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# Intracardiac germ cell tumour in neonate with patent ductus arteriosus: An unusal case

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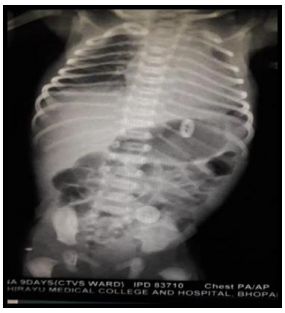
**Abstract**
Primary cardiac tumors are rare in pediatrics age group. Germ cell tumor although relatively common in mediastenum, but intracardiac mature teratoma with PDA are extremely rare histological finding reported in our department of pathology in 9 days old full term neonate.

**Keywords:** chronic myeloid leukemia, Philadelphia chromosome, e13a3, fluorescence

## Introduction
Primary cardiac tumors are rare forms of heart disease reported in both adult and children and first described by Realdo Colombo in 1559 [1, 2]. The autopsy prevalence of primary cardiac tumors reported in literature varies greatly 1 in 2000 while proportion of heart surgeries that are performed for tumors was only 0.45-0.85% while it is 0.093% in pediatrics series [3, 4]. 0.5% of neonate are born with congenital heart diseases which are more common in premature. Hence condition may have genetic reason or can be caused by environmental factors like infections or drug [3]. most of the congenital heart diseases causes obstruction in flow and other causes for flow obstruction are primary and secondary tumors of heart and cysts.
Intracardiac germ cell tumors are neoplasms of germ cell origin that arises with in myocardium or cardiac chambers. Manifestation depend on its location include heart block, right ventricular outflow tract obstruction with murmurs, seizer, and sudden death [5].

## Case Report
Full term female on her 9 days of life was referred to Cardio-thoracic vascular surgery unit of Chirayu Medical College and Hospital Bhopal Madhya Pradesh for evaluation of cardiac murmur detected during afebrile illness and complain of breathlessness and tachycardia. Physical examination revealed active alert, well nourished afebrile height of 46cm, and weight 2.2kg. Chest examination was normal and on auscultation of heart yielded a single soft S2 with grade 3/6 ejection systolic murmur loudest at left lower sternal edge, leading to clinical diagnosis right ventricular outflow tract obstruction. The liver was not enlarged.

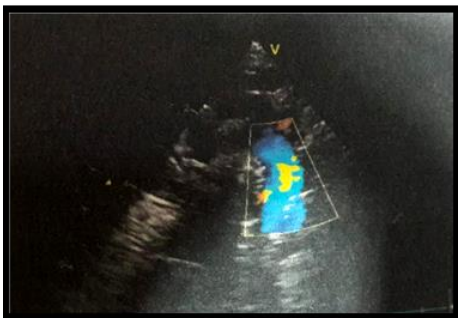
## Investigation
On radiography Chest PA/AP showed large homogeneous mediastinal opacity with mediastinal enlargement.




**PA view showed mediastinal enlargement**

**On 2D echocardiography and color doppler**

Shows tiny persistent foramen ovale(L-R shunt), conotruncus including single large mass attached to pulmonary valve causing severe RVOT obstruction. Gradient 60 mmHg with patent ductus arteriosus 1.5 mm PDA L-R shunt.



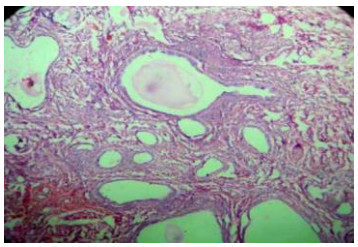
**On Gross examination**

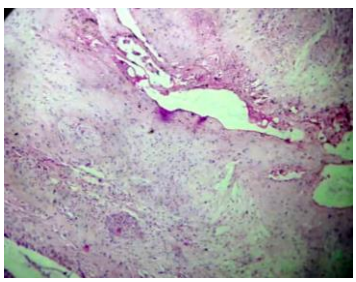
Single tubular soft tissue mass of size 4.5 *1.5*1 cm were noted on cut section shows irregular cystic cavities filled with mucinous material surrounded with solid areas.

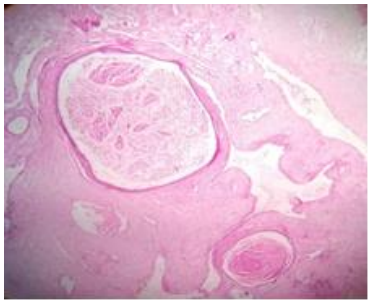
**Elongated soft tissue mass with mucin filled cavity.**

**On Microscopic examination**

Section from tumor shows multiple variable cystic cavities lined by simple columnar mucous secreting epithelium to a pseudo stratified ciliated epithelium surrounded by smooth muscles in addition aggregates of pericystic seromucous glands were present, Foci of squamous epithelium were identified with epidermal cyst filled with keratin, rare foci of cartilaginous differentiation, bone formation were also noted. The Histological study of the following resected intracardiac tumor supported the final diagnosis of benign teratoma.







**Discussion**

Pediatric cardiac tumors are extremely rare, and most (~90%) are histologically benign. In pediatric population, nearly 20% of primary cardiac tumours present within first year of life. Rhabdomyomas comprise most common pediatric cardiac tumors, followed by teratomas,fibromas and myxomas [6, 7, 8, 9]. Cardiac teratoma are the second most common primary tumor in neonates and fetuses.though most common germ cell tumor is intrapericardial teratoma, which attached to pulmonary artery and aorta. Intramyocardial teratoma typically rare and manifests with congestive heart failure [10]. Morphologically these tumors can show complex and varied aspects due to histological variety of their constituents, teratomas are distinct as they derived from one or several layers of germ cells. These finding explain the difficulties in diagnosis, faced sometimes, when the tumor is not well differentiated or immature. In our case tumor consisted of well differentiated mature teratomatous elements allowing a morphology based diagnosis with no need for immunohistochemistry studies. Teratoma is rare in any age group, but more than 75% occur below 15 years of age. It unusually arises in the pericardial cavity and often adherent to the great vessels; however intracardiac origin rarely occur [6].

Intracardiac teratoma attached to pulmonary valve and leads to right ventricular out flow obstruction is a rare finding which was encountered clinically and confined among pediatric age group in cardiac tumour mass.

Cardiac tumours produces large variety of symptoms through many mechanisms.

1. Mass obstruction leads to haemodynamic disturbance.
2. Local invasion can lead to arrhythmias or cardiac failure.
3. Causes pericardial effusion and cardiac tamponade.
4. Can embolize causing infarcts.
5. Causes mechanical hemolysis of red blood cells
6. Have biochemical effect with elevated ESR [11].

These sign can be detected early by radiography. 2D echocardiography play a crucial role in the diagnosis of intracardiactumours particularly when produces mass effect and produce breathlessness and tachycardia as in our case.

The differential diagnosis of intracardiac teratomas is represented by other most frequent cardiac tumours, specially the epithelial cyst of the myocardium. Other cardiac tumours may also have a cystic presentation particularly: the mesothelioma of the atrioventricular node, the myxoma, and vascular tumours such as haemangioma and sarcoma [12]

The definite diagnosis is made by surgical excision of mass from right ventrical and PDA was corrected simultaneously. On microscopy, areas squamous differentiation with

epidermal inclusion cyst and respiratory epithelium with seromucinous glands and smooth muscle noted some foci of cartilage was also seen give the definite diagnosis of mature teratoma.

To our knowledge, this is the first report of PDA associated with mature teratoma. The relationship between PDA and Mature teratoma is unknown in this case.

In conclusion, thorough large sampling of resected specimen should be performed before considering resected mass as benign, and intracardiac mature teratoma must be kept in mind when dealing with cardiac tumours.

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