Case report

Unusual scapular winging — A case report

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ARTICLE INFO

Article history:
Received 27 September 2015
Received in revised form
6 December 2015
Accepted 13 December 2015

Keywords:
Scapular winging
Weakness
Facioscapulohumeral dystrophy
Physiotherapy

ABSTRACT

Scapular mobility has a central role in maintaining normal upper limb function. Scapular winging is characterized by a failure in the dynamic stabilization of the scapula against the thoracic wall resulting in a condition in which the medial border of the scapula is prominent.

The following case describes a patient who was referred to physiotherapy due to abnormal scapular protrusion. The main findings of the physical examination showed weakness of the scapular stabilizers more prominent on the right side than of the left. Additionally, the physical examination demonstrated weakness of the abdominal muscles, hip adductors, and ankle dorsi-flexors, as well as some facial muscles. The electromyography results were inconclusive. Further examination led to clinical suspicion of Facioscapulohumeral Dystrophy (FSHD) as a diagnosis, which was confirmed by genetic testing.

Facioscapulohumeral Dystrophy is characterized by symptoms related to motor function and in most cases becomes evident in patients in their 20s and 30s. The disease signs and symptoms are often identified in a clinical setting. Currently, there are no reports describing an effective treatment for the disease. However, physiotherapy, moderate physical exercise, counselling, and use of suitable aids and orthoses may help improve functionality and mobility.

This case report aims to increase the awareness of musculoskeletal physiotherapists to this unique dystrophy, when encountering complex presentations with scapular winging.

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1. Introduction

Scapular winging is often a cause for pain, disability and functional limitation in the upper extremities (Zatz et al., 1998). Combined stabilization and scapular mobility is vital for normal shoulder function. When scapular stabilization is impaired, shoulder mobility may be limited to 90° abduction or less, and this impairment may impact the ability to perform loaded motion, such as lifting heavy objects as well as overhead activities of daily living (ADL) such as dressing and hair combing (Kibler, 1998). Scapular winging may also be distressing for esthetic reasons (Kauppila and Vastama ki, 1996; Martin and Fish, 2008).

Scapular winging is characterized by a failure in the dynamic stabilization of the scapula against the thoracic wall resulting in a condition where the medial border of the scapula is prominent. Appropriate functional control of the scapula during shoulder elevation involves coupled activation of upper and lower trapezius, along with the serratus anterior and rhomboids. Both the upper and lower trapezius together rotate the scapula upwards, while the serratus anterior, a scapula protractor, stabilizes it to the chest wall and assists in upward rotation, and the rhomboids maintain traction of the medial border (Kibler, 1998).

Injury to either the nerves supplying these muscles, the muscles themselves, or their attachments to the scapula may lead to winging (Gooding et al., 2013). However, in the literature the most commonly described conditions leading to winging are serratus anterior palsy or long thoracic nerve injury (Gooding et al., 2013). Less common scapular winging presentations include brachial plexus injury, structural anomalies of the scapula including osteochondroma, fractures, neuropathies, or dystrophies (Fiddian and King, 1984; Kuhn et al., 1995).

The following case describes a patient who was referred to physiotherapy due to observed scapular winging. However, the findings of the assessments revealed an uncommon presentation.

2. Patient presentation

A 29-year-old male was referred to physiotherapy evaluation by a Pilates instructor who identified abnormal scapular position,
unusual difficulty in shoulder elevation, and restricted trunk mobility.

2.1. Subjective examination

Pain was not a predominant complaint. The patient’s aim was to improve posture for esthetic reasons in view of the family history of a very kyphotic grandfather. Mild functional disability (e.g. performing overhead activities, transition from supine to sitting position) was associated with reduced endurance and strength of the shoulders and abdominal muscles, but the patient presumed it was within normal range. Although pain free at rest, lower back discomfort (intensity 4/10) was reported. The discomfort was aggravated by activities such as playing basketball or standing still for over 10 min.

Moderate diffuse low back pain (LBP) started when the patient was 18-19 years old with no previous abnormality. A bone scan and x-ray were reported as normal. At age 20 he received the first physiotherapy treatments for his LBP. Four years later when attending physiotherapy for LBP, abnormal scapular mobility was noted but treatment focused on his posture. Additionally, he participated in personal training sessions for two years aimed at improving general fitness, endurance, and strength.

2.2. Physical examination

The patient was tall and lean. The abdomen was protruding and there was visible evidence of bilateral atrophy of the pectoralis major and biceps brachii. Bilateral scapular winging (R>L) was also noted along with shoulder height asymmetry (lower on the right dominant side). Fig. 1 presents static posture as observed in the physical examination.

Full passive ROM was found in all upper and lower limbs and in the spine. Active shoulder elevation produced scapular winging bilaterally (R>L).

Neurological examination was overall negative with normal muscle tone, stretch reflexes and sensation in all body parts. Table 1 presents the results of manual muscle testing of the shoulder girdle, trunk and lower limb. No abnormalities (5/5 MMT) were found around the elbow, wrist, hand, knee, and toe joints. The same testing revealed weakness of the hip adductors (3/5), ankle dorsi flexors (3+/5) (functionally, he was unable to walk on his heels), and most significant weakness in the scapular and abdominal muscles.

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Table 1
Manual muscle testing results in physical examination.

<table>
<thead>
<tr>
<th>Resisted Movement</th>
<th>Primary muscle tested</th>
<th>Left</th>
<th>Right</th>
</tr>
</thead>
<tbody>
<tr>
<td>Upper limb</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Scapular elevation</td>
<td>Upper trapezius</td>
<td>5/5</td>
<td>5/5</td>
</tr>
<tr>
<td>Scapular adduction</td>
<td>Rhomboids</td>
<td>4/5</td>
<td>4/5</td>
</tr>
<tr>
<td>Scapular abduction and upward rotation</td>
<td>Serratus anterior</td>
<td>3/5</td>
<td>1/5</td>
</tr>
<tr>
<td>Shoulder extension</td>
<td>Latissimus dorsi, Teres major, Posterior deltoid</td>
<td>4/5</td>
<td>4/5</td>
</tr>
<tr>
<td>Shoulder flexion</td>
<td>Anterior Deltoid, Supraspinatus, Coracobrachialis</td>
<td>4/5</td>
<td>2/5</td>
</tr>
<tr>
<td>Shoulder horizontal adduction</td>
<td>Pectoralis major</td>
<td>4/5</td>
<td>4/5</td>
</tr>
<tr>
<td>Shoulder abduction</td>
<td>Middle Deltoid, Supraspinatus</td>
<td>4/5</td>
<td>2/5</td>
</tr>
<tr>
<td>Shoulder external rotation</td>
<td>Infraspinatus, Teres minor</td>
<td>5/5</td>
<td>5/5</td>
</tr>
<tr>
<td>Shoulder internal rotation</td>
<td>Subscapularis</td>
<td>4/5</td>
<td>4/5</td>
</tr>
<tr>
<td>Elbows, wrists &amp; fingers</td>
<td></td>
<td>5/5</td>
<td>5/5</td>
</tr>
<tr>
<td>Trunk</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Trunk flexion</td>
<td>Rectus abdominis, internal/external obliques</td>
<td>2/5</td>
<td></td>
</tr>
<tr>
<td>Hip abduction</td>
<td>Adductor magnus/longus/brevis, pectineus, gracilis</td>
<td>3/5</td>
<td>3/5</td>
</tr>
<tr>
<td>Hip Flexion/Extension/ABD/IR/ER</td>
<td></td>
<td>5/5</td>
<td>5/5</td>
</tr>
<tr>
<td>Knee Extension/Flexion</td>
<td></td>
<td>5/5</td>
<td>5/5</td>
</tr>
<tr>
<td>Ankle dorsiflex</td>
<td>Tibialis anterior</td>
<td>3+/5</td>
<td>3+/5</td>
</tr>
<tr>
<td>Ankle Plantar-flexion/Eversion/Inversion, toes</td>
<td></td>
<td>5/5</td>
<td>5/5</td>
</tr>
</tbody>
</table>

The bold in table is to emphasize the main weak muscle groups.

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Fig. 1. Observation from the front, back and side: Body form was tall and lean, postural changes include a protruding abdomen, bilateral atrophy of pectoralis major and biceps brachii, bilateral scapular winging (R>L), and asymmetrical shoulder’s height (lower on the right – dominant side).

Fig. 2. Shoulder girdle muscle atrophy and wasting: Notice the ‘Trapezius Hump’ on the right-the upper scapular border is protruding upwards through the upper trapezius muscle, and is palpable as a bony structure where usually the upper trapezius muscle can be palpated.
2.3. Clinical reasoning

The muscle weakness pattern did not resemble a localized muscle or nerve injury. There were no history or symptoms of a cervical spinal cord injury nor of central nervous system origin. In light of the unrelated sites involved the authors performed a literature search for a relevant diagnosis using the aforementioned physical findings of weakness and atrophy. The results included two relevant diagnoses: Limb Girdle Muscular Dystrophy (LGMD), and Facioscapulohumeral Dystrophy (FSHD). Both dystrophies described high Creatine Phosphokinase (CPK) levels as a possible physiological characteristic, and abnormal electromyography signals. Accordingly, the patient was referred for these tests. CPK levels were found to be within normal range. The EMG report concluded there was no evidence for peripheral neuropathy nor typical myopathy, but lower than normal amplitudes were reported in the thoracodorsal and long thoracic nerves possibly due to muscle atrophy. In addition, the EMG findings were inconclusive to support congenital myopathy, and the neurologist suggested further investigation with a muscle biopsy. Further neurological examination revealed bilateral Orbicularis Oculi weakness, and mild weakness of Orbicularis Oris. The weakness of these facial muscles supported the diagnosis of FSHD.

3. Discussion

Muscular dystrophies are inherited myogenic disorders characterised by progressive muscle wasting and weakness of variable distribution and severity (Emery, 2002). Facioscapulohumeral dystrophy (FSHD) was first described in 1884, (Landouzy and Dejerine, 1884) and is a relatively slowly progressing disease and is inherited in an autosomal dominant pattern (Richards et al., 2012). The reported prevalence of the disease is 1:15,000 (Flanigan et al., 2001) to 1:20,000 (Padberg, 2004), and its clinical presentation may vary considerably, from asymptomatic individuals to patients who are wheelchair-bound (20% of cases) (Tupler and Gabellini, 2004).

3.1. Clinical presentation

The age at clinical onset may also vary. Most patients become symptomatic in their 20’s and in about 95% of the cases the first symptoms may be identified by the age of 20 (Padberg, 2009).
FSHD is characterized by muscle weakness that spreads throughout the body usually starting with the facial muscles (Oribicularis oculi, Orbicularis oris), followed by the scapular muscles (Figs. 2–4), Deltoid and Biceps brachii muscles weakness, often asymmetrical (Fig. 6), and trunk weakness, characterised by abdominal bulging, lumbar hyper lordosis and the positive Beevor’s sign discussed below (Figs. 1, 4 and 5).

Lower extremities weakness is often distal, typically in the anterior leg compartment causing a foot-drop (Tawil and Van Der Maarel, 2006). It often takes a long time between the appearance of the initial symptoms and the diagnosis due to the slow progression of the disease.

The classic clinical sign typical of FSHD is the Beevor’s sign (Awerbuch et al., 1990; Shahrizaila and Wills, 2005; Eger et al., 2010). Beevor’s sign is described as an abnormal upward movement of the umbilicus when the assessed patient is in a supine position attempting to raise his head and neck towards his chest (Fig. 5). This demonstrates the lower abdominals being unable to provide satisfactory stabilization with contraction of the upper abdominal muscles. Beevor’s sign has shown 90%–95% sensitivity and 93%–100% specificity to differentiating FSHD from other neuromuscular diseases. Beevor’s sign can also be seen in patients suffering from a spinal cord injury at T10 level. It has also been reported in some cases of Amyotrophic lateral sclerosis and other myopathies (Awerbuch et al., 1990; Shahrizaila and Wills, 2005).

In addition, Pradhan (2002) introduced the “Poly-hill Sign” which identifies a typical topographic anatomic finding of multiple bulges and indentations in the shoulder area (Fig. 6) (Pradhan, 2002). The Poly-hill Sign is observed in the upright position, with patient’s arms raised to 90° shoulder abduction and external rotation. The Sign is considered positive when the following characteristics are observed: (1) indentation due to Trapezius atrophy; (2) bulge formed by superior angle of the scapula; (3) indentation due to Trapezius atrophy; (4) acromio-clavicular joint (ACJ) bulge; (5) indentation due to atrophy of the upper quarter of the Deltoid adjacent to the ACJ; (6) bulge due to slight enlargement (at the early stages) or relative preservation (at a later stage) of three quarters of distal Deltoids; (7) indentation due to Biceps atrophy; (8) bulge caused by Brachioradialis or Extensor Digitorum Communis hypertrophy. Additional bulges arise by the inferior angle of scapula, and by slight hypertrophy of the infraspinatus (Fig. 6) (Pradhan, 2002).

The Poly-Hill Sign was found in 17/18 FSHD patients (aged 16–24), which was the highest rate of appearance compared to other dystrophies. Its high diagnostic value arises also from its presence in FSHD cases with no facial weakness or more severe lower limb weakness (Pradhan, 2002).

Other clinical symptoms may include high-pitch hearing loss (in about 75% of cases), or retinal telangiectasia (60% of cases) (Padberg et al., 1995). In rare cases, FSHD patients may suffer from respiratory muscle weakness (Wohlgemuth et al., 2004), myocardial conduction defects (Laforet et al., 1998), cardiac arrhythmia (Trevisan et al., 2006) and central nervous system disorders associated with the severe form of the disease (Saito et al., 2007).

FSHD is an autosomal-dominant genetic disease. The genetic mutation is typically familial and found in chromosome 4. Specifically, it is a polymorphic repeat array (D4Z4) on the distal end of the long arm (Tawil and Van Der Maarel, 2006).

The prognosis of FSHD is variable, and often is less severe in women (Zatz et al., 1998). Life expectancy may be unaffected, to variable levels of disability, with about 20% of patients who require a wheelchair as a mobility aid (Padberg, 2009). A sub-population of patients with infantile onset of the disease may develop severe disabilities already at a young age.
### 3.2. Physiotherapy and other treatments

Unfortunately, no effective therapeutic strategies have been reported in the literature to either slow down or stop the progression of muscular weakness and atrophy (Rose and Tawil, 2004). Nonetheless, a number of therapeutic interventions can provide symptomatic and functional improvement in quality of life for many patients. Orthoses should be considered for advancing walking, mainly due to dorsi-flexors weakness, and therefore a fixed or hinged ankle foot orthoses for foot drop may be useful. Abdominal supports/braces and binders are often recommended by many clinicians claiming it may reduce pressure on the lower back when engaging in physical activities (Pandya et al., 2008) and in selected patients may reduce back pain (King and Kissel, 2013). However, there is a lack of evidence in the literature to support the benefits attributed to these braces for patients with FSHD.

### 3.3. Physical training

In the past, the recommendations for patients diagnosed with muscular dystrophy were not to engage in physical training, due to concerns that muscle tension may lead to muscle weakness due to overuse (Fowler, 1983; Petrof, 1998). A Cochrane Review on muscle strengthening and aerobic training in patients diagnosed with muscular dystrophies, concluded that moderate intensity strength training in FSHD patients (as well as in Myotonic Dystrophy patients) does not seem to cause any harm, although no sufficient data was found to determine its effectiveness (Voet et al., 2010). These findings are also corroborated by an earlier review (Cup et al., 2007) that examined the benefits of different types of physical training and physiotherapeutic methods on various dystrophies and neuromuscular diseases. Therefore, there is a paucity of evidence to establish physical training is beneficial for muscle strength training (Cup et al., 2007, Voet et al., 2010). However, some indications show aerobic training may have positive benefits on the body functions, activities and participation. A recent review of various neuromuscular diseases was more hesitant in its conclusions and stated that based on the available research no recommendation can be provided regarding any specific type, duration, or intensity of physical training (Gianola et al., 2013).

Despite the inconclusive evidence, published guidelines for physical training in FSHD patients recommend the following: (Pandya et al., 2008).

- Maintain an active lifestyle for its physical and psychological benefits.
- Flexibility training including ROM and stretching exercises is recommended to avoid limited mobility associated with muscular weakness.
- Moderate intensity strengthening exercises may be beneficial for treating muscle weakness at the earlier stages of the disease, in muscles that maintained at least 3/5 of their strength.
- Moderate intensity aerobic exercise can contribute to maintaining or improving aerobic capacities.

The emphasis on moderate intensity training is due to the concern that exercises associated with eccentric muscle contraction or high intensity training may have a detrimental impact on the muscles due to weakness of the cell membranes. According to a Cochrane systematic review, the indications for surgery, optimal technique and possible complications are not entirely clear. However, case reports and case series suggest that operative interventions of scapular stabilization in patients with FSHD may produce significant benefits (Orrell et al., 2010). More recent studies seem to support Thoracoscopicar arthrodesis for improving shoulder function in patients with FSHD (Van Tongel et al., 2013; Cooney et al., 2014; Goel et al., 2014).

### 4. Conclusion

The last decade has seen significant contributions to the understanding of the genetics and clinical characteristics of FSHD. Although early diagnosis of the disease cannot prevent its progression, it is important for minimizing possible complications, relieving pain, and contributes to maintaining an active and functional lifestyle. Furthermore, it is important to advise these patients to undergo genetic testing, prior to planning family life (Lunt and Harper, 1991).

The physiotherapist may be a key clinician in the diagnosis and treatment of patients with FSHD. Physiotherapists are encouraged to refer these patients to extensive laboratory and genetic testing and to provide therapeutic guidance regarding proper training, exercises, and fitness maintenance while preventing fatigue, as well as recommending suitable aids and orthoses, when relevant.

The case presented here aims to acquaint the musculoskeletal physiotherapist with the characteristics of FSHD, which are totally different than orthopedic presentations with scapular winging. Correct and early identification of patients suffering from FSHD will prevent inappropriate management, and will advance their wellbeing and quality of life. Further research is needed to find effective therapeutic training for these patients.

### Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.math.2015.12.006.

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Please cite this article in press as: Dori Z, Sarig Bahat H. Unusual scapular winging — A case report, Manual Therapy (2015), http://dx.doi.org/10.1016/j.math.2015.12.006
Please cite this article in press as: Dori Z, Sarig Bahat H, Unusual scapular winging – A case report, Manual Therapy (2015), http://dx.doi.org/10.1016/j.math.2015.12.006