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ABSTRACT

This case report describes a patient who presented with cervical spinal pain and headaches associated with atlanto-axial subluxation (AAS) secondary to rheumatoid arthritis (RA). For physiotherapists, especially less experienced clinicians, the significant risks associated with using manual assessment and treatment techniques in such a patient require careful consideration right at the start of a consultation. The focus of the case is therefore on the recognition of AAS in this patient with RA, highlighting the clinical findings that alert clinicians to this possibility and explaining the requisite knowledge and skills required to safely and effectively manage this patient. The use of screening tools to help clinicians identify possible RA in its pre-diagnosis stage and the clinical signs and symptoms that raise the index of suspicion for AAS, are discussed. The relevant contraindications and precautions associated with manual treatments directed at the upper cervical spine, and which may have potentially serious negative consequences, including quadriplegia and mortality, are addressed. Finally, the implications for the use of manual assessment and treatment of patients with RA and co-morbid AAS are addressed.

Key words: rheumatoid arthritis, systems screening, red flags, risk benefit analysis, triage, cervical spine instability

Word count: (2978)

INTRODUCTION

For physiotherapists using manual treatments in the assessment and management of patients with rheumatoid arthritis (RA), awareness and identification of potentially serious articular and peri-articular manifestations of the disease, including instability of the cervical spine, is essential. While there is a need for vigilance with respect to extra-articular manifestations of RA (see accompanying Professional Issue by Briggs et al., 2013), this paper focuses on the upper cervical spine. The most frequently occuring instability in the cervical spine is anterior atlantoaxial subluxation (AAS) (Wasserman et al., 2011; Yurube et al., 2012), where progressive loss of the primary and secondary ligamentous integrity combined with bony erosion of the odontoid process, associated with systemic inflammation as part of the RA disease process, can result in dire consequences, including quadriplegia or death (Paus et al., 2008; Wasserman et al., 2011). A high index of suspicion for AAS in patients with RA should alert clinicians to the potential risks associated with manual assessment and treatment and help ensure safe and effective patient care. The following case report on a patient with RA and associated AAS takes a clinical practice focus, highlighting the importance of the requisite clinical knowledge, reasoning and skills required to guide appropriate assessment and management.

CASE REPORT

Clinical History

A 55 year-old female with a 35-year history of seropositive RA was referred to physiotherapy for assessment and management of persistent, bilateral neck pain and headaches (Figure 1).

INSERT Figure 1 here

The neck pain radiated bilaterally from the suboccipital area to occipital and parietal areas, with occasional shooting pain to both temples. She described hearing “clanking” and "crunching" sounds in her neck, mainly on neck flexion or extension. The pain had been present for five years, with recurrent episodes of increased neck pain associated with increased bilateral suboccipital/occipital and parietal headaches. The neck pain and headaches had noticeably worsened in the past two years and coincided with a change in her occupational duties, which involved increased computer work requiring more sustained postural demands and more frequent and repeated flexion/extension movements of the head and neck. Pain was rated as moderately severe (average VAS 4-6 over 24 hours) and irritability varied with workload, from moderate to low. Considered over a 24-hour period, her neck pain and headaches worsened in the afternoons and improved in the mornings and on non-working days, consistent with sustained postures associated with computer work. She had not had any prior physiotherapy for her neck pain and headaches, relying primarily on simple analgesia and regular exercise (walking and tai chi).

She denied experiencing any vertebrobasilar insufficiency (VBI) symptoms, or dysaesthesiae of her lips or tongue, although she reported transient paraesthesia of her left foot and left distal arm, which were not behaviourally linked to her neck pain and headaches and usually resolved quickly, once she had adjusted her head and neck posture. There was no gait disturbance, upper or lower limb weakness, or change in bowel or bladder function to suggest cauda equina syndrome. Her RA was well controlled with a combination of disease-modifying anti-rheumatic drugs (DMARDS). Her medical history is summarised in Table 1.

Based on her description of the neck “clanking”, and prior to her physiotherapy consultation, she had been referred by her rheumatologist for plain radiographs and magnetic resonance imaging (MRI) of the cervical spine. Plain radiographs revealed erosion of the odontoid peg,

and 5mm anterior subluxation of C1 on C2 on flexion (Figure 2a), which reduced to normal (≤3mm) with cervical extension (Figure 2b). MRI of the cervical spine revealed advanced arthropathy at the articulation between the lateral masses of C1 and C2 on the left, associated with marrow oedema. There was no evidence of cervical cord compression or an intrinsic spinal cord signal abnormality.

INSERT Figure 2 here

INSERT Table 1 here

Physical examination

Examination of the cervical spine revealed a loss of the normal cervical lordosis. Active cervical ranges were limited globally and associated with crepitus through range and end range pain: extension to approximately 10 degrees with restriction throughout the entire cervical spine; flexion was limited to 30 degrees, occurred primarily in the upper cervical spine and was associated with an audible 'clunk' that was reproducible and not associated with any transient cord symptoms or signs; lateral flexion and rotation were restricted bilaterally to approximately 30 degrees. At rest, paraspinal cervical muscle overactivity was evident bilaterally, primarily in the suboccipital region and the sternocleidomastoid muscles. Examination of the hands revealed no evidence of active synovitis, with typical RA-type joint deformities involving the wrists, metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints, ankles and metatarsophalangeal joints. There was mild swelling and tenderness in bilateral 1st carpo-metacarpophalangeal joints.

Neurological examination of both upper and lower limbs revealed normal power (5/5; grade 1 on the Ranawat classification (Ranawat et al., 1979)). With the exception of absent ankle jerks bilaterally, reflexes were normal. Sensory testing indicated no loss or gain of sensitivity to light touch, pressure and thermal stimuli in upper and lower limbs. Babinski was down-going and Hoffman's sign was absent, plus there was no evidence of clonus.

Treatment

Physiotherapy treatment was conservative and comprised upper cervical isometric stabilisation exercises, soft tissues stretches to pain onset, advice on use of active range of motion rotational and side flexion exercises, and information about symptoms that would prompt the patient to seek further assessment. Given the evidence for RA-associated AAS and subaxial cervical subluxation in this case, the use of manual techniques was considered inappropriate as these can further progress the AAS with potentially serious negative consequences (see discussion). Treatment resulted in significant improvement of her neck pain, although the neck pain and headaches still persisted. Appropriate ergonomic adjustments were recommended for her workstation, along with regular change of position and avoidance of sustained postures.

Discussion

Safely and effectively managing patients with a systemic disease like RA, especially for less experienced clinicians, is challenging (Fary et al., 2012) . The challenge for clinicians includes both knowledge (the ‘know’) and skills (the ‘do’) (Briggs et al., 2012). The ‘know’ includes knowledge of RA as a clinically significant disease with associated co-morbidities; red flag issues such as AAS; what to assess, why, when and how to interpret the clinical findings. ‘Know’ also includes the clinical indicators for when and how to facilitate timely access to

specialist care. The ‘do’ includes clinical skills to enable early diagnosis of RA and screening for risk factors such as AAS; performing a standardised neurological examination; appropriate use of screening tools; and safe assessment and management procedures.

The patient history, associated rheumatology referral and radiological images in this case provided the clinician with timely, accurate information about RA-associated AAS, enabling sound clinical decision-making and safe care. Clinicians need to be aware that involvement of the cervical spine can occur early in RA (Paimela et al., 1997), and is common. The RA-associated incidence of laxity, instability and subluxation of the AAS ranges from 17-85% (Bouchaud-Chabot and Liote, 2002; Wolfs et al., 2009) with AAS being the most frequent deformity (Wasserman et al., 2011), occurring in up to 50% of cases (Mukerji and Todd, 2011). The clinical presentation can be variable (including minor pain with loss of function, significant pain with loss of function, or loss of function with minimal pain), with symptoms commonly associated with neck pain or compressive myeloradiculopathy (Mukerji and Todd, 2011), highlighting the need for a thorough medical history and appropriate screening for red flags.

In this case, the clinical presentation suggested a significant nociceptively-mediated contribution to her cervical pain and headaches, with the RA-associated inflammatory component well controlled. If her cervical pain and related headache tended to occur in parallel with peripheral joint disease flares, or if the symptoms fluctuated in a similar pattern to the peripheral joint disease, consistent with nociceptive inflammatory pain, then management with DMARDs would be required to address disease activity as a priority. As her widespread peripheral joint problems were well controlled, it is likely that the mechanical factors associated with the AAS and subaxial cervical subluxation were significant contributors to her symptoms. Evidence consistent with this interpretation includes cervical symptoms that

were dominantly mechanically-patterned rather than inflammatory: better in the morning without prolonged pain and stiffness; no sleep disturbance; and cervical pain and headaches worsened with sustained postural load and computer work (and evidence for some symptomatic relief associated with avoiding sustained load and workstation adjustments), worse at the end of day and during the working week. Further, physical examination findings are consistent with a dominant mechanical contribution to her cervical pain and headache, as demonstrated by stimulus-response movement-related pain behaviours and limited soft tissue sensitivity and an absence of any neurological compromise.

As the body chart was mapped to reflect the effective management of the RA condition (using a treat-to-target approach), her additional disease-related widespread pain areas and secondary osteoarthritis were not charted. While appropriate in this instance (based on the preceding rationale), one advantage of mapping the additional pain areas on the body chart, regardless of disease-activity, would be to facilitate clinical pattern recognition (Jones and Edwards, 2008), thereby highlighting the need to expand questioning and screen for systemic diseases such as RA. In this regard, current best practice guidelines (Royal Australian College of General Practitioners, 2009; Royal College of Physicians, 2009) recommend timely on-referral within 2 weeks of consultation if patients present with any of the following:

- the small joints of the hands or feet are affected;
- more than one joint is affected;
- there has been a delay of 3 months or longer between onset of symptoms and seeking medical advice.

This approach would be particularly important if RA is not yet diagnosed, and when the clinician is working in a primary care setting, acting as a first-contact practitioner and is inexperienced.

Clinical practice point: clinical pattern recognition and use of screening tools

Mapping all the pain areas can help to facilitate the visual recognition of potential systemic condition and expand the clinical reasoning process to screen for systemic diseases and motivate on-referral. Use of standardised screening tools may assist clinicians identify early (pre-diagnosis) RA in primary care. The gait, arms, legs and spine (GALS) locomotor screening examination for RA (recently tested for use among physiotherapists) has high specificity, suggesting utility as a physical screening test in primary care settings (Beattie et al., 2011). A self-administered early inflammatory arthritis detection (EIA-3 Detection tool), developed for use in primary care (Bell et al., 2010), may also be helpful. This history-based tool consists of 11 questions with Yes/No responses, covering dimensions of pain, stiffness and swelling.

This case demonstrated both AAS and subaxial cervical subluxation, consistent with epidemiologic data indicating that the three most common presentations of cervical spine involvement in RA-associated instability include AAS (65%), basilar invagination (20%) and subaxial cervical subluxation (15%) (Wasserman et al., 2011). The “clanking” sound described by the patient in this case, is a classic sign of instability and should have raised the index of suspicion, particularly since a large proportion of cervical instabilities can be otherwise asymptomatic (Collins et al., 1991; Neva et al., 2006). Plain radiolographic images confirmed AAS, demonstrating a 4-5mm anterior atlanto-dens interval (AADI) and subaxial cervical subluxation. The AADI is the distance from the posterior margin of the anterior ring of C1 to the anterior surface of the odontoid peg (Mukerji and Todd, 2011). The AADI typically increases with progressive ligamentous laxity of primary (transverse ligament) and secondary (alar ligaments) atlanto-axial restraints, with an anterior subluxation of greater than 10-12mm implying destruction of all ligamentous restraints. In this case, the 4-5mm implies secondary

193 restraints are intact. However, the AADI does not correlate well with the risk of developing a
194 neurological deficit or with the extent of any neurological deficit because patients have
195 variable spinal canal diameters. Thus, the effect of a given degree of slip in a patient with a
196 wide canal will be less than that in a patient whose canal is congenitally narrow. The posterior
197 atlanto-dens interval (PADI) is considered a better method because the PADI directly measures
198 the spinal canal and can better indicate the degree of canal narrowing associated with AAS.
199 The PADI is the distance between the posterior surface of the odontoid peg and the anterior
200 margin of the posterior ring of the atlas (the normal spinal canal measures 17-29 mm at C1)
201 and a minimum PADI of 14 mm is required to avoid cord compression (Boden et al., 1993). The
202 sagittal diameter of the subaxial cervical spinal canal also better correlates with the presence
203 and/or extent of myelopathy and patients with subaxial cervical canal diameters of 13 mm or
204 less are at increased risk of myelopathy (Boden et al., 1993). In this case, while AAS and
205 subaxial cervical subluxation were present, there was no evidence of significant cervical cord
206 compression, suggesting ample canal space.
207
208 While not evident in this case, reduced ROM has been described in RA-associated instabilities
209 as alternating with increased ROM, termed ‘pseudostabilisation’ by (Wasserman et al., 2011).
210 This ROM variability can be an indication of basilar invagination and is reportedly present in
211 40% of people with RA with the dens entering the foramen magnum and thereby reducing
212 available ROM (Boden, 1994; Boden et al., 1993). Basilar invagination (also termed ‘superior
213 migration of the odontoid’, ‘cranial settling’ or ‘vertical subluxation’), involves the axis
214 telescoping into the atlas, driving the odontoid peg upwards and this can cause brainstem
215 compression, producing facial sensory disturbance, dysphagia, or abnormalities in the lower
216 cranial nerves (Murkeji and Todd 2011). Suboccipital pain (as present in this case and typically

a consequence of C2 nerve root involvement (Heywood et al., 1988), is commonly associated with AAS and occasionally subaxial cervical subluxation and basilar invagination.

Clinical practice point: importance of neurological examination at baseline

If upper cervical instability is suspected, undertaking a comprehensive neurological examination at the first consultation is recommended. This should include upper and lower limb reflexes, Hoffman, Babinski, clonus, motor and sensory function and checking gait for imbalance. Clinical signs of a myelopathy include hyperreflexia, upgoing plantar (Babinski) responses, positive Hoffman's signs or clonus, together with motor and sensory deficits. Neurologic impairment can be classified using systems such as the Ranawat classes I-IIIB (Ranawat et al., 1979), to establish the degree of neurologic compromise or Steinbocker's grades I-IV to classify functional limitation (Steinbrocker and Blazer, 1946). If still in doubt, refer on for a medical review, as patients may be asymptomatic even in the presence of upper cervical spine instability.

In the absence of radiological evidence or a specific diagnosis of AAS from the rheumatology referral, and based on the body chart and behaviour of the cervical pain and headache alone, the patient's symptoms might have been interpreted as cervicogenic in origin, thereby missing AAS and the subaxial cervical subluxation. In that instance, assessment and treatment directed at the upper cervical spine would be associated with clear risk as the performance of manual techniques may further progress the AAS with potentially serious negative consequences, including quadriplegia and mortality. In this regard, the use of craniovertebral instability (CVI) tests requires consideration, although currently there are no related guidelines or diagnostic criteria. Further, although the reliability of CVI (anterior shear and tectorial membrane) screening tests has been reported in normal volunteers as moderate to substantial

(Osmotherly et al., 2012), according to a recent survey of Australian physiotherapists (Osmotherly and Rivett, 2011) their use in clinical practice appears to be inconsistent. In the absence of any clear guidance, the default position relies on clinicians having sound knowledge and applying sound clinical reasoning to analyse the risk/benefit associated with these CVI tests. In this clinical case, a common-sense approach based on the medical ethical precept '*primum non nocere'*, would be not to perform CVI tests: the potential risk to the patient far outweighed any clinical benefit of establishing evidence for AAS. Furthermore, in the absence of any prior medical review or radiology, it is advisable to request a medical review and raise an index of suspicion for a CVI prior to undertaking any manual assessment or treatment of the upper cervical spine. A mandatory review of radiological images prior to manual assessment or treatment is appropriate subsequent to a medical review. If, as in this case, there is evidence of osteoporosis or a suspicion of bone fragility, the use of strong manual treatments requires further consideration.

SUMMARY

Clinical guidelines provide a mechanism by which physiotherapists can ensure best evidence practice in assessing and managing patients with RA. However, knowledge must sit alongside advanced clinical skills and sound clinical reasoning to ensure safe and effective care. For first contact practitioners in particular, a high index of suspicion regarding AAS should assist in the early recognition of a potential red flag pathology that contraindicates manual treatment and requires immediate on-referral and appropriate investigation. The use of a systems approach (triage and screening; appropriate on-referral), can facilitate the effective implementation of an evidence-informed and safe approach to the assessment and management of patients with RA.

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Figure legends

Figure 1. The location of the patient's reported neck pain and headaches are shown in this body chart. Note that the patient's widespread rheumatoid arthritis-associated joint involvement and osteoarthritis are not mapped on this chart, indicating an effective treat-to-target approach, consistent with the current best practice for management of rheumatoid arthritis.

Figure 2. The plain radiographs of the cervical spine reveal : (a) a 5mm of anterior subluxation (arrowed) of C1 on C2, reflected in an increased anterior atlanto-dens interval (AADI) on flexion (Figure 2a); which is reduced with cervical extension (Figure 2b). There is also multilevel cervical spondylosis with mild instability between flexion and extension at C2/3, as well as further variable grade 1 spondylolistheses between C3/4 and C5/6 (not annotated).

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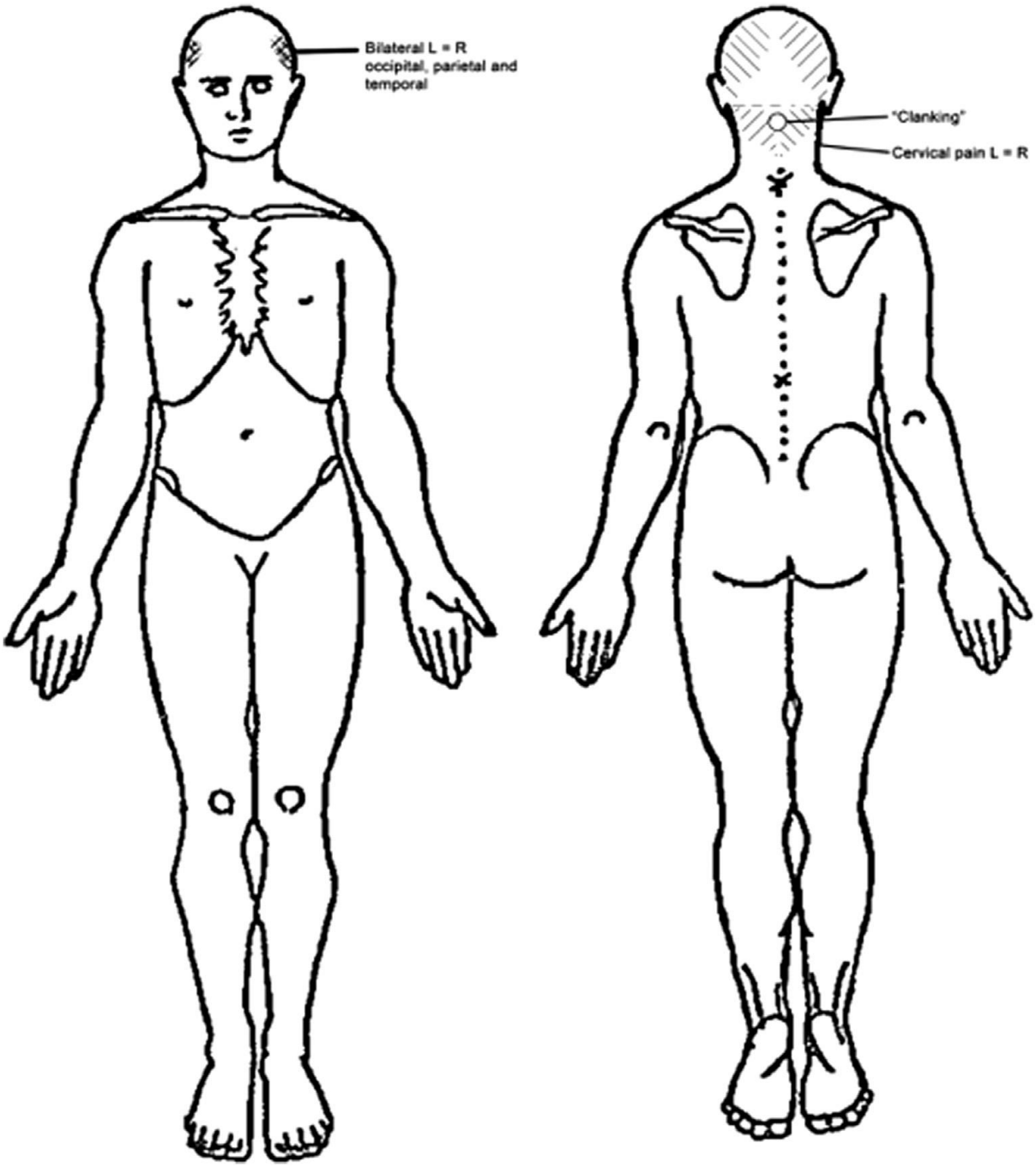
Bilateral L = R
occipital, parietal and
temporal
"Clanking"
Cervical pain L = R

Fig. 1

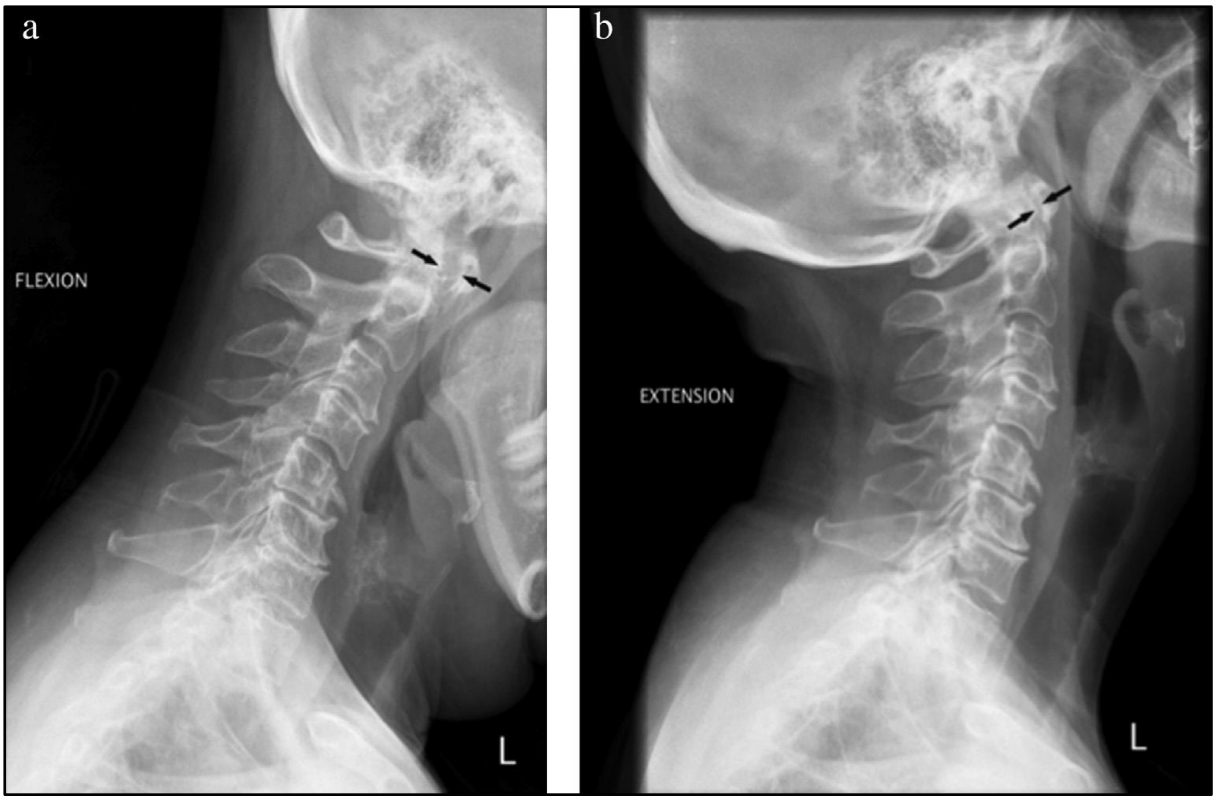

Fig. 2