A Case of Chaotic Atrial Tachycardia and Noonan’s Syndrome*

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Abstract
Although chaotic atrial tachycardia (CAT) is a rare arrhythmia in children, some cases of Noonan’s syndrome have been reported to be related with CAT. We encountered an infant with CAT and Noonan’s syndrome. The present case showed atrial wall thickening and pulmonary interstitial consolidation just prior to the onset of CAT, which may indicate a relation between CAT and Noonan’s syndrome.

Case
A male baby with a birth weight of 1670g was born to nonconsanguineous parents at 30 weeks’ gestation. His medical history was significant for polyhydramnios and decreased fetal movement prenatally. His Apgar score was 4/6 at 1 and 5 minutes, respectively. A chest roentgenogram showed bilateral moderate pleural effusion. He was intubated and undertook continuous pleural drainage to relieve the respiratory distress. The patient had dysmorphic features consistent with Noonan’s syndrome, including hypertelorism, micrognathia, epicanthic fold, and a low-set, posteriorly rotated ear. Echocardiography demonstrated no abnormal findings including the atrial wall. At 8 days of age, he was fed using a nasogastric tube. Subsequent pleural drainage produced a considerable amount of chyloous fluid. The chylothorax did not respond to intravenous dexamethazone treatment. At 37 days of age, minocyclin pleurodesis was performed, which successfully eliminated the storage of pleural fluid. However, soon after the procedure, frequent atrial premature beats were recorded by an electrocardiograph monitor. The atrial premature beats progressed to frequent episodes of atrial tachycardia with a ventricular rate of more than 250 beats/min. A bolus injection of adenosine triphosphate revealed multiple ectopic P waves including several episodes of atrial fibrillation, which was diagnosed as CAT (Figure 1). A repeated echocardiography demonstrated a thickened interatrial septum (4.5 mm in thickness) and atrial free wall (Figure 2A). Chest X-ray showed a newly-appeared patchy bilateral consolidation with diffuse reticular interstitial shadow (Figure 3). At 53 days of age, the patient was extubated and spontaneously breathed oxygen. At 4 months of age, mild dyspnea and CAT persisted. However, the ventricular rate was controlled under the administration of digoxin (0.01mg/kg/day) and propranolol (2mg/kg/day). At 8 months of age, the thickness of the interatrial septum returned to normal (2.8mm in thickness), with improvement of CAT (Figure 2B).

Discussion
CAT is a rare tachyarrhythmia in infants and children. The condition is defined by the following electrocardiographic criteria [1]: a minimum of three P-wave morphologies; a lack of a dominant atrial rhythm; isoelectric intervals between P waves; varying PP, RR, and PR intervals; and average atrial rates greater than 100/min during tachycardia. Patients with CAT often have other atrial arrhythmia, including atrial fibrillation, atrial flutter, and atrial premature beats. With regard to the mechanism, little evidence is available, although it is known that CAT is related with acute or chronic obstructive lung disease in adults. However,
in children, several cases of Noonan’s syndrome have been reported to have this rare arrhythmia [2, 3], suggesting that there is a certain relation between Noonan’s syndrome and CAT. In those patients, one patient with Noonan’s syndrome had pleural effusion [2].

Noonan’s syndrome is a developmental disorder characterized by facial dysmorphism, short stature, cardiac defects, and skeletal malformations. Lymphatic dysplasia is present in less than 20% of patients with Noonan’s syndrome [4]. Complications of the abnormal lymphatic vessels include chylothorax, chyloperitoneum, and lymphedema [5-7].

In the present case, the thickened atrial wall could not be explained hemodynamically, and the general-
ized lung reticular consolidation appeared just prior to the onset of CAT. CAT appeared soon after the pleurodesis. Although we have not obtained histological data, one possible speculation is that lymphangiectasia may have occurred at the atrial wall and the lung interstitium because the storage capacity in the pleural space was eliminated due to the minocyclin pleurodesis. Lymphangiectasia may induce atrial wall lymphedema that results in the thickened atrial wall, and may cause the atrial electrical instability that progresses to CAT. Since, the patient showed no infectious signs before and after birth, it is unlikely that viral myocarditis caused the thickened atrial wall. As far as we are aware, this is the first case that suggests a relation between lymphangiectasia and the onset of CAT, and this may provide an important insight into the mechanism of CAT in patients with Noonan’s syndrome.

References