Massive biventricular rhabdomyoma in a neonate

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ABSTRACT

Rhabdomyoma is a well characterised entity in a neonate. Herein, we report a massive biventricular rhabdomyoma in a neonate presenting with cyanosis and congestive heart failure which was confirmed on autopsy. The report is for documentation of an unusually large tumour.

Keywords: Everolimus, rhabdomyoma, tuberous sclerosis

Rhabdomyoma as the most common benign cardiac tumor presenting in infancy is a well-characterized entity, and its association with tuberous sclerosis is also well known.[1] Most patients with small tumors are asymptomatic, but some patients may present with arrhythmias, obstruction, and rarely embolism.[2] Spontaneous partial or complete regression of rhabdomyomas during growth is the rule.[1,3] We recently encountered a 20-day-old neonate in congestive heart failure with cyanosis resulting from massive multiple biventricular rhabdomyoma [Figures 1a and b and Supplementary Videos 1a and b] and wish to document the same in view of its size. The neonate also had macules over the chest suggestive of tuberous sclerosis, but further evaluation could not be performed. He was planned for debulking surgery but had ventricular fibrillation from which he could not be revived. An autopsy confirmed the diagnosis [Figures 2a-c]. More recently, salutary effects of everolimus (mammalian target of rapamycin) in

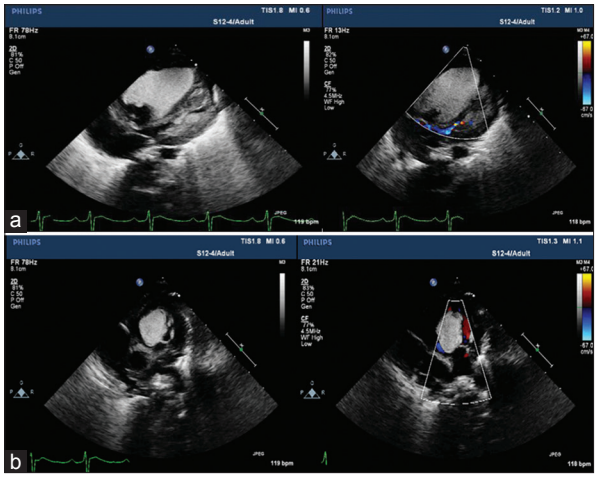

Figure 1: (a) Subcostal view. (b) Parasternal short axis

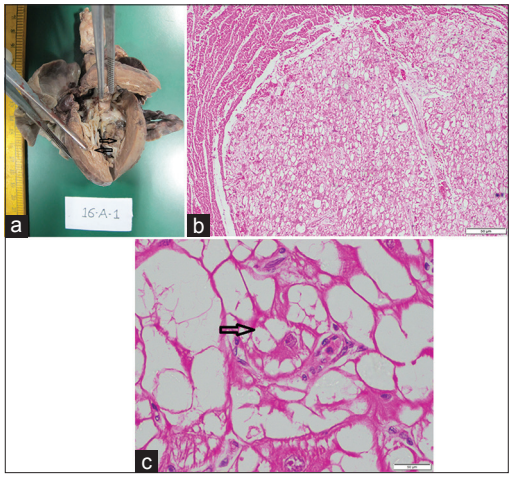

Figure 2: (a) Cardiac rhabdomyoma: Gross picture of the ventricular cavity shows multiple sessile gray-white nodules of varying sizes attached to the ventricular wall (arrows). (b) Microscopy of these nodules revealed a well-demarcated tumorous lesion with adjacent compressed normal myocardium (H and E, ×4). (c) Microscopy of these nodules on high magnification shows classical spider cells (arrow) of rhabdomyoma (H and E, ×20)

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regression of rhabdomyomas have been reported and possibly could have been utilized in this neonate.[4,5]

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Conflicts of interest

There are no conflicts of interest.

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