Case Report

An Unusual Co-occurrence of Lown-Ganong-Levine Syndrome with Neonatal Focal Lipomatous Hypertrophy of Interatrial Septum with Patent Foramen Ovale

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Abstract

We report an extremely rare co-occurrence of focal lipomatous hypertrophy of the interatrial septum (LHIS) with patent foramen ovale (PFO) in a neonate and electrocardiogram evidence of Lown-Ganong-Levine syndrome (LGL). Although atrial arrythmias in the form of supraventricular tachycardia like atrial premature contractions, atrial fibrillation, and atrioventricular block have been described in lipomatous hypertrophy interatrial septum, co-occurrence of LGL syndrome with LHIS has not been described in literature so far. Baby presented in the cardiology outpatient department for presurgical cardiological evaluation of congenital hydrocephalus. Typically, LHIS spares fossa ovalis and affects both side of the interatrial septum making interatrial septum dumbbell shaped. There is even no literature description of focal hypertrophy of interatrial septum in early infancy and child hood including neonatal period. Our case is a unique illustration of a rare co-occurrence of LGL syndrome in focal lipomatous hypertrophy of interatrial septum in a neonate with PFO.

Keywords: Focal, interatrial septum, lipomatous hypertrophy, patent foramen ovale

Introduction

Lipomatous hypertrophy of interatrial septum (LHIS) is characterized by benign fatty infiltration of the interatrial septum. It is commonly found in the elderly and obese patients as an incidental finding during echocardiography. The prevalence of LHIS is approximately between 1% and 8% in general population with higher incidence in women.[1] The incidence of LHIS increases with age, body mass, and chronic corticosteroid therapy. Focal LHIS in a neonate with LGL syndrome has not been described in literature so far. Both conditions are usually incidentally detected and are asymptomatic in nature.[2] Although focal hypertrophy of interatrial septum can present with atrial arrhythmias or superior vena cava syndrome if superior vena cava is encased, co-occurrence of LGL with LHIS has not been described. Histologically, LHIS is characterized by fatty infiltration between the myofibers of the interatrial septum.[3] Lipomatous hypertrophy of the interatrial septum typically occurs anterior to the fossa ovalis and it characteristically spares it.[3] It involves both sides of the interatrial septum with a characteristic dumbbell-shaped appearance of interatrial septum. Rarely, it also presents like a cardiac mass. The present case is unique to illustrate a focal LHIS involving the lower half of the septum only.

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Case Report

A 2-day-old new born baby boy presented to the cardiology outpatient department for presurgical routine cardiological evaluation of congenital hydrocephalus secondary to aqueductal stenosis. The child was term baby with normal birthweight of 2.8 kg without any history of perinatal asphyxia. The child was born out of nonconsanguineous marriage with no maternal history of exanthematous fever suggestive of congenital TORCH infection. Cardiovascular system examination was within normal limit. Electrocardiogram (EKG) revealed the presence of sinus rhythm with heart rate of 128 beats/min with the presence of short PR interval of 82 msec without the presence of delta wave suggestive of Lown-Ganong-Levine (LGL) syndrome [Figure 1]. ECG of Wolf Parkinson White (WPW) syndrome reveals short PR interval with delta wave and Mahaim fiber presents with only delta wave without the presence of short PR interval. Paroxysmal junctional re-entrant tachycardia remains a specific entity in the neonatal period with presence of short PR and delta wave and EKG evidence of right posteroseptal pathway, presenting as incessant tachycardia more than 12 h and characteristic initiation with a sinus beat. Echocardiography revealed the presence of focal echoluscent bulge below fossa ovalis suggestive of LHIS [Figure 2] without the presence of any capsule. Baby had an associated patent foramen ovale of 2 mm size with left to right shunt [Figure 3]. Interestingly, it spared the fossa ovalis along with upper half of the septum [Figures 4 and 5] and septum did not appear dumbell shaped. Baby had intact interventricular septum, had no associated patent ductus arteriosus or coarctation of aorta with normal biventricular function. Serum electrolytes, blood glucose, calcium, magnesium, and serum thyroid-stimulating hormone levels were within normal limit. Baby boy had LGL syndrome which is eventually rare to encounter in routine clinical practice and presence of LGL syndrome in LHIS is not yet described. We encountered a case of focal LHIS which is extremely rare to observe in a neonate; paradoxic to the rule that incidence of incidental detection of LHIS increases with age.

Discussion

LHIS is defined as fatty infiltration of the interatrial septum which is rare to encounter in routine clinical practice and seen in adults. It was first described by American pathologist John T Prior (1917-2007) at Syracuse College of Medicine, New York in 1964.[4] The incidence of LHIS increases with age. Characteristically, LHIS spares fossa ovalis and the septum looks dumbell shaped. Diagnosis of LHIS is made in computed tomography (CT) when a smooth well-marginated expansion of the interatrial septum is identified exceeding 2 cm in transverse diameter in adults. Cardiac MRI delineates LHIS with characteristics bilobar interatrial septal thickening with homogeneous high signal intensity similar to subcutaneous fat tissue. T_1 weighted MRI reveals high signal, T_2 weighted non-fat suppressed MRI reveals high signal, and T_2 fat suppressed MRI reveals low signal. Fatty nature of LHIS is well appreciated in fat-suppressed imaging. Fluro deoxyglucose (FDG) positron emission tomography reveals a moderate degree of FDG uptake in lipomatous hypertrophy. Previously, it was thought due to metabolic activity of brown adipose tissue,[5,6] recently, it is attributed to the presence of subclinical inflammation in the adipose tissue.[7]

Only differential diagnosis that exists in a neonate is a cardiac lipoma which has a characteristic encapsulated appearance in echocardiography. Although LHIS most often behaves as a benign anomaly, it may present with severe superior vena cava obstruction or intractable arrhythmia in which cases surgical excision with reconstruction of the interatrial septum is sometimes considered. As lipomatous hypertrophy occurs in the area of anterior and middle internodal pathways, their interruption could be the major reason for occurrence of supra ventricular arrhythmias in the form of atrial premature contractions, atrial fibrillation, atrioventricular block, syncope, and sudden cardiac death.[8,9] Lipomatous septal hypertrophy if very large gives rise to obstructive flow symptoms including dyspnea.[1,10]

Pathologists coin lipomatous hypertrophy as "lipomatous hyperplasia" because of unencapsulated deposition of mature adipose tissue with cells resembling brown fat with vacuolated cytoplasm with more centrally placed nuclei.[11,12] Typically, the fatty mass extends from above the aortic root to the level

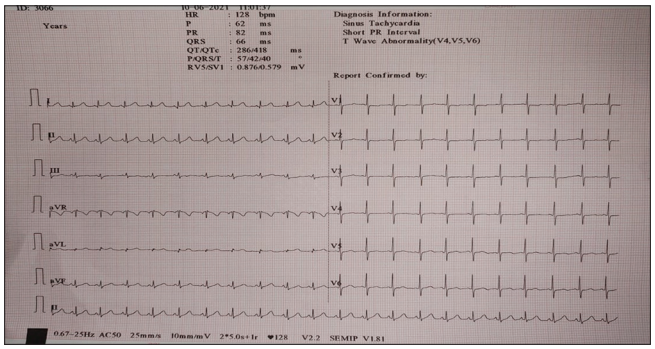

Figure 1: Lown-Ganong-Levine syndrome (PR interval 82 ms without the presence of delta wave)

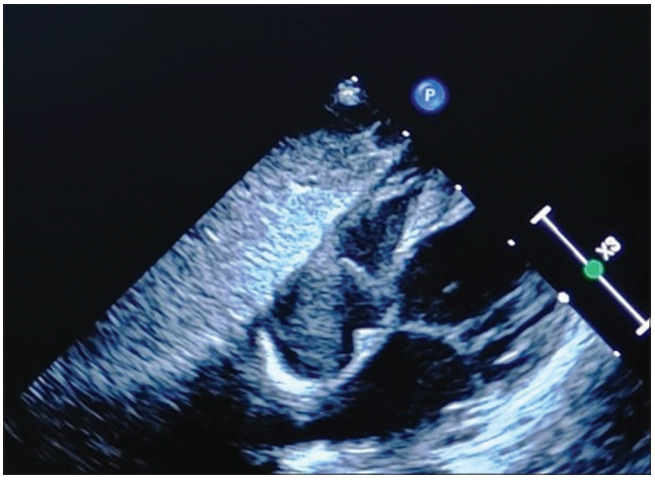

Figure 2: Focal lipomatous hypertrophy of lower IAS

of coronary sinus[13] but interestingly, in our case, it was a focal fatty infiltration limited to the lower interatrial septum only. Co-occurrence of LGL syndrome with focal LHIS has not been described in the literature so far.

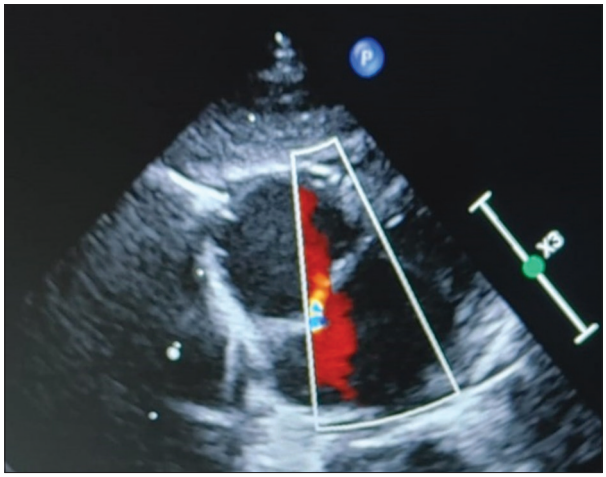

Figure 3: Presence of patent foramen ovale with L-R shunt

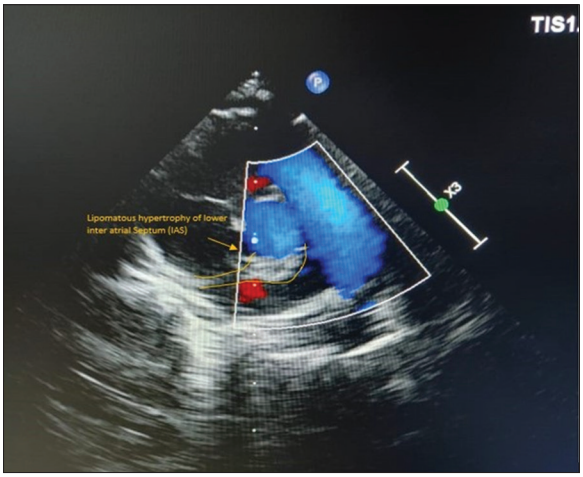

Figure 4: Lipomatous hypertrophy of lower inter atrial septum (IAS) in parasternal short axis (PSAX) view

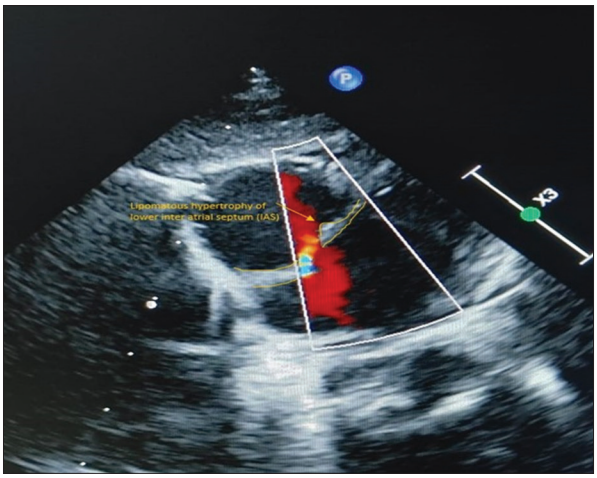

Figure 5: Lipomatous hypertrophy of IAS in subcostal modified apical 4 chamber view

LGL syndrome is a clinical syndrome consisting of paroxysms of tachycardia with ECG evidence of short PR interval (<120 ms) with normal QRS duration through atriohisian accessory pathway or James fibers. Even there is not any consensus about it, clinical tachycardia should be present for LGL diagnosis and otherwise short PR interval without delta wave is named as enhanced av nodal conduction. Clerc, Levy and Critesco in 1938 first described the occurrence of frequent paroxysms of tachycardia in patients with short PR interval and normal QRS duration.[14] This syndrome was again described by LGL in 1952 forming todays eponym.[15] James described atriohisian fibers originating in the low atrium and terminating in the His bundle.[16] Criteria for LGL include a PR interval <120 ms with normal QRS complex duration of <120 ms.[17] The most common tachycardia that occurs in LGL syndrome is atrioventricular nodal re-entrant tachycardia followed by accessory pathway-mediated tachycardia, atrial fibrillation, atrial flutter, and ventricular tachycardia.[18,19]

Lown and associates described tachyarrhythmia in 17% of patients with LGL syndrome.[15] The frequency of LGL is approximately 0.5% of the adult population. The incidence of paroxysmal supraventricular tachycardia in LGL syndrome appears to be 11%.[20] The average age of onset of tachycardia in LGL syndrome is 33.5 years.[20] The newborn had no documented supraventricular tachycardia in the past 24 h. No studies have shown an increased rate of sudden death or decreased survival in patients with LGL syndrome. They have a higher risk of developing tachyarrhythmia with sympathomimetics which is commonly used in children in cough remedies.[20] LGL described six patients with paroxysmal atrial fibrillation two of whom developed sudden cardiac death due to faster conduction through James fiber to the ventricle causing ventricular fibrillation.[15]

Adult patients are advised not to drive or operate public transport for 6 months after the most recent episode of syncope. The absence of a rapid heart rate does not exclude LGL as a possible diagnosis as the tachycardia in LGL is paroxysmal.[20] No specific therapy is indicated although there should be a focus on preventing tachyarrhythmia thhrough the accessory pathway. In acute setting of tachycardia, identifying the cause of tachycardia and controlling the ventricular rate is of utmost importance.[21] Baby was quite asymptomatic, no documented tachycardia in the last 24 h, both LHIS and LGL syndrome are relatively benign and incidentally detected, we manged the baby conservatively and advised the baby to undergo requisite surgery as advised by neurosurgeons.

Conclusion

Our case is unique illustration of co-occurrence of LGL syndrome in a case of LHIS in a neonate. Although both conditions are benign and incidentally detected, it further warrants a close scrutiny in adulthood if at all symptoms in the form of rhythm abnormality occurs. Baby should not be administered sympathomimetics, especially the

cough suppressants as both conditions may precipitate supraventricular tachycardia which can be catastrophic in the presence of accessory James fiber in LGL syndrome with rapid conduction to the ventricles.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that name and initials will not be published and due efforts will be made to conceal patient identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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