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CASE REPORT
Arrhythmias/electrophysiology

A case report of deglutition triggered atrial fibrillation in a patient with Laing distal myopathy

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Background Deglutition-induced atrial fibrillation is a rare clinical entity with a reported prevalence of 0.6%. Laing distal myopathy is a rare autosomal dominant muscular dystrophy that is the result of mutations within the slow skeletal muscle fibre myosin heavy chain gene (MYH7). Atrial fibrillation has not been previously reported in patients with Laing distal myopathy. We describe the first reported case of deglutition triggered atrial fibrillation in a female with a history of Laing distal myopathy.

Case summary A 44-year-old female with a history of Laing distal myopathy diagnosed at age 32, began experiencing intermittent episodes of pre-syncope and palpitations which occurred after deglutition with food. An ambulatory 30-day patient triggered event monitor recorded episodes of atrial fibrillation with rapid ventricular response. Family history was significant for Laing distal myopathy, atrial fibrillation, as well as sudden cardiac death. Laboratory data, transthoracic echocardiogram, cardiac magnetic resonance imaging, and an exercise treadmill SPECT Imaging stress test were normal. An oesophagram revealed a mild oesophageal dysmotility with no other abnormalities. She was started on flecainide 50 mg p.o. every 8 h and verapamil 40 mg p.o. every 8 h with no further episodes of atrial fibrillation.

Discussion Given the strong genetic component of this myopathy, one could postulate as to a possible genetic component in the development of atrial fibrillation in our patient. Although we cannot make definite correlation between deglutition-induced atrial fibrillation and Laing myopathy, it is important to report this unusual association which has not been described before.

Keywords Case report • Deglutition triggered atrial fibrillation • Laing distal myopathy • Wide-complex tachycardia • Flecainide and non-dihydropyridine calcium channel blockers

ESC Curriculum 5.3 Atrial fibrillation • 5.5 Supraventricular tachycardia

Learning points

- Recognize the unusual association of deglutition-induced atrial fibrillation in a patient with Laing distal myopathy.
- Flecainide in combination with non-beta blocker atrioventricular blocking agents such as verapamil can be effectively used for the treatment of vagally mediated atrial fibrillation.

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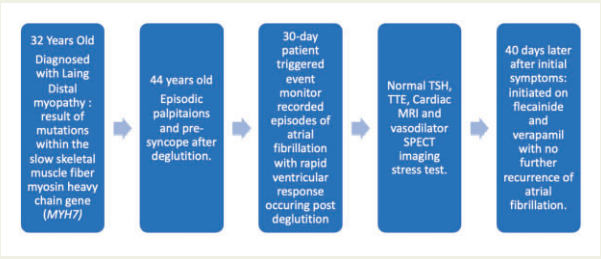
Introduction

Deglutition-induced atrial fibrillation is a rare clinical entity with approximately 50 reported cases since its initial description in 1926 and a reported prevalence of 0.6%.[1–3] Laing distal myopathy is a rare autosomal dominant muscular dystrophy with an unknown prevalence that is the result of mutations within the slow skeletal muscle fibre myosin heavy chain gene (*MYH7*). We describe the first reported case of deglutition triggered atrial fibrillation in a middle-aged female who has a history of Laing distal myopathy.

Timeline



Case presentation

A 44-year-old female with a past medical history of hyperlipidaemia and Laing distal myopathy diagnosed at age 32, began experiencing intermittent episodes of pre-syncope and palpitations which occurred intermittently after swallowing, predominantly during deglutition with food. Episodes of palpitations and pre-syncope post-deglutition occurred mostly with warm liquids or food intake and less frequently with cold liquids. There was no accompanying chest pain, shortness of breath, or odynophagia. She did not consume alcohol, caffeine and was a non-smoker. An ambulatory 30-day patient triggered event monitor recorded episodes of atrial fibrillation with rapid ventricular response (*Figure 1*) and wide-complex tachycardia with heart rates up to 242 b.p.m. (*Figure 2*). Each patient triggered event occurred after eating, associated with lightheadedness and/or palpitations. Family history was significant for Laing distal myopathy in her father who is still alive at 72 years old and atrial fibrillation in her mother.

Given the findings on ambulatory event monitoring, she was admitted to our hospital for further evaluation. Patient blood pressure on presentation was 123/64 mmHg, pulse 76 b.p.m. and regular, respiration rate 18 breaths/minute, weight 109 pounds, and body mass index of 22.07 kg/m^2. Oxygen saturation was 100% on ambient air. Remainder of the physical exam including respiratory exam was unremarkable. Laboratory data including a complete metabolic panel, troponin I, N-terminal prohormone of brain natriuretic peptide, and thyroid function tests were within normal limits.

Resting baseline electrocardiogram demonstrated sinus rhythm, right axis deviation. Transthoracic echocardiogram and cardiac magnetic resonance imaging were normal. An exercise treadmill SPECT Imaging stress test revealed normal left ventricular systolic function and no evidence of ischaemia or infarction. A fluoroscopic oesophagram revealed a mild oesophageal dysmotility.

During hospitalization, she had several episodes of atrial fibrillation with rapid ventricular response which coincided with deglutination. She was initially placed on metoprolol tartrate 12.5 mg twice daily with mild improvement of palpitations; however, she was still experiencing episodes of atrial fibrillation with rapid ventricular response after deglutination. Metoprolol tartrate was subsequently discontinued, and she was started on flecainide 50 mg p.o. every 8 h and verapamil 40 mg p.o. every 8 h. During her hospitalization, there were no recorded episodes of ventricular tachycardia. After initiation of flecainide and verapamil, there has been no documented recurrence of atrial fibrillation and her symptoms of palpitations have resolved.

Discussion

Laing distal myopathy is a rare autosomal dominant muscular dystrophy caused by mutations within the myosin heavy chain gene 7 (MYH7). The MYH7 gene encodes a beta-myosin heavy chain isoform that is expressed primarily in the heart and also in type I skeletal muscle fibres.[4] Mutations in the MYH7 gene is associated with various cardiac conditions including myosin storage myopathy, left ventricular non-compaction, familial restrictive, dilated and hypertrophic cardiomyopathy.[5]

Atrial fibrillation has been previously associated with several other myopathies including myotonic dystrophy type I, Emery–Dreifuss muscular dystrophy, as well as Duchenne and Becker muscular dystrophies. After an extensive review of the literature, we could not find any previous reports of atrial fibrillation in patients who were diagnosed with Laing myopathy. Thus far, the only previously

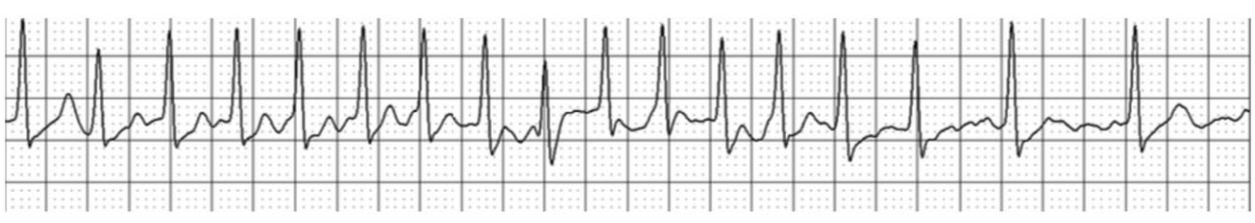


Figure 1 Ambulatory patient triggered cardiac event strip demonstrating atrial fibrillation with rapid ventricular response at 170 b.p.m. Event was triggered after deglutition.

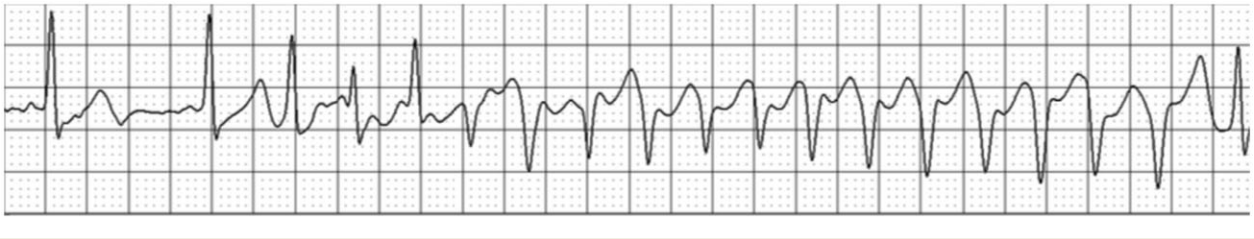


Figure 2 Ambulatory patient triggered cardiac event which reveals sinus rhythm with 75 b.p.m. followed by 13 beats of wide-complex tachycardia at 242 b.p.m. Event was triggered after deglutition.

reported cardiac involvement in a patient with Laing myopathy was one report of a cardiomegaly, however, no reports of atrial arrhythmias.[6]

Emerging evidence that supports a genetic and familial basis for atrial fibrillation is becoming increasingly recognized and is an area of research. Variants in genes encoding ion channel subunits, cardiac gap junctions, as well as loss in cytoskeletal proteins such as cytoskeletal protein 4.1R have been implicated in the arrhythmogenesis associated with atrial fibrillation.[7,8] Given the strong genetic component of this myopathy, one could postulate as to a possible genetic component in the development of atrial fibrillation in our patient. Furthermore, given the family history of our patient which includes paternal Laing myopathy and maternal history of atrial fibrillation, future studies may better elucidate the shared genetic component of patients with atrial fibrillation.

Although the pathophysiology behind deglutition triggered atrial fibrillation has not been fully elucidated, various mechanisms have been proposed. One such mechanism involves direct vasovagal parasympathetic stimulation via the afferent and efferent branches of the vagus nerve. These branches are activated during a rise in intraoesophageal pressure with preferential vagal discharges to the atria and not the sinus node, triggering atrial fibrillation without precipitation of bradycardia.[9] Vagal activation can also reduce the atrial effective refractory period.[10]

Initial management of deglutition-induced atrial fibrillation involves avoidance of food triggers, limiting large food boluses, caffeine, and alcohol use. For recurrent deglutition triggered atrial fibrillation despite conservative changes, pharmacologic medications can be considered. There is no universally agreed upon treatment for swallowing-induced atrial fibrillation, however, Class IC agents including propafenone or flecainide have been shown to be effective.[11,12] Amiodarone has also been successfully used,[11] however, given the potential long-term adverse effects, we opted to not use this agent in our relatively young patient. We therefore choose to prescribe flecainide given the lack of structural heart disease in our patient. In addition, we added a non-dihydropyridine calcium channel blocker verapamil to decrease the risk of 1:1 atrioventricular conduction with flecainide.[13] Verapamil can also provide a degree of suppression of triggered activity which might be responsible for atrial arrhythmogenesis.

Catheter ablation with pulmonary vein isolation (PVI) has been attempted successfully, although it has been reserved mostly for cases which are refractory to medical treatment.[14,15] Given the potential role that the autonomic nervous system may have in vagally mediated atrial fibrillation, catheter ablation of ganglionated plexi (GP), may potentially be curative for these patients. Calò *et al*. successfully utilized ablation of the GP in the right atrium in patients with vagally mediated atrial fibrillation.[16] Utilizing activation mapping of the left atrium to detect atrial tachycardia during swallowing, Challapudi *et al.*[1] were able to perform PVI of the right superior pulmonary vein and right anterior GP with resolution of symptoms in follow-up. Finally, combined PVI along with GP ablation have shown inconsistent results, however, increased success has been seen in patients with more pronounced autonomic triggers for atrial fibrillation.[10] Our patient wanted to avoid an invasive treatment approach and given that she was well controlled with medications, this was reasonable.

We choose against oral anticoagulation for our patient given that she had no risk factors other than being female which traditionally would give her a CHA2DS2-VASc of 1. Without any additional risk factors, female sex is only a risk modifier, is age dependent, and there is no difference in stroke risk based on gender alone.[13] Our patient was well controlled on both flecainide 50 mg orally every 8 h and verapamil 40 mg orally every 8 h with no further symptoms or documented recurrence of atrial fibrillation on subsequent 30-day cardiac event monitoring 1 year later.

Lead author biography

Chikezie K. Alvarez was born in 1986 in the Caribbean country of Trinidad and Tobago. He went to medical school at St. George's University which is also located in the Caribbean Island of Grenada. He is a second year Cardiology fellow at University of Connecticut/ Hartford Hospital and he hopes to pursue a career in electro physiology.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

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