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Uhl Anomaly as a Maximum Variant of Arrhythmogenic Right Ventricular or Biventricular Cardiomyopathy

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ABSTRACT

The cause of Uhl anomaly remains unclear. A maximum variant of arrhythmogenic right ventricular or even biventricular cardiomyopathy is possible or a distinct unknown form of origin. **Method:** The shape of the right ventricle, histology and appearance the ECG findings were analyzed. **Results:** As a maximum variant of arrhythmogenic right ventricular or biventricular cardiomyopathy the shape of the intraventricular septum turns to a S-shaped form overlapping the left ventricle. In histology, lipomatosis appeared in new-born and early childhood and in later stages turns to pure fibrosis. The ECG of patients are likely to be arrhythmogenic right ventricular or in this single case biventricular cardiomyopathy but without localized right precordial QRS prolongation and terminal activation delay. In later stages complete right bundle branch block is more and more evident. **Conclusions:** There is evidence that Uhl anomaly might be the maximum variant of aarhythmogenic right ventricular or biventricular cardiomyopathy

Keywords: Uhl Anomaly, Arrhythmogenic Right Ventricular Cardiomyopathy, Low Voltage in Limb Leads, T-Wave Inversions, Fibrosis.

INTRODUCTION

In a little more than 40 cases typical Uhl anomaly are documented with a paper-thin right ventricular wall, no aneurysms and no trabecularisations of the right ventricle [1]. The right ventricle is grossly enlarged and poor contracting with a S-shaped aspect of the intraventricular septum overlapping the left ventricle [2]. The right ventricle is either completely [3] or in parts involved with parchment-like areas [4]. In histology, Uhl anomaly is characterised of missing heart muscle and missing fatty infiltration. Fibrosis is the dominant feature of Uhl anomaly possibly due to intrauterine apoptosis [5].

We recently reported on an 18-year old asymptomatic male with Uhl anomaly [6]. In this case the right ventricle was entirely paper-thin, had a diameter of 5 cm and was poorly contracting. The right ventricle had no aneurysms and no trablecularisation with late enhancement, the tricuspidal valve was slighty insufficient. The left ventricle was normally contracting and revealed no late enhancement by cardiac MRI.

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The question is whether Uhl anomaly and arrhythmogenic cardiomyopathy are distinct diseases or a progressive disease form arrhythmogenic cardiomyopathy to Uhl anomaly.

METHOD

This single patient was compared to experiences from 489 patients (310 males, mean age 42.1 ± 12.8 years) with right dominant, biventricular of left dominant forms of arrhythmogenic cardiomyopathy. In only 6 publications standard ECG was mentioned with right bundle branch block in most cases [2,7]. In the case of the young patient the ECG revealed low voltage in limb leads [8], epsilon waves [9], QRS fragmentation [10], T-wave inversions in inferior and precordial leads from lead V1 to V5 [8] and typical appearance of lead aVR with large Q waves, small R waves and negative T-waves [11]. ECG features include low voltage in limb leads, epsilon waves, typical features in lead aVR, QRS fragmentation, and T-wave inversions in inferior and precordial leads V1 to V5 – all typical criteria of arrhythmogenic biventricular cardiomyopathy, but localized right precordial QRS prolongation and terminal activation delay are missing.

Apoptosis can be suggested as a mechanism of arrhythmogenic cardiomyopathy in later stages [12] and in Uhl anomaly intrauterine [5].

RESULTS

A bulging of the left ventricular septum is present [13] due to increased right ventricular pressure, but without late enhancement of the left ventricle. Uhl anomaly is electrocardiographically defined as arrhythmogenic biventricular cardiomyopathy [3] without localized right precordial QRS prolongation and terminal activation delay as a very unique finding. But by the age of the patient it is a reasonable suggestion that intrautrine apoptosis is the correct mechanism of the development of Uhl anomaly as the maximum variant of arrhythmogenic right ventricular or biventricular cardiomyopathy with age-dependant loss of fatty infiltration and complete loss of myocardium with entire paper-thin right ventricular wall diagnosed by cardiac MRI of the case.

An earlier progression of the disease in new-born or early childhood is the development of complete right bundle branch block as documented in other papers [2,6] is a typical electrocardiographic finding of arrhythmogenic right ventricular cardiomyopathy [14] and Uhl anomaly, too. Even this electrocardiographic finding seems to be a hint that Uhl anomaly might be the maximum variant of arrhythmogenic right ventricular or biventricular cardiomyopathy.

What is generally missing, is a thorough ECG analysis of all case reports of Uhl anomaly from new-borns, early childhood and elder patients.

The theory is, that Uhl anomaly due to intrauterine apoptosis is the maximum variant of arrhythmogenic right ventriculara or biventricular cardiomyopathy. In other cases, apoptosis in older patients leads to arrhythmogenic cardiomyopathy – right dominant, biventricular or left dominant.

In the described patient the family history is interesting, as the father oft he patient died form sudden cardiac death a few weeks after appendektomy. Whole sequencing of desmosomal and non-desmosomal mutations of arrhythmogenic cardiomyopathy are analyzed.

DISCUSSION

In this case the key to diagnosis is standard ECG. The case represents electrocardiographically arrhythmogenic biventricular cardiomyopathy, but in fact, the left ventricle is not involved neither echocardiographically nor by cardio MRI. The mechanism of ECG is unclear, suggesting that Uhl anomaly might be the maximum variant of arrhythmogenic right ventricular cardiomyopathy. Early intrauterine apoptosis may be a very progressive development even in new-born or early childhood. In this means, I remember a slide from Guy Fontaine in a 3 months old patient with a little fatty infiltration and a huge amount of fibrosis and missing myocardium from autopsy findings. In later years Uhl anomaly is characterised by missing myocardium and fibrosis without fatty infiltration – a feature of disease progression.

In more than 40 cases the ECG's must be described correctly and sampled case by case. Only in this manner the question can be answered correctly whether Uhl anomaly is the maximum variant of arrhythmogenic right ventricular cardiomyopathy. Another cause must be found why low voltage in limb leads is not a depolarization criterium of left ventricular disease. With regard to the dominance of right ventricular disease with S-shaped intraventricular septum overlapping the left ventricle

it is a suggestion that the role of left ventricle is irrelevant and low voltage in limb leads is not a sign of additional left ventricular disease, but a hint of beginning heart failure.

The ECG suggests that the right diagnosis is arrhythmogemic biventricular cardiomyopathy, although the left ventricle is normal by echocardiography and cardiac MRI. May be,

the ECG of the patient is the early hint of left ventricular dysfunction.

CONCLUSIONS

There is evidence, that Uhl anomaly might be a maximum variant of arrhythmogenic right ventricular or biventricular cardiomyopathy due to a shape of the right ventricle overlapping the left ventricle, to histology with a great amount of lipomatosis in new-born and early childhood and in later years with pure fibrosis and the appearance of ECG findings with typical signs of arrhythmogenic biventricular cardiomyopathy but without localized right precordial QRS prolongation and terminal activation delay ending up in complete right bundle branch block.

LIMITATIONS

There are only 44 publications on Uhl anomaly with a great lack of ECG and histologic findings. Most reports are on new-born or early childhood patients and in few cases are on adult cases with and without any complaints.

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CONFLICTS OF INTEREST

The author declares that no conflicts of interest.

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