Posterior spinal instrumented fusion for idiopathic scoliosis in patients with multisystemic neurodegenerative disorder: a report of two cases

Kwong Weng Loh,1 Chris Yin Wei Chan,2 Chee Kidd Chiu,1 Mohd Shahnaz Bin Hasan,2 Mun Keong Kwan1
1 Department of Orthopaedic Surgery, University of Malaya, Kuala Lumpur, Malaysia
2 Department of Anaesthesiology, University of Malaya, Kuala Lumpur, Malaysia

ABSTRACT

Mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke (MELAS) syndrome is a progressive multisystemic neurodegenerative disorder. MELAS syndrome impairs oxidative phosphorylation and predisposes patients to lactic acidosis, particularly under metabolic stress. We report 2 siblings with MELAS-associated idiopathic scoliosis who underwent posterior spinal instrumented fusion with measures taken to minimise anaesthetic and surgical stress, blood loss, and operating time.

Key words: MELAS syndrome; mitochondrial diseases; scoliosis

INTRODUCTION

Mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke (MELAS) syndrome is a progressive multisystemic neurodegenerative disorder caused by a single base A3243G point mutation in the mitochondrial transfer RNA \(^{Leu} \) (UUR) gene; the onset of symptoms is typically between the ages of 2 and 20 years.1 Scoliosis is not a typical clinical feature of MELAS.2 We report 2 siblings with MELAS-associated idiopathic scoliosis who underwent posterior spinal instrumented fusion with measures taken to minimise anaesthetic and surgical stress, blood loss, and operating time.

Case reports

Patient 1

In April 2007, a 10-year-old female presented with adolescent idiopathic scoliosis with a 38° main thoracic Lenke 1BN curve. She was first treated with a Boston brace, but the curve progressed to 55° 2 years later (Fig. a).

A posterior spinal instrumented fusion by 2 surgeons simultaneously was planned. The induction anaesthesia comprised intravenous propofol, atracurium, and fentanyl. During surgery, boluses

Address correspondence and reprint requests to: Kwong Weng Loh, Department of Orthopaedic Surgery, University of Malaya, Jalan Universiti, 50603 Kuala Lumpur, Malaysia. Email: melvinloh@gmail.com