Kearns-Sayre syndrome: electro-vectorcardiographic evolution for left septal fascicular block of the his bundle

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Received 1 March 2008

Abstract

The Kearns-Sayre syndrome is a neuromyopathic disorder associated with mitochondrial abnormalities and characterized by the triad of chronic external ophthalmoplegia, atypical pigmentary retinopathy, and progressive conduction system disorders. Ragged red muscle fibers that seem to contain an excess of altered mitochondria are observed. The disease affects both sexes alike, during the first or the second decade of life. The following manifestations are observed: central bilateral sensorineural deafness, pyramidal signs, ataxia, asymmetrical ptosis, external ophthalmoplegia, and progressive muscular weakness secondary to myopathy associated with a significant increase of proteins of cephalorachidian liquid. A variety of endocrinopathies may occur.

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Keywords: Kearns-Sayre syndrome; Prominent anterior forces; Fascicular blocks; Left septal fascicular block; Trifascicular block

Case report

A 23-year-old man of African descent was referred to a cardiologist for evaluations based on identified alterations in retinal examination that are commonly associated with cardiac disease.

History of current problems

The patient reported progressive muscular weakness over the previous 2 years, which required interruption of regular practice of soccer 3 months before this presentation. Sudden and intermittent double-vision developed 20 days ago, and ptosis of his left eye occurred 10 days previously resulting in referral for a detailed ophthalmologic evaluation. On examination of his visual fields, he was shown to have tubular vision, whereas the presence of external ophthalmoplegia explained the double vision. He was also found to have an early nuclear cataract in the right eye. Angiofluoresceinography of the retina showed atypical retinal pigmentary degeneration. The presence of the retinal pigmentation and external ophthalmoplegia raised concerns about Kearns-Sayre Syndrome, and he was referred for a cardiac evaluation.

In 1986, at the age of 6 years, he had a preoperative electrocardiogram (ECG) that was interpreted as normal. At the age of 9 years, an ECG obtained before an appendectomy showed incomplete right bundle branch block (IRBBB) (Fig. 1). Because of the abnormal ECG, an echocardiogram was obtained and reported as being normal. Three serological evaluations for Chagas disease were all
negative. These included the enzyme-linked immunosorbent
assay, indirect immunofluorescence, and indirect hemoagglutination tests.

Physical examination

The patient appeared to be in good health, had an ectomorphic build with a height of 1.56 m, weight of 64 kg, and blood pressure of 120/70 mm Hg. He had a normal precordial examination, left ptosis, and slight deafness (bilateral). Deep hyperreflexia was present bilaterally, and a Babinsky sign was positive in the right foot.

Supplementary examinations

- ECG (Fig. 2)
- Vectocardiogram (Fig. 3)
- An echocardiogram demonstrated indirect signs, a decrease in ventricular compliance consistent with diastolic dysfunction.

Discussion

Cardiac involvement in the Kearns-Sayre syndrome may manifest by different degrees of atrioventricular block through the His-Purkinje system, giving rise to bundle branch blocks (right or left), frequently associated to left fascicular blocks. The symptoms have an evolutionary character, with the potential of reaching complete or third-degree atrioventricular block requiring implantation of a permanent pacemaker.

In this report, we present the electrocardiographic evolution of the carrier of the syndrome, who initially had normal ECG tracing, evolved into IRBBB, and over the course of 17 years developed into a rare trifascicular block: complete right bundle branch block (CRBBB), left anterior fascicular block (LAFB), and left septal fascicular block (LSFB) (Figs. 2 and 3).

Since the pioneer publication of Durrer et al, the left branch of the His bundle divides into 3 anatomical-functional divisions. These serve to simultaneously activate the endocardium of the left septal surface at 3 points: the central region of the left septum, the anterosuperior region at the base of the papillary muscle of the mitral valve, and the posteroinferior in the base of the posteroinferior papillary muscle of the same valve.

The existence of a third division that originates from the main trunk of the left bundle has been labeled by a multiplicity of names including the left septal fascicle, middle or anteromedial division, and centroseptal fasciculus of the left branch. Its presence has been demonstrated by both anatomical dissections in humans and in dogs and careful analysis of the ventricular activation displayed on a 12-lead ECG or vectorcardiogram. Conduction disorders involving the left septal fascicle have been shown as a progressive or intermittent abnormality and by electrophysiologic study and surface endocardial mapping of both right and left ventricles. Using invasive methodology, Dhala et al demonstrated that the left septal fascicular delay/block manifests in ECG after transcatheter right bundle branch ablation. The multifascicular nature of left intraventricular conduction is more apparent when diseased and unmasked by concomitant block in the right bundle branch.

The left septal fascicle has, in most cases, fibers that originate in the angle formed by the emergence of other subdivisions of the left branch, as a separate tract. Less frequently, it originates as a ramification of the left anterior fascicle or the left posterior fascicle as a fan that cannot be individualized, or rarely (15%), it may not exist.
The left septal fascicle activates the middle-septal region of the left septal surface, being responsible for the vector from the initial 10 to 20 milliseconds, which is directed to the front and the right (85% of the cases) or to the front and the left (remaining 15%) depending on ventricular orientation.

When there is concomitant conduction delay in left septal fascicle and left anterior fascicle of the left branch, and by the right bundle branch (trifascicular block), initial septal activation depends exclusively on the left posterior fascicle, which is directed posteriorly, manifested by an initial small q wave that is usually seen in the right precordial leads from V1 to V2, or from V1 to V3, because in these circumstances, such activation heads anterior chest leads.

The diagnosis of LSFB is based on the presence of (a) prominent anterior QRS forces (PAF) not explained by right ventricular enlargement or posterior infarction; (b) the presence of a qR pattern from V1 to V3 in absence of clinical causes of right ventricular enlargement, misplacement of electrocardiographic precordial leads; (c) R wave of V2 greater than R wave in V3; (d) progressive decrease of R wave voltage from V4 to V6; (e) loss of septal initial q waves in left precordial leads V5-V6 by initial activation from right to left in the posteroinferior region of the left ventricle. There is also prolonged ventricular activation time longer than 35 milliseconds in V1-V2. The diagnosis of LSFB should always be based on clinical electro-vectorcardiographic criteria in absence of others clinical causes of PAF. Other causes of anterior forces include normal variant with counterclockwise rotation of the heart around the longitudinal axis; right ventricular enlargement (types A or B vectorcardiographic); lateral infarction; obstructive and nonobstructive forms of hypertrophic cardiomyopathy; preexcitation variant of Wolff-Parkinson-White syndrome, with accessory anomalous pathway located posteriorly; diastolic left ventricular enlargement secondary to septal hypertrophy; progressive muscular dystrophy, or Duchenne cardiomyopathy; dextroposition; and any combination of the above.

The presence of bifascicular block usually manifested as right bundle branch block and LAFB in the Kearns-Sayre syndrome is a class I indication for pacemaker implantation according to American College of Cardiology/American Heart Association guidelines. This is the first electro-
vectorcardiographically documented report of the presence of LSFB in the Kearns Sayre syndrome, as part of a variant of a very rare trifascicular block.

References