vascular congestion and acute focally hemorrhagic edema, associated with FDM, perimedial fibroplasia type. Chemical- toxicological analysis for research of Mepivacaina levels showed non- toxic concentration.

Conclusions: In both described cases death was referable to complications of FMD (aneurismatic dissection, arterial obstruction) most likely triggered by chest trauma in the first case and by possible cardiotoxicity due to association local/general anestesia.

Fibromuscolar Dysplasia, Pulmonary Arteries, Histopathology

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