Taking a look inside: MRI of the wrist in pediatric mixed connective tissue disease (MCTD) patients

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Learning objectives

Mixed connective tissue disease (MCTD) constitutes a distinct entity, presenting combined clinical findings of four systemic disorders: systemic lupus erythematosus (SLE), systemic sclerosis (SSc), rheumatoid arthritis (RA) and polymyositis/dermatomyositis (PM/DM) [1]. As MCTD in children shows similarities to juvenile idiopathic arthritis (JIA), discrimination between these two entities, with respect to both clinical presentation and imaging features, can be difficult. MRI is the modality of choice in the imaging of inflammatory arthritic diseases [2]. In contrast to the quickly expanding research on MRI in JIA, MCTD-specific MRI features in children have not yet been described in the literature [3].

In this study we will attend the following objectives:

1. To describe findings on MRI of the wrist in a series of pediatric MCTD patients
2. To assess potential presence of MCTD-specific features in children discriminating this disorder from JIA.
Background

Clinical presentation

The term Mixed Connective Tissue Disease was initialised by Sharp et al. in 1972 [4]. They described a distinct systemic entity, which is characterised by high serum titres of auto-antibodies to nuclear RNP, along with the appearance of common clinical features of other systemic disorders, such as SLE, SSc, RA and PM/DM [4]. Thus, MCTD constitutes a clinical syndrome, commonly presenting itself with Raynaud's phenomenon, arthralgias (with or without arthritis), sclerodactyly, edematous fingers/hands and muscle disorders [5].

The disease appears to have a predilection for younger patients (mean age 31 years, SD ± 14) with a preponderance of female genre (approximately 9:1) and a mean disease duration of 15 years. Until now, there are no efficient data to estimate the prevalence of this entity [6;7].

Because childhood MCTD shows significant similarities to JIA with respect to the symptoms of arthritis, differentiation between these two entities based on clinical presentation is usually not straightforward. Besides clinical evaluation and laboratory testing, imaging of the affected joints can be helpful in a better distinction between JIA and MCTD.

Imaging in MCTD

Musculoskeletal involvement in MCTD has been thoroughly investigated with conventional radiography and is most likely to present in the wrist and knee joints with features of the aforementioned overlapping syndromes: periarticular osteopenia, joint space narrowing and marginal erosions, indicative of RA, tuftal erosions, periarticular osteolysis and calcifications suggestive of SSc, bony ankylosis and osteonecrosis, as commonly found in SLE [8]. To the best of our knowledge, only two MCTD cases with arthritic complaints investigated with MRI have been reported, by Cimmino et al. [9].

Based on that article and MR imaging findings of the overlapping syndromes, MCTD of the wrist in pediatric patients is expected to show abnormalities indicating inflammatory activity (e.g. synovial hypertrophy, tenosynovitis). Damage in the osteochondral structures of the wrist is also hypothesized, due to the often severe disease course of MCTD. Finally, the common involvement of skin and muscles might be also be visualized on MRI (e.g. soft tissue edema indicating cellulitis or myositis).
Imaging findings OR Procedure details

Patients (n=5)

To acquire information on our learning objectives for this educational poster, we evaluated wrist contrast-enhanced MRI datasets of five MCTD -clinically and laboratory-confirmed patients, using the Alarcón-Segovia criteria [1]. All MR examinations were performed at open-bore 1.0 Tesla scanner using a phased-array coil, with the patients placed in supine position. Coronal/axial T1- and T2-weighted images before intravenous contrast (gadolinium) administration were acquired. After intravenous contrast administration, coronal and axial (with fat saturation) sequences were obtained to enable assessment of synovial enhancement.

Four out of five patients showed moderate to severe soft-tissue abnormalities: extensive enhancing synovial hypertrophy (confined to radiocarpal, distal radio-ulnar and midcarpal region) and tenosynovitis in both flexors and extensors. One patient showed mild abnormalities: minimal synovial hypertrophy and only extensor tenosynovitis. Bone marrow edema and bone erosions were rarely seen (in two and one patient respectively) involving distal radius/ulna and miscellaneous carpal bones. Additional features, such as cellulitis and myositis (specific for SSc) were only found in one patient [10].

Figures

Figure 1

Synovial hypertrophy and tenosynovitis are common findings in MRI of the wrist in pediatric MCTD patients, recognizable as a combination of high signal intensity on T2-weighted images (figure 1a and 1c) and enhancement after IV contrast administration on T1-weighted images (figure 1b and 1d). The extent of these inflammatory features know large variation, indicated by the difference between two patients (upper images show minimal synovial hypertrophy and tenosynovitis, in contrast to the severe abnormalities on the lower images).

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Figure 2

The midcarpal region shows extensive synovial hypertrophy (asterix) and tenosynovitis (arrows) in the extensor tendons on axial T2-weighted images (a) and T1-weighted images with fat saturation after IV contrast (b), as also seen in very severely affected JIA patients.

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**Figure 3**

This patient presents multiple locations affected with bone marrow edema on coronal T2-weighted images of both wrist joints.

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**Figure 4**

Coronal T1-weighted images before IV contrast demonstrate extensive structural damage throughout the entire wrist. Bone erosions are visualized in distal radius, distal ulna, lunate, hamate and trapezoid bones (arrows).

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**Figure 5**

Axial T2-weighted image visualizing involvement of muscle (arrow) and skin (double arrow) with high signal intensity, indicative of soft tissue edema. Also bone marrow edema in the trapezoid and tenosynovitis in both the flexor- and extensor-compartment is present in this slice.

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**Fig. 1:** Synovial hypertrophy and tenosynovitis

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**Fig. 2:** Synovial hypertrophy and tenosynovitis

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**Fig. 3:** Bone marrow edema

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**Fig. 4:** Bone erosions

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Fig. 5: Soft tissue edema

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Conclusion

MRI findings in MCTD patients are similar to those seen in JIA: common presence of synovial hypertrophy and tenosynovitis, while bone marrow edema and bone erosions were rarely seen. The most frequent sites of involvement were distal radius/ulna, radiocarpal and midcarpal region, while carpometacarpal region was mostly spared. In our experience, unique findings for MCTD include extensive synovial hypertrophy and involvement of the flexor tendons, present in the majority of our MCTD patients, while only occurring in clinically very severely affected JIA patients. Furthermore, besides signs of soft tissue edema, MCTD-specific features were not easily identified in this case series. Future investigation should engage larger series of patients in order to extract a safer conclusion about the potential presence of unique discriminating imaging findings of this entity.
References


