Neonatal Marfan Sydrome with Pulmonary Artery Cystic Degeneration

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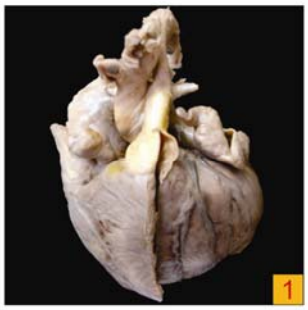
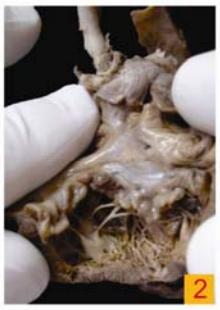


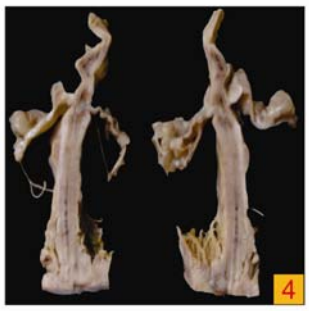
CASE PRESENTATION

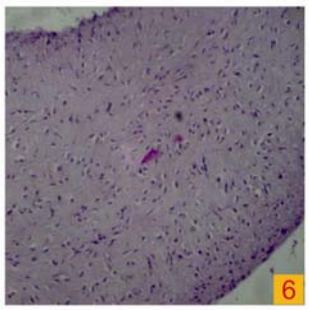
The infant was a female at the 31st week of gestation referred because of cardiomegaly associated with tricuspid and mitral dysmorphogenesis, leading to severe insufficiency detected on ultrasonogram. The child was born at 32nd week of gestation, needed resuscitation, persisted with respiratory discomfort and died on 2nd day of life.



AUTOPSY FIDINGS

Autopsy examination revealed micrognathia and arachnodactyly. Cardiomegaly with concordant cardiac connections, soft and billowy mitral and tricuspid valves were observed. Right ventricle and atrium were dilated. In addition, ascending pulmonary artery showed a sacular dilatation (fig 1). Histopathology revealed elastin fragmentation (fig 11) with alcian blue positive deposits (fig 8) caracterizing cystic degeneration of pulmonary artery´s media. These findings were not observed in aorta. Mitral and tricuspid valves showed myxomatous stromal degeneration (fig 6). Other relevant findings were bilateral pneumonia, hyaline membrane disease and passive congestion of liver and spleen. A post-mortem diagnosis of Marfan syndrome was made.





CONCLUSIONS

Marfan syndrome frequently affects aorta causing cystic media degeneration in adults. In this case report, authors suggest that pulmonary artery´s hypertension caused by peculiar neonatal circulation, associated with bilateral pneumonia, resulted in media lesions on pulmonary artery but not on aorta.