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Original Article

Ultrasound and magnetic resonance imaging assessment of joint disease in symptomatic patients with cystic fibrosis arthropathy

G. Fitch [a], K. Williams [a], J.E. Freeston [b,d], S. Dass [b], A. Grainger [c,d], R. Hodgson [d], L. Horton [d], P. Whitaker [a], D. Peckham [a,*]

[a] Leeds Adult CF Unit, Leeds Teaching Hospitals NHS Trust, UK
[b] Department of Rheumatology, Leeds Teaching Hospitals NHS Trust, UK
[c] Department of Radiology, Leeds Teaching Hospitals NHS Trust, UK
[d] NIHR Leeds Musculoskeletal Biomedical Research Unit, Leeds Teaching Hospitals NHS Trust, UK

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Abstract

Objectives: Cystic fibrosis arthropathy (CFA) is a term commonly used for joint pain with and without swelling seen in some patients with CF. Early studies into CFA focused on the presence of rheumatoid factor and immunological changes on synovial biopsy, with parallels drawn between respiratory and joint activity. Identification of anti-cyclic citrullinated peptide antibodies (anti-CCP) as a marker of rheumatoid arthritis (RA), along with increased access to sensitive imaging techniques including ultrasound (US) and magnetic resonance imaging (MRI), offer great potential to investigate and more accurately understand the type(s) of inflammatory arthritis that may underlie CFA. The aim of this study was to phenotype an active CFA cohort using serology and imaging, as a basis for further work in this understudied area.
Methods: This was a prospective observational cohort study of symptomatic CFA patients presenting with joint pain. Participants underwent serological testing, clinical and US joint and entheseal assessment, as well as MRI of the most symptomatic joint/joint area.
Results: Ten symptomatic patients were studied with 9/10 having positive clinical findings. Inflammatory changes on US were seen in 8/10 cases. Five patients had positive findings on MRI (3 of whom had received IV gadolinium contrast). This included patients with significant erosive changes. One patient was anti-CCP positive suggestive of RA, and two were anti-nuclear antibody positive.
Conclusion: Imaging, and to a lesser extent serology, identified inflammatory joint pathology in a proportion of cases, providing important data to explore in a large CFA cohort examining the clinical and imaging phenotype of this group.

Keywords: Cystic fibrosis; Arthropathy; Ultrasound; MRI; Joint count

1. Introduction

Cystic fibrosis arthropathy (CFA) is the most common form of joint pain in patients with cystic fibrosis (CF) and can be associated with significant morbidity [1,2]. The clinical picture may vary from a minimally swollen joint to a polyarthritis with swollen, tender joints resembling those seen in rheumatoid arthritis (RA). Joint disease in patients with CF may also be associated with adverse drug reactions and gout [3]. CFA can follow a palindromic pattern with a remitting and relapsing course and symptoms mostly disappearing between attacks.

Cystic fibrosis arthropathy is usually described as a form of polyarthritis unique to CF that cannot be classified as any other rheumatological disorder [4]. However, there is no formalised definition or diagnostic criteria for this condition and little understanding of its aetiology, pathogenesis or sequelae. CFA can affect any joint and while symptoms may be self-limiting,

* Corresponding author.

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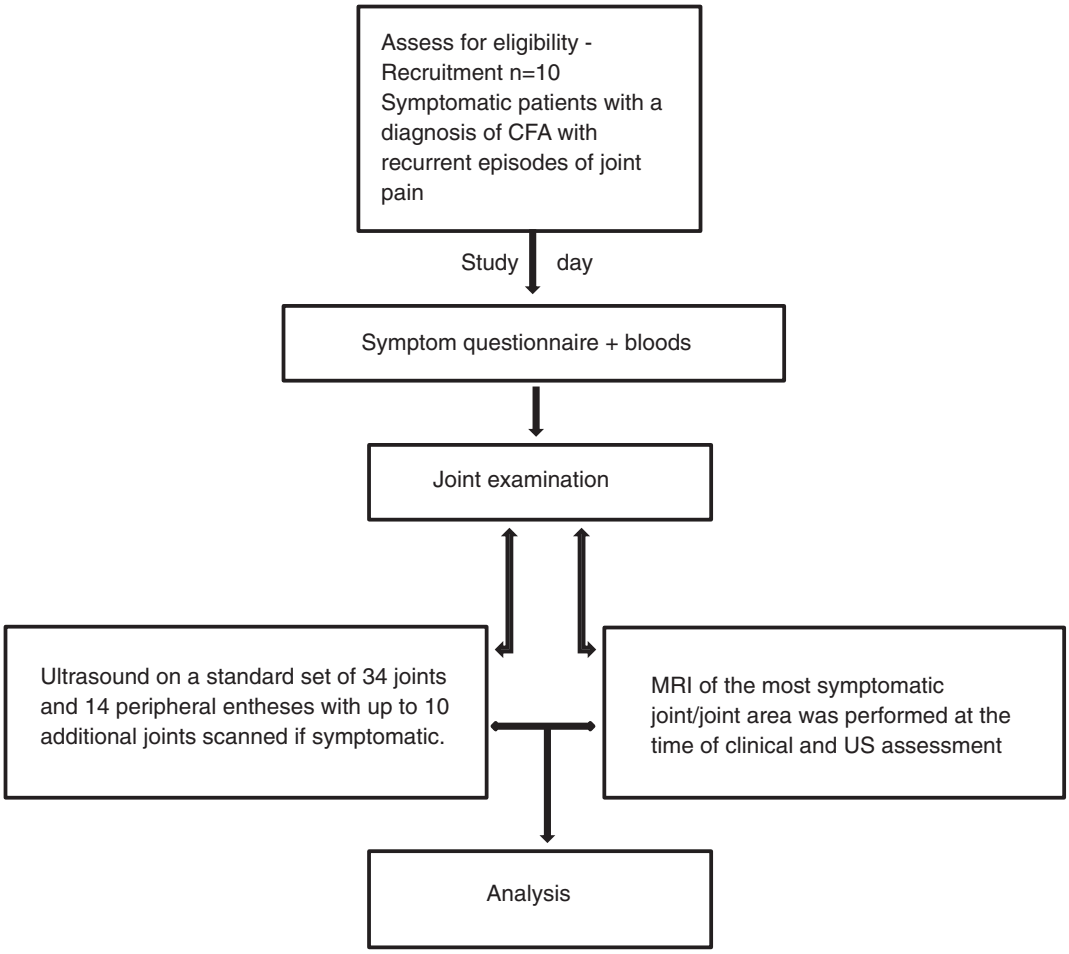

Fig. 1. Consort diagram.

they can also be severe, persistent and difficult to manage. Arthropathy may also be associated with joint effusions and in some cases a vasculitic rash [5].

Despite CFA becoming an increasing cause of morbidity in an ageing population, there is little data available about the spectrum of disease, underlying mechanism of the disease process, imaging and effectiveness of treatment [6–9].

There is no published literature describing the radiological ultrasound (US) and magnetic resonance imaging (MRI) features of symptomatic arthropathy in patients with CF.

The aims of this study were to describe the clinical phenotype of CFA, identify potential correlations between the history, examination and imaging and to assess the relative sensitivity of US and MRI as tools for assessing joint pathology in this group of patients.

2. Subjects and method

2.1. Patients

This was a prospective observational cohort study of symptomatic adult patients with CF presenting with joint pain with or without joint swelling. All patients were attending the Leeds Adult CF Centre and had classical features of CF in conjunction with either two mutations or two abnormal sweat tests. Patients presenting with symptomatic joint pain were recruited to the study (Fig. 1). Participants had serological testing followed by a clinical assessment, joint US and MRI on the same day. The study was approved by the Yorkshire and Humber Ethics committee 11/YH/0205.

2.2. Clinical assessment

Demographic, clinical and pathology data were collated from the CF unit's electronic patient record [10]. Recorded parameters are summarised in Table 1. A detailed symptom questionnaire covering respiratory, dermatological, gastrointestinal and joint symptoms as well as past medical history, drug, family and occupational history was undertaken with each subject prior to a quantitative joint assessment by an experienced physician (SD) who was blinded to the imaging results. As there is no validated joint assessment tool for CFA, a comprehensive 62 joint count was recorded, with joints assessed for pain, tenderness and swelling. The following areas were assessed (bilaterally where applicable): temporo-mandibular joint, acromio-clavicular joint, sterno-clavicular joint, shoulder, elbow, wrist, metacarpophalangeal joints1–5, proximal inter-phalangeal joints 1–5, distal inter-phalangeal joints 1–5, cervical spine, lumbar spine, sacro-iliac joints, hip, knee, ankle and metatarsophalangeal joints 1–5.