Atlantoaxial Rotary Subluxation in Marfan Syndrome

John Ong Ying Wei[1], Tin Chan[1], William Lo[1], Buddhika Wimalachandra[1]

Abstract

Atlantoaxial rotary subluxation (AARS) is a rare condition that may cause persistent torticollis if not treated appropriately. AARS is associated with ligamentous abnormalities, which may result from acquired or congenital disorders. We report the case of a paediatric patient with congenital Marfan syndrome and AARS due to a minor traumatic head injury. A 9-year-old boy with a known diagnosis of Marfan syndrome (and extensive family history) encountered a traumatic head injury that presented as torticollis with a typical "cock-robin" head and neck orientation. AARS was diagnosed through a head and neck CT scan. He underwent initial conservative treatment involving a muscle relaxant (diazepam) and Miami-J collar. This was followed up with manipulation under anaesthesia (MUA) and further cervical traction, which resolved the subluxation without more invasive treatment. To the best of our knowledge, AARS associated with Marfan syndrome has been rarely reported in literature. It is postulated that the ligamentous laxity associated with Marfan syndrome would increase the patient's predisposition to AARS and more importantly, the propensity to require more invasive treatment (internal fixation). However, our patient unexpectedly responded well to conservative management, namely MUA and cervical traction. This illustrates that despite the increased ligamentous laxity in Marfan syndrome, it is still advisable to conservatively manage AARS before deciding to perform invasive internal fixation.

Keywords: Atlantoaxial rotary subluxation (AARS), persistent torticollis, congenital Marfan syndrome

DOI: http://dx.doi.org/10.31344/ijhhs.v5i0-2.333

1. Faculty of Medicine, University of Birmingham