



Mixed connective tissue disease with temporomandibular joint ankylosis: A case report



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ABSTRACT

Mixed connective tissue disease (MCTD) is a rare systemic autoimmune disorder, which can affect the temporomandibular joint (TMJ). The literature on TMJ dysfunction in patients with MCTD is however sparse. This is a case report of a 35-year old woman with MCTD, who presented with severe pain from the temporomandibular region. She had a disc-derangement and intra-articular erosions of her left TMJ and developed a fibrous ankylosis of her right TMJ. The conditions led to arthroplasty with interposition of a temporalis muscle flap (TMF) bilaterally. Follow-up showed re-establishment of the cortical borders of the condyles and glenoid fossae and a pain free craniomandibular articulation within a normal range of motion. This is to our knowledge the first report on TMJ ankylosis and arthroplasty with the interposition of TMFs in a patient with MCTD. The report describes the clinical progression, the surgical intervention and presents a narrative review of the literature.

1. Introduction

MCTD is a rare systemic autoimmune disorder, defined by the presence of serum antibodies targeting the U1 small nuclear ribonucleoprotein (U1snRNP) and overlapping clinical features of Systemic Lupus Erythematosus (SLE), Systemic Sclerosis (SSc) and Polymyositis/Dermatomyositis (PM/DM) [1,2]. Clinical features include Raynaud's phenomenon, polyarthralgia/polyarthritis, myositis, sclerodactyly, oesophageal hypomobility, interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH) [1–4].

The estimated mean annual incidence of MCTD is about 2.1 per million per year [2]. 7–23% of the total MCTD population have a juvenile (<18 years) onset (JMCTD) [2,3]. Polyarthritis is a common symptom, mainly limited to the hands and feet [1–3]. Joint erosions tend to develop 10–15 years after the onset of JMCTD/MCTD [1,5], and are present in about half of the JMCTD/MCTD population [2,5]. It is suggested that MCTD with erosive arthritis is a separate phenotype, less often presenting with ILD or PAH [4]. The extent of joint damage in general remains unclear [2] and very little is known about TMJ involvement, though there seems to be a higher prevalence of TMJ symptoms and severe TMJ erosions in patients with MCTD compared to healthy controls [6].

The basic immune-modulating treatment in MCTD is hydroxychloroquine (HCQ). Corticosteroids (CS), methotrexate (MTX), cyclosporine, azathioprine and mycophenolate mofetil (MMF) are used in addition when treating MCTD-related arthritis. There are

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some reports on therapy-resistant arthritis successfully treated with rituximab, and indications that TNF- α inhibitors could be used treating severe MCTD-arthritis [2].

This report presents a rare case of MCTD with severe TMJ degeneration and fibrous ankylosis, treated with bilateral arthroplasty with the interposition of TMFs.

2. Case report

The patient, a 35-five-year-old female, had an onset of general arthralgia at the age of seven and was diagnosed with MCTD at sixteen. TMJ pain developed in her late twenties. Initial TMJ-treatment included jaw exercises, occlusal splint therapy and intra-articular CS injections (IACS) by a specialist in stomathognathic physiology. As her symptoms got worse, she was referred to our department for arthrocentesis. The medical treatment was directed by her rheumatologist, and included HCQ, ibuprofen, amitriptyline, pregabalin, meperzolate, omeprazole and also recurrent IACS in the hands and feet. For reasons unknown, one had chosen not to use MTX or MMF, nor try rituximab or TNF- α inhibitors.

At referral the patient reported on a constant pain from the temporomandibular region at rest, 8 on a visual analogue scale (VAS), and intense pain when moving the jaw (VAS 10). Clinical examination demonstrated a maximal interincisal opening (MIO) of 20mm, with severe pain from the TMJs and the masticatory muscles. Bilateral arthrocentesis was performed twice within a period of two months. There was a slight increase of the MIO, a decrease in pain in the right TMJ, but persisting pain in the left TMJ.

Magnetic resonance imaging (MRI) showed a perforated, oedematous and anteromedial displaced disc of the left TMJ. On the right side the disc was intact and non-dislocated, and a small condylar subcortical cyst was present. Computed tomography (CT) revealed erosions of the articular surfaces of the eminence and glenoid fossa in the left TMJ. Both condylar heads had well-defined cortical borders. A radiopaque structure was noted in the soft tissue inferior to the articular eminence on the right side (Fig. 1).

The symptoms, clinical and radiographic findings indicated removal of the left articular disc. Due to the erosive lesions, interposition of a TMF was advocated to prevent postoperative ankylosis.

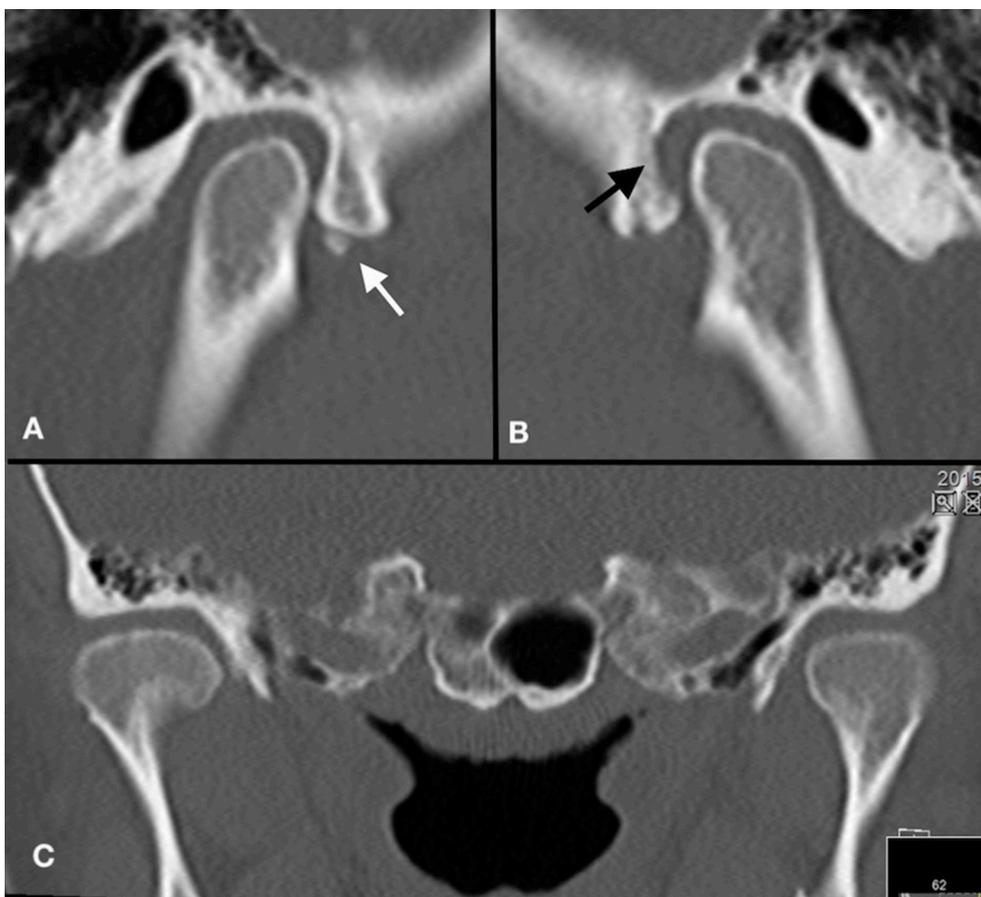


Fig. 1. The first pre-operative CT-scan. a) Sagittal view of the right TMJ, showing the radiopaque structure inferior to the articular eminence (white arrow). b) Sagittal view of the left TMJ, with erosions of the articular surface of the eminence (black arrow). c) Coronal view.

2.1. Surgery of the left TMJ

Surgery was performed under general anaesthesia. The left TMJ was exposed through an extended pre-auricular incision. The articular disc was perforated and adherent to the articular eminence, with degenerative lesions of the articular cartilage. Inflamed synovial membrane and the articular disc were removed. The eminence and the condylar head were reduced and the bony surfaces smoothed. A thin pedicled TMF was raised and folded over the zygomatic arch. The flap was interposed in the joint space and fastened with resorbable sutures. The tissues were thereafter closed in layers.

Gentle jaw movements were allowed postoperatively, and jaw exercises were initiated after two weeks.

2.2. Follow up

After seven months the MIO was 36mm. The left TMJ presented with minimal pain, but the right TMJ was distinctly painful. A new CT-scan revealed erosions of the right condylar head and the glenoid fossa, and also intra- and extracapsular bone formation. The left TMJ presented with new bone formation within the glenoid fossa, and a reduction of the condyle and eminence in accordance with previous surgery (Fig. 2).

After another three months the MIO had decreased to 29 mm with minimal laterotrusion and protrusion. There was severe pain from the right TMJ in rest (VAS 7) and on function (VAS 10). A firm swelling was palpable lateral to the joint. The clinical and radiological findings indicated an ankylosing process of the right TMJ, and the decision was made to perform an arthroplasty with the interposition of a TMF.

2.3. Surgery of the right TMJ

Exposure of the TMJ was identical to previous surgery. The joint capsule presented as a thick fibrous tissue with multiple ossified bodies, with a fusion of the lateral condyle, the zygomatic process and the articular eminence. The joint cavity was accessed through incision, and the articular disc was found absent. Thickened synovial membrane, intra- and extracapsular ossifications were removed. The eminence and the condylar head were thereafter reduced and bony surfaces were smoothed. Interposition of the TMF and wound closure were performed as previously described.

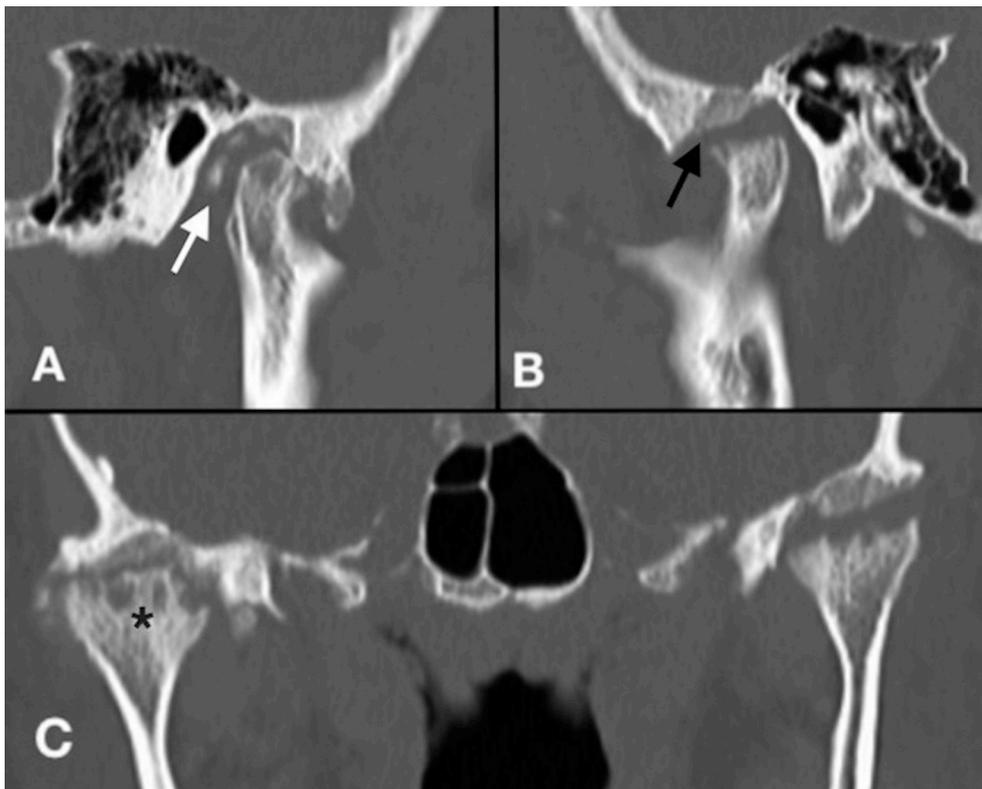


Fig. 2. The second CT scan taken 7 months after the left TMJ-surgery, 10 months after the first scan. a) Sagittal view of the right TMJ, with heterotopic bone formation within the glenoid fossa (white arrow). b) Sagittal view of the operated left TMJ, with bone formation within the glenoid fossa (black arrow). c) Coronal view, showing subcortical cysts of the right condyle (black asterisk), heterotopic bone formation and cortical erosions.

Histopathological examination showed osseous, chondromatous and fibrous tissue, with no signs of atypia or dysplasia. These findings were consistent with heterotopic bone formation, and the fibrous ankylosis was regarded as a reactive process.

2.4. Follow up

Three months postoperatively the patient had minimal pain from the temporomandibular region, a MIO of 36 mm and unrestricted laterotrusion and protrusion. After another four months she presented with an anterior open bite, which gradually worsened. The occlusion stabilised after about one and a half year postoperatively, and a third CT-scan was taken to evaluate the TMJs. This showed new bone formation within the glenoid fossae, with re-establishment of the intra-articular cortical borders bilaterally (Fig. 3). With no signs of progressive TMJ-disease, the patient underwent surgical correction of the open bite with bilateral sagittal split osteotomies (BSSO). 6 months postoperatively the TMJs were painless, the mandibular motion was within normal range and the occlusion was stable.

3. Discussion

This report presents a MCTD patient with severe TMJ involvement, who underwent bilateral arthroplasty with TMFs and subsequently BSSO.

A search in Pub-Med and Embase for TMJ disorders in MCTD, including case reports, case series and clinical studies in English literature, resulted in four papers comprising two adult MCTD populations [6–9]. No reports on TMJ ankylosis, nor TMJ-surgery in patients with MCTD were found.

Helenius et al. [6,8,9] studied a population of 15–19 patients with MCTD. Panoramic radiographs revealed TMJ erosions in 19% of the population, whereas only 1% of healthy subjects had erosions. There was also a significantly higher prevalence of pain from the TMJ area, compared to healthy controls [8]. Symptoms from the TMJs and masticatory muscles correlated with structural changes on radiographs and MRI [8,9]. The MRI findings of TMJ abnormalities in MCTD were less frequent and less severe than seen in RA [9]. Konttinen et al. [7] described a series of 10 patients with MCTD. 7 patients had subjective discomfort in the TMJs and/or masticatory muscles and 4 patients had radiographic signs of erosions of the TMJs. Clinical and radiographic TMJ involvement is reported in both MCTD, SLE, SSc and PM/DM, though generally less frequent and less severe compared to RA and JIA [10,11]. The joint alterations in this case were assumed to be MCTD-related, taking into consideration the bilateral degeneration of disk and articulate cartilage, the

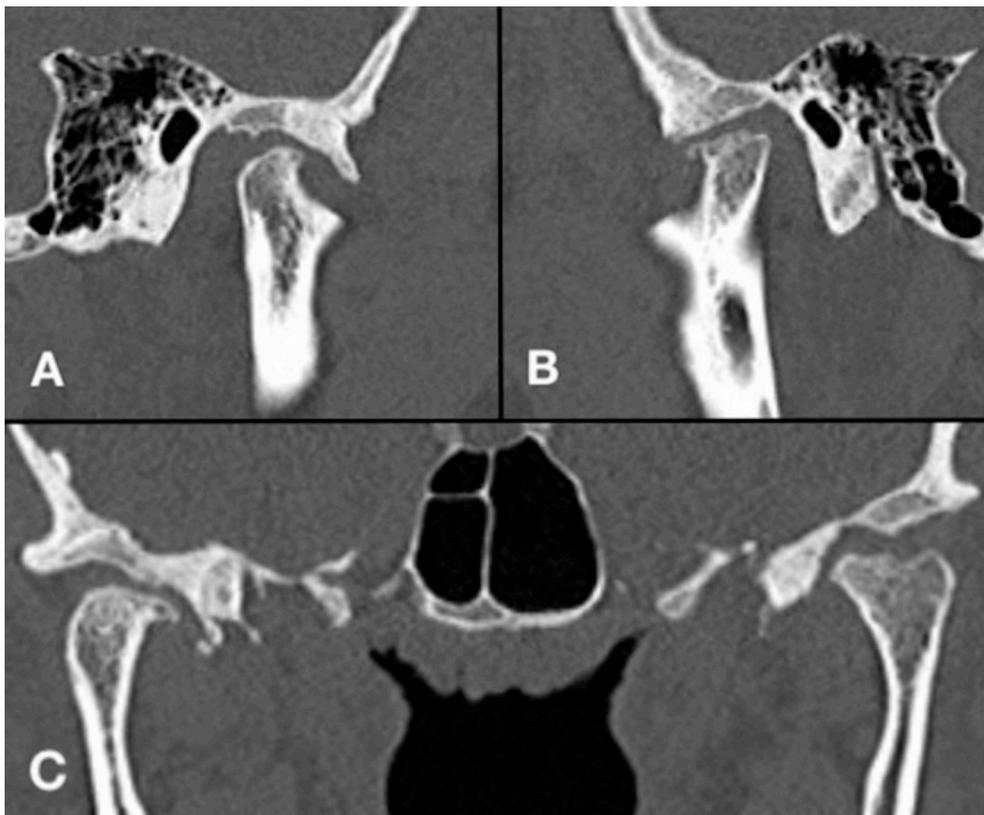


Fig. 3. The third CT scan taken 19 months after the right TMJ-surgery and 32 months after the left TMJ-surgery, showing re-established cortical borders of the glenoid fossae and the condyles. a) Sagittal view of the right TMJ. b) Sagittal view of the left TMJ. c) Coronal view.

synovial changes, the subchondral cysts and the erosions of the glenoid fossae [9,12,13]. The heterotopic bone formation was probably caused by the severe arthritis, although the multiple IACS could be a contributing factor [14,15].

Alloplastic reconstruction could have been a treatment option in this case. It is an effective long-term management of end-stage TMJ disease [16], and would have prevented the subsequent need for BSSO. The estimated lifetime risk of revision of TMJ prosthesis is though unknown, and could be an important factor considering when treating younger patients [17].

At our department the TMF is used in conjunction with discectomy and arthroplasty when there is considered being an increased risk of TMJ-ankylosis, e.g. concurrent rheumatic disease or previous ankylosis. The risk of postoperative malocclusion has to be taken into consideration, and it is possible that postoperative treatment with arch bars and elastics could help stabilise the occlusion [18]. The importance of daily physiotherapy is emphasised, as mobilisation of the TMJ is crucial in preventing re-ankylosis [19].

The use of the TMF in treating TMJ-ankylosis is well known, and could be beneficial in preventing re-ankylosis compared with no graft [19]. It has been proposed that connective tissue diseases can have an adverse effect on autogenous TMJ-grafts, with a considerable risk of re-ankylosis [16]. There are no previous reports on the use of TMFs in patients with MCTD though, and in this case the TMFs have served as stable interposition grafts.

4. Conclusion

MCTD has overlapping features of both SLE, SSc and PM/DM, diseases which all can present with TMJ disorders. One could therefore expect that TMJ involvement in MCTD would have a complex pathogenesis with varying clinical features and a diverse response to treatment. Very little is known about the prognostic factors and the progression of TMJ disorders in the MCTD population. This case report indicates that the TMF could serve as a stable interposition graft after TMJ arthroplasty in patients with MCTD.

Declarations of interest

None.

Ethical approval

Not applicable.

Patient consent

Written consent was obtained from the patient.

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