

## Unusual involvement of right ventricle in patient with Rosai–Dorfman disease 🎧

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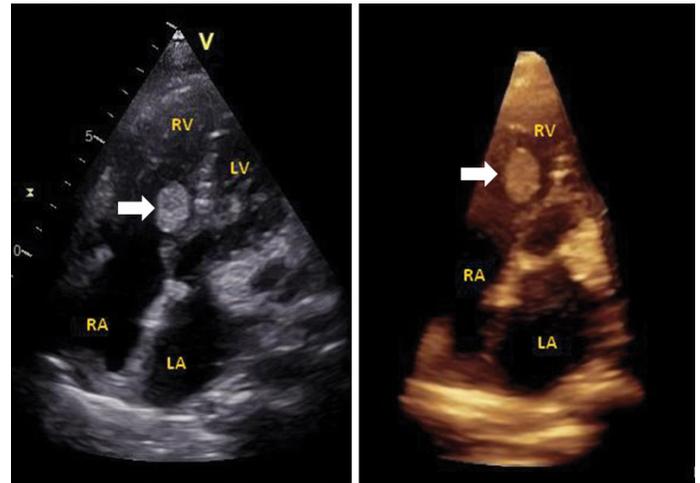
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### Introduction

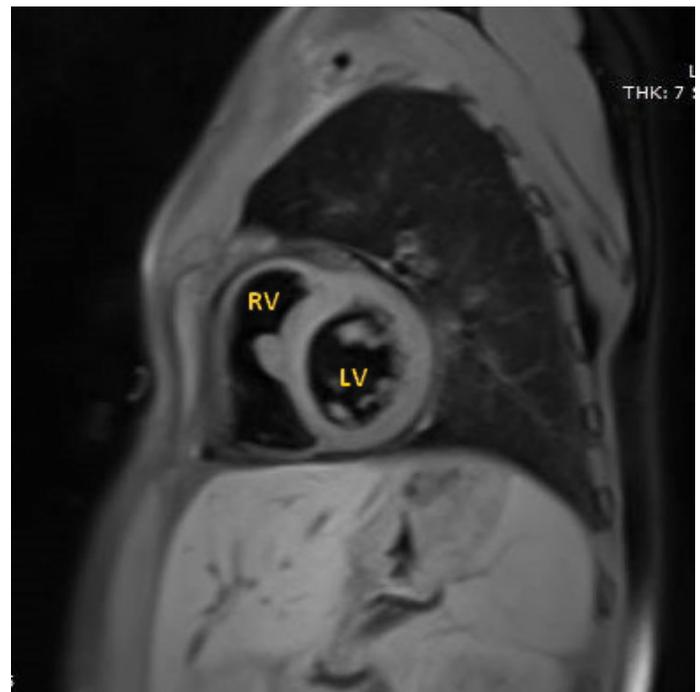
Rosai–Dorfman disease (RDD) is a benign form of histiocyte proliferation with unknown etiology that was first described by Rosai–Dorfman in 1969 (1). RDD is mostly presented with cervical lymphadenopathy, and also involves the extranodal system including skin, nasal cavity and paranasal sinuses, orbit, upper respiratory tract, and bone (2). RDD rarely involves the heart. There are only 19 reported cases of RDD in the literature, with none involving the right ventricle. In this case, we present RDD with the parotid gland, mediastinum, and cardiac involvement.

### Case Report

A 32-year-old female with type 1 diabetes mellitus, presented with bilateral swelling in her cheeks which had progressed during the past years. Ultrasonographic evaluation showed a 2 cm×3 cm mass in the right parotid gland. She underwent excisional biopsy and pathological diagnosis revealed sinus histiocytosis confirming RDD. She was referred to our cardiology department for the evaluation of extranodal involvement of disease. The patient had no cardiac symptoms at admission. In physical examination, bilateral swelling was found in her cheeks. She had no hepatic or splenic enlargement on physical examination. Electrocardiography showed a normal sinus rhythm. Echocardiography revealed a 15 mm×11 mm mass in the right ventricle with prolongation to the interventricular septum (Fig. 1). Magnetic resonance imaging (MRI) confirmed a well-circumscribed lesion in the right ventricle which was associated with interventricular septum (Fig. 2). Also, other masses were seen in the anterior mediastinum and nearby superior vena cava which were 12 mm×22 mm and 31 mm×22 mm×27 mm, respectively, and had properties similar to the cardiac mass. According to MRI results, lesions were described as cardiac and mediastinal involvement of RDD. Surgery was recommended for cardiac involvement; however, the patient refused. At 1 year follow up, the patient was asymptomatic, and there was no enlargement of the cardiac mass.



**Figure 1.** 2D and 3D echocardiography showed a 15 mm×11 mm mass in the right ventricle



**Figure 2.** Magnetic resonance imaging confirmed a well-circumscribed lesion in the right ventricle which was associated with interventricular septum

### Discussion

In this case, we presented a patient with right ventricle mass which was the cardiac involvement of RDD. Also, she had lymphadenopathy in the anterior mediastinum and right parotid gland; excisional biopsy and pathological assessment of this condition revealed the presence of RDD.

RDD primarily presents with painless cervical lymphadenopathy (up to 90% of cases) in childhood or adulthood. Extranodal sites are found in up to 43% of patients including skin, paranasal

sinuses, nasal cavity, orbit, soft tissues, and respiratory system. Less common affected extranodal sites are the central nervous system, urogenital, and gastrointestinal system (3, 4). Cardiac involvement of RDD is seen in <1% and there were 19 reported cases in the literature, out of which, 3 were reported in children (5). In adults, cardiac involvement is most often seen in the right atrium (n=7), followed by left atrium (n=3), epicardium (n=4), left ventricle (n=3), and pulmonary artery (n=2). In childhood cases, interatrial septum (n=2) and tricuspid as well as pulmonary valve (n=1) are involved as the extranodal sites (6).

Etiology of RDD is still unknown with several possible mechanisms proposed such as disorder of immune regulation or viral infections (Herpes virus-6, Epstein-Barr virus, cytomegalovirus) (4). Diagnosis of RDD is based on the pathological assessment. In immunohistochemical analysis, there is a positive reactivity of histiocytic cells with lymphocytes and macrophages due to emperipolesis, which is the abnormal phagocytosis of the autologous lymphocytes by histiocytes (1). In our case, the biopsy results verified the diagnosis of RDD in the parotid gland. We could not perform a biopsy of the heart mass because the patient refused surgery or any invasive treatment. According to the assessment of the MRI results, the mass in the heart had properties similar to the masses in the mediastinum and parotid gland.

Most of the patients with RDD have a stable disease; however, some of them have an aggressive pattern with the involvement of the extranodal sites. Generalised lymphadenopathy is a poor prognostic factor for RDD. Prognosis of RDD with cardiac involvement is largely unknown because of the limited cases. Treatment options are surgical excision, radiotherapy, corticosteroids, and chemotherapy. For patients with resectable lesions, surgical excision is a well-established treatment option with good survival. Besides this, O'Gallagher et al. (3) reported a patient with an intracardiac mass of RDD who was treated with corticosteroids, which led to little change in the size of the mass.

## Conclusion

In conclusion, we presented RDD with right ventricle, parotid gland, and mediastinum involvement. At 1 year follow up, the patient was asymptomatic and did not undergo any treatment such as radiotherapy, corticosteroids, and chemotherapy. Although cardiac involvement of RDD is a rare condition, it should be considered in patients who have been diagnosed with cardiac mass. However, because of the limited number of cases in the literature, the prognosis and treatment modalities with cardiac involvement are largely unknown.

**Informed consent:** An informed consent was obtained from the patient.

**Video 1.** Right ventricle mass was seen in the apical 4-chamber view.

## References

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## Successful right anteroseptal manifest accessory pathway cryoablation in a six-month infant with dyssynchrony-induced dilated cardiomyopathy

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## Introduction

Ventricular preexcitation via an accessory pathway (AP) results in an asynchronous spread of ventricular depolarization that may lead to an abnormal regional wall motion and systolic dysfunction in patients with Wolff-Parkinson-White (WPW) syndrome, even without documented supraventricular tachycardia (1-3). It is known that adults and children with preexcitation-associated cardiomyopathy can completely recover following resynchronization