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Review

Focal myositis: A review

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Abstract

Amongst the heterogeneous group of inflammatory myopathies, focal myositis stands as a rare and benign dysimmune disease. Although it can be associated with root and/or nerve lesions, traumatic muscle lesions and autoimmune diseases, its triggering factors remain poorly understood. Defined as an isolated inflammatory pseudotumour usually restricted to one skeletal muscle, clinical presentation of focal myositis is that of a rapidly growing solitary mass within a single muscle, usually in the lower limbs. Electromyography shows spontaneous activity associated with a myopathic pattern. MRI reveals a contrast enhanced enlarged muscle appearing hyper-intense on FAT-SAT T2 weighted images. Adjacent structures are spared and there are no calcifications. Serum creatine kinase (CK) levels are usually moderately augmented and biological markers of systemic inflammation are absent in most cases. Pathological histological features include marked variation in fibre size, inflammatory infiltrates mostly composed of T CD4+ lymphocytes and macrophages, degenerating/regenerating fibres and interstitial fibrosis. Differential diagnoses are numerous and include myositis of other origin with focal onset. Steroid treatment should be reserved for patients who present with major pain, nerve lesions, associated autoimmune disease, or elevated C reactive protein or CK.

Keywords: Focal myositis; Inflammatory myopathy; Muscle biopsy; Intramuscular tumour

1. Introduction

Focal myositis (FM) is defined as a focal inflammatory pseudotumour usually restricted to one skeletal muscle whose prognosis is usually benign with self-regression in most cases [1–5]. Although focal inflammation of muscle has been reported since 1958 in patients with weakness of a single limb progressing to generalised polymyositis [6,7], the first cases of isolated FM were initially described by Heffner et al. in 1977 [1]. Since then, approximately 200 cases have been reported [1–5,8]. In 1993, Flaisler et al. defined focal myositis as a "myopathy affecting a single skeletal muscle without systemic manifestation with a histologically proven inflammatory myositis process" [9]. Its clinical presentation is that of a rapidly growing solitary mass within a single muscle, usually in the lower limbs. Pathological histological features are circumscribed within one muscle and include marked heterogeneity of fibre sizes including hypertrophic and regenerating fibres, inflammatory infiltrates mainly composed of macrophages and T cells, and fibrosis [4]. Because FM can mimic any disease presenting as a solitary inflammatory intramuscular mass lesion, differential diagnostics are multiple, amongst which nodular or granulomatous myositis and soft tissue tumours are the most challenging. The aetiology of FM remains unknown although some cases suggest the occurrence of triggering factors such as dysimmune diseases or denervation in the context of increased genetic susceptibility [8,10,11].

2. Clinical presentation

FM can occur at any age but preferentially during mid-adult age range (Table 1) [1–5,10]. Males are affected as well as females. FM usually presents as a circumscribed intramuscular mass or swelling within one specific muscle. However, six of the 16 cases initially reported by Heffner et al. presented involvement of more than one muscle [1]. Following this first description, the fact that FM can either involve part of a muscle, a whole muscle or multiple adjacent muscles was further

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Table 1
Demographic characteristic and clinical features in main FM series.

Authors	Heffner et al.	Smith et al.	Sekiguchi et al.	Auerbach et al.	Gaeta et al.
Year of publication	1977	2000	2004	2009	2009
Number of cases	16	8	4	115	8
Sex ratio	1M:1F	3M:1F	4M:0F	62M:53F	1M:1F
Mean age (years old)	39	40	37.5	41	44
Mean size (cm)	4,9	N/A	N/A	3,9	N/A
Pain	10/16	5/8	4/4	31/115	8/8
Systemic symptoms	0/16	1/8	N/A	2/115	N/A
Treatment	0/16	2/8	4/4	N/A	2/8
Recurrence	0/16	2/8	3/4	2/115	1/8

N/A: not available.

supported by recently published series of patients [3,5,8]. Therefore Gaeta et al. proposed a classification depending on the extent of muscle involvement: classes 1, 2 and 3 refer to FM involving a part of muscle, a whole muscle, and adjacent muscles respectively [5].

The mass is dense and firm, of variable size, and reported as painful in 14%–75% of patients [1,4,8,11]. In the largest series of 115 FM cases documented by muscle biopsy, the mean size was 3 cm, ranging from 1 to 20 cm, and pain was reported in 31 patients [27%] [4]. The mass grows insidiously for days to months and patients usually complain of swelling within one muscle eventually painful. Although typically affecting the calf or other muscles of the extremities, FM has been reported in many other unusual intramuscular sites such as the paraspinal muscles, proximal limb muscles, mentalis muscle, tongue, and orbicularis muscle [4,12–16].

There are usually no systemic symptoms (except fever which was encountered in 2 out of the 115 cases reported by Auerbach and co-authors) [1,3,4]. Neurological examination is normal and patients do not usually complain of any weakness or sensory impairment or arthritis.

3. Laboratory investigations

Laboratory studies including serum creatine kinase (CK), erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP) are often within normal ranges and search of known auto-antigens is negative (Table 2). However, elevated CK and ESR were encountered in 25% of cases reviewed by Yamnaz [2] and 50% of the cases reported by either Gaeta et al. or Lunde et al. [5,8]. In the large series of patients reported by Auerbach et al., the serum CK and ESR were normal but they were evaluated in less than 10% of patients [4]. Interestingly, it has been noted that patients with increased serum CK or elevated ESR were more likely to develop a diffuse inflammatory myopathy [1,9,17,18]. Such abnormalities should therefore be considered as atypical features.

4. Electrodiagnostic studies

Nerve conduction studies and electro-neuro-myography (ENMG) are useful to search for myogenic abnormalities and to determine whether the disease is focal or multifocal and/or is associated with nerve involvement that will change the treatment strategy. Little is known about electrodiagnostic data. Nerve conduction studies were normal in cases unrelated to a nerve lesion [3]. Profuse spontaneous activity with complex repetitive discharges were observed in three out of the eight patients reported by Smith et al. in the absence of nerve lesion [3], probably in relation to muscle fibre necrosis as classically described in other myogenic and necrotic processes. In most of the cases associated with S1-radiculopathy, spontaneous activity with fibrillation potentials and positive sharp waves related to denervation were reported [8,11,19]. Lastly, during contraction, a myopathic pattern is observed exclusively in the affected muscle with short-duration and small-amplitude polyphasic motor unit potentials [3].

5. Radiological features

5.1. MRI

MRI is one of the key diagnostic tools in the assessment of inflammatory myopathy (Fig. 1, Table 2) [3,5,10,20]. MRI typically reveals a circumscribed mass within a single muscle

Table 2
Investigation results in main FM series.

Authors	Heffner et al.	Smith et al.	Sekiguchi et al.	Auerbach et al.	Gaeta et al.
Year of publication	1977	2000	2004	2009	2009
Increased CK	0/16	3/8	1/4 (sub normal)	0/115	4/8
Inflammatory blood marker (ESR/CRP)	N/A	Normal or mildly elevated	Increase 4/4	Normal or mildly elevated	N/A
MRI enlargement	N/A	4/4	3/4	N/A	7/8
T1		−	N/A		6/8 iso, 2/8+
T1 gadolinium		Patchy	N/A		+
T2		+	+		+
STIR		N/A	N/A		+

CK: creatine kinase; CRP: C reactive protein; ESR: erythrocyte sedimentation rate; MRI: magnetic resonance imaging; N/A: not available; −: hypointense; iso: isointense; +: hyperintense.