MRI Verification of a Case of Huge Infantile Rhabdomyoma

Naser Ramadani[1,3], Kreshnike Dedushi Kreshnike[1,4,5], Sefedin Muçaj[1,3], Serbeze Kabashi[1,4], Astrit Hoxhaj[6], Naim Jerliu[1,3], and Ramush Bejiçi[1,2]

[1]Faculty of Medicine, Pristine University, Pristine, Kosovo
[2]Pediatric Clinic, Department of Cardiology UCCK, Pristine, Kosovo
[3]National Institute of Public Health of Kosovo, Pristine, Kosovo
[4]Department of Radiology, Diagnostic Centre, UCCK, Pristine, Kosovo
[5]International Health Center "IHC" Pristine, Kosovo
[6]Hospital Hygeia, Tirana, Albania

Corresponding author: Dedushi Kreshnike, MD. Faculty of Medicine, Pristine University, Department of Radiology–Diagnostic Centre, UCCK and International Health Center "IHC" Pristine, Kosovo. ORCID ID: http://orcid.org/0000-0003-3639-0891. Tel.: +377 45 266-015. E-mail: kreshnikededushi@gmail.com

doi: 10.5455/aim.2016.24.146-148

ACTA INFORM MED. 2016 APR; 24(2): 146-148

Received: JAN 15, 2016 • Accepted: MAR 11, 2016

ABSTRACT

Introduction: Cardiac rhabdomyoma is type of benign myocardial tumor that is the most common fetal cardiac tumor. Cardiac rhabdomyomas are usually detected before birth or during the first year of life. They account for over 60% of all primary cardiac tumors. **Case report:** A 6 month old child with coughing and obstruction in breathing, was hospitalized in the Pediatric Clinic in UCCK, Pristine. The difficulty of breathing was heard and the pathological noise of the heart was noticed from the pediatrician. In the echo of the heart at the posterior and apico-lateral part of the left ventricle a tumoral mass was presented with the dimensions of 56 x 54 mm that forwarded the contractions of the left ventricle, the mass involved also the left ventricle wall and was not vascularized. The right ventricle was deformed and with the shifting of the SIV on the right the contractility was preserved. Aorta, the left arch and AP were normal with laminar circulation. The pericard was presented free. Radiography of thoracic organs was made; it resulted on cardiomegaly and significant bronchovascular drawing. It was completed with an MRI and it resulted on: Cardiomegaly due to large tumoral mass lesion (60x34 mm) involving lateral wall of left ventricle. It was isointense to the muscle on T1W images, markedly hyperintense on T2W images. There were a few septa or bant like hypointensities within lesion. On postcontrast study it showed avid enhancement. The left ventricle volume was decreased. Mild pericardial effusion was also noted. Surgical intervention was performed and it resulted on the histopathological aspect as a huge infantile rhadbomyoma. **Conclusion:** In most cases no treatment is required and these lesions regress spontaneously. Patients with left ventricular outflow tract obstruction or refractory arrhythmias respond well to surgical excision. Rhabdomyomas are frequently diagnosed by means of fetal echocardiography during the prenatal period.

Key words: Rhabdomyoma, fetal cardiac, epidemiology, MR, UCCK- Pristine.

1. INTRODUCTION

Cardiac rhabdomyomas are usually detected before birth or during the first year of life. They account for over 60% of all primary cardiac tumors (1, 2). Studies have demonstrated that the incidence of cardiac rhabdomyoma is 0.002-0.25% at autopsy, 0.02-0.08% in live-born infants, and 0.12% in prenatal reviews (3). Cardiac rhabdomyoma is the most common primary pediatric tumor of the heart and is considered to be a hamartoma of developing cardiac myocyte s (4-6). Although the behavior of a cardiac rhabdomyoma is benign, the positioning within critical areas in the heart can lead to lethal arrhythmias and chamber obstruction. The natural history of cardiac rhabdomyomas is one of spontaneous regression. Rhabdomyomas may be either cardiac or extra cardiac. Extracardiac forms of rhabdomyoma are sub classified into three distinct types: adult type, fetal type and genital type. Epidemiology: Cardiac rhabdomyomas are often multiple and can represent up to 90% of cardiac tumors in the pediatric population. The majority are diagnosed before the age of 1 year. The estimated incidence is at ~1 in 20,000 births (7). The majority of cardiac rhabdomyomas are asymptomatic although there can be wide clinical spectrum. On occasion they may present with left ventricular outflow tract obstruction or refractory arrhythmias (8). In most cases no treatment is required and these lesions regress spontaneously. Patients with left ventricular outflow tract obstruc-

tion or refractory arrhythmias respond well to surgical excision. The overall prognosis is dependent on the number, size and location of the lesions as well as the presence or absence of associated anomalies.

2. CASE REPORT

A 6 month aged patient, in October 2015, is hospitalized at the Pediatric Clinic at the University Clinical Center of Kosovo (UCCK) on the pulmomogic unit. He was showing some non-specific symptoms like: cough and difficulty in breathing. Lab data: SE/30; Er: 3.78, Le: 10.4; Hgb: 10.5; Hct: 30.5; PLT: 191; Lymph: 56.4; Mono: 8.2; Gran: 35.4; Glycaemia: 10.16, 8.2., 9.7, 6.6; Creatinine: 57.3; Urea: 2.29; Total Protein: 68.5; Albumin: 47.0; ALT: 19; AST: 45; Total bilirubin: 1.9; CK: 86; CRP: 4.0; pH: 7.38 ,7.48; pCo 2: 40, 34; pO2: 41,71; Na: 137, 135; K: 4.7..,4.7; Ca: 1.30; HCO3: 23.7, 25.3; BE: 1.3, 2.1. The difficulty of breathing was heard and pathological noises of the heart was noticed from the pediatrician. EKG result: rhythm is sinus, HR=120/min, left axis, T negative in D1, AVL, there are no other pathological changes (Figure 1). Echography results on: in the echo of the heart at the posterior and apico-lateral part of the left ventricle a tumoral mass presented with dimensions of 56 x 54 mm that forwarded the contractions of the left ventricle, the mass involves also the left ventricle wall and was not vascularized. The right ventricle is deformed and with the shifting of the SIV on the right the contractility was preserved. Aorta, the left arch and AP were normal with

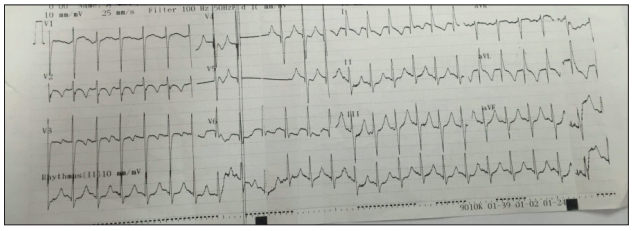

Figure 1. ECG: rhythm is sinusal, HR= 120/ min, left axis, T negative in D1, AVL, there are no other pathological changes. (Pediatric Clinic of the University Clinical Center of Kosovo, Pristina).

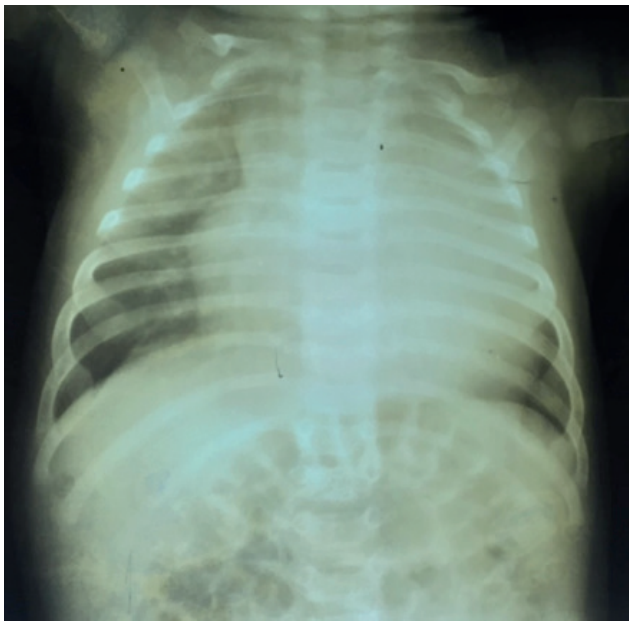

Figure 2. Radiography of thoracic organs results with cardiomegaly and significant bronchovascular drawing. (Department of Radiology in the University Clinical Center of Kosovo, Pristina).

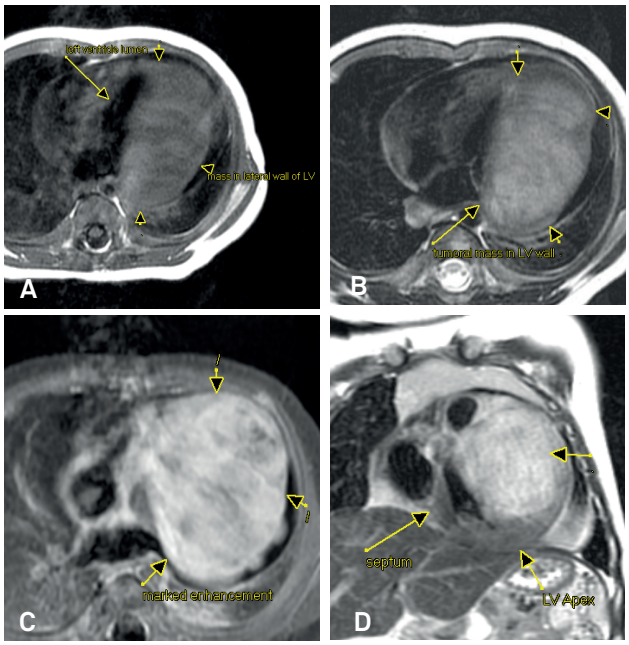

Figure 3. Pre contrast MRI images of the chest were obtained using TSET1W sequence in axial planes (a) Cardiomegaly due to large tumoral mass lesion (60x34 mm) involving lateral wall of left ventricle. It is isointense to muscle on T1W images; Pre contrast MRI images of the chest were obtained using TSE/T2W sequence with fat suppression in axial and coronal planes (b), it is markedly hyperintense on T2W images. There are a few septa or bant like hypo intensities with in lesion; Post contrast images were done using TSE / T1W sequence in axial planes (c, d) On post contrast study it show avid enhancement. Left ventricle volume is decreased. Mild pericardial effusion is also noted. (International Health Center "IHC" Pristina City, Kosovo)

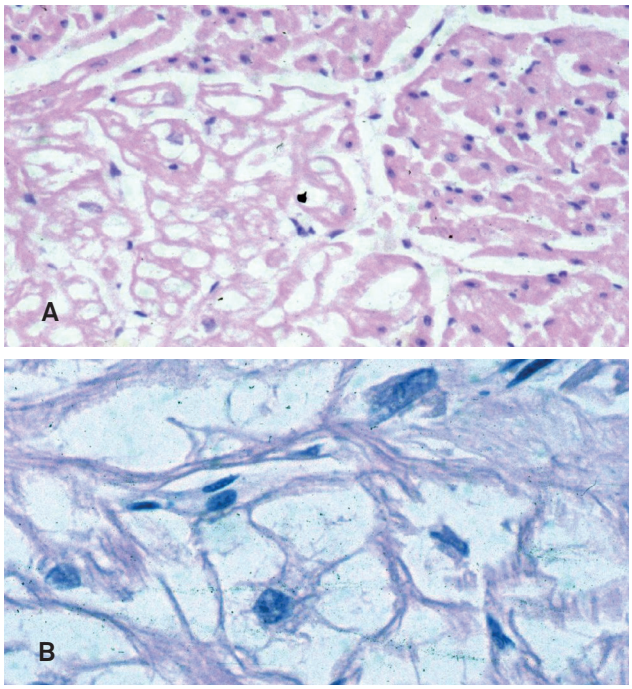

Figure 4. Histopathological images (a, b) – Huge infantile Rhabdomyoma (Institute of Pathology in the University Clinical Center of Kosovo, Pristina).

laminar circulation. The pericard is presented free. Radiography of thoracic organs is made; it results on cardiomegaly and significant bronchovascular drawing, (Figure 2). It was completed with MRI and resulted on: Cardiomegaly due to large tumoral mass lesion (60x34 mm) involving lateral wall of left ventricle. It was isointense to

muscle on T1W images (Figure 3 a), markedly hyperintense on T2W images (Figure 3 b), there were a few septa or bant like hypointensities within the lesion. On post contrast study it showed avid enhancement. Left ventricle volume is decreased. Mild pericardial effusion is also noted (Figure 3c, 3d). The patient was referred to me with final diagnosis: Tumor left ventricle massive, insufficient cardiorespiratory to a specialized Cardio-surgical centre in Tirana where he underwent a surgical intervention. The histopathological analysis proved that this was a case with huge infantile rhadbomyoma (Figure 4a, 4b). Patient stayed at intensive care and after 3 days will be released home. After 2 days his condition complicated again and he gets hospitalized in intensive care in UCCK and after 8 days of monitoring and caring the patient dies (exitus letalis).

3. DISCUSSION

Cardiac rhabdomyoma is a type of benign myocardial tumor and is most commonly fetal (9). A rhabdomyoma is a benign tumor of striated muscle. Rhabdomyomas may be either "cardiac" or "extra cardiac" (occurring outside the heart). Extracardiac forms of rhabdomyoma are sub classified into three distinct types: Adult type, fetal type and genital type. Cardiac rhabdomyomas are the most common primary tumor of the heart in infants and children. It has an association with tuberous sclerosis (10). Compared to other soft-tissue tumors; rhabdomyoma is extremely rare in the United States. Specifically, in the category of benign primary tumors of the heart, rhabdomyoma has a relative incidence of 5.8%. Worldwide, rhabdomyoma is also rare. Exact data regarding its incidence within particular populations has not been cited. In world literature, 14 cases of multifocal adult head and neck rhabdomyoma are reported (11). Adult rhabdomyoma occurs in older adults (usually >40 years). Fetal rhabdomyoma occurs between birth and the age of 3 years. Genital rhabdomyoma is observed in young and middle-aged women. Cardiac rhabdomyomas occur chiefly, but not exclusively, in the pediatric age group; a 45-year single-institution review found rhabdomyoma to account for 58% of cardiac neoplasms in 64 pediatric patients (age <18 years) who presented for surgical evaluation of a cardiac tumor (12). Rhabdomyomatous mesenchymal hamartoma of the skin is observed in newborns and infants. Adult rhabdomyoma has been diagnosed mostly in men. Some reports exist of cases in women. Fetal rhabdomyoma affects boys. Genital rhabdomyoma affects young and middle-aged women. Cardiac rhabdomyoma is observed in men and women. Rhabdomyomatous mesenchymal hamartoma of skin is observed in male and female newborns and infants. Rhabdomyoma has been identified in all racial groups. No predilection for any particular racial group exists.

Ultrasound/echocardiography: May be seen as one or more solid hyper echoic mass/es located in relation to the myocardium. Small lesions can mimic diffused myocardial thickening. They frequently occur in relation to the ventricles. The size for lesion detected in the uterus may range from ~10-50 mm (13). *MRI:* T1: relatively well defined mass/es isointense to adjacent myocardium, T2: relatively well defined mass/es hyperintense to adjacent myocardium

4. CONCLUSION

In most cases no treatment is required and these lesions regress spontaneously. Patients with left ventricular outflow tract obstruction or refractory arrhythmias respond well to surgical excision. The overall prognosis is dependant on the number, size and location of the lesions as well as the presence or absence of associated anomalies. Rhabdomyomas are frequently diagnosed by means of fetal echocardiography during the prenatal period. The prognosis of patients with rhabdomyomas is chiefly determined by the size and location of the lesion. Tumors larger than 20 mm in diameter are more likely to cause hemodynamic disturbances or arrhythmias, which are associated with an increased risk of death. Rhabdomyomas that obstruct the inflow or ventricular outflow tracts or that alter valve function and lead to regurgitation also carry a poor prognosis. Because the natural history of the rhabdomyoma is one of regression, most patients can be managed conservatively. There is a well known association with tuberous sclerosis, with over 50% of all cardiac rhabdomyomas found in patients with later confirmed tuberous sclerosis and congenital renal anomalies. Patients with rhabdomyomas should be aware of the nature of their particular tumors and the type of surgical procedures available for treatment. Any surgical procedure has a risk of a malpractice claim.

• Conflict of Interest: The authors have not conflict of interest to declare.

REFERENCES

1. Grebenc ML, Rosado de christenson ML, Burke AP. et al. Primary cardiac and pericardial neoplasms: radiologic-pathologic correlation. Radiographics. 2000 Jul- Aug; 20(4): 1073-103.
2. Uzun O, Wislon DG, Vujanic GM. Cardiac tumors in children. Orphanet J Rare Dis. March 2007; 2: 11.
3. Isaacs H Jr. Fetal and neonatal cardiac tumors. Pediatr Cardiol. 2004 May-Jun; 25(3): 252-73.
4. Becker AE. Primary heart tumors in the pediatric age group: a review of salient pathologic features relevant for clinicians. Pediatr Cardiol. 2000 Jul-Aug; 21(4): 317-23.
5. Elderkin RA, Radford DJ. Primary cardiac tumours in a paediatric population. J Paediatr Child Health. 2002 Apr; 38(2): 173-7.
6. Holley DG, Martin GR, Brenner JI, et al. Diagnosis and management of fetal cardiac tumors: a multicenter experience and review of published reports. J Am Coll Cardiol. 1995 Aug; 26(2): 516-20.
7. Entezami M, Albig M, Knoll U. et al. Ultrasound Diagnosis of Fetal Anomalies. Thieme. Read it at Google Books - Find it in Amazon, 2003. ISBN: 1588902129.
8. D'addario V, Pinto V, Di naro E. et al. Prenatal diagnosis and postnatal outcome of cardiac rhabdomyomas. J Perinat Med. 2002; 30 (2): 170-5.
9. Chao AS, Chao A, Wang TH et al. Outcome of antenatally diagnosed cardiac rhabdomyoma: case series and a meta-analysis. Ultrasound Obstet Gynecol. 2008; 31(3): 289-95.
10. Evans JC, Curtis J. The radiological appearances of tuberous sclerosis. Br J Radiol. 2000; 73 (865): 91-8.
11. Delides A, Petrides N, Banis K. Multifocal adult rhabdomyoma of the head and neck: a case report and literature review. Eur Arch Otorhinolaryngol. 2005 Jun; 262(6): 504-6.
12. Linnemeier L, Benneyworth BD, Turrentine M, Rodefeld M, Brown J. Pediatric cardiac tumors: a 45-year, single-institution review. World J Pediatr Congenit Heart Surg. 2015; 6(2): 215-9.
13. D'addario V, Pinto V, Di Naro E et al. Prenatal diagnosis and postnatal outcome of cardiac rhabdomyomas. J Perinat Med. 2002; 30(2): 170-5.