International Journal of Life Sciences, Biotechnology and Pharma Research Vol. 13, No. 7, July 2024 Online ISSN: 2250-3137
Print ISSN: 2977-0122

DOI: 10.69605/ijlbpr_13.7.2024.14

ORIGINAL RESEARCH

Surgical Management of Gynaecological Issues in Patients Suffering From Auto Immune Connective Tissue Disorders: A Case Series of Perioperative Considerations

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Received Date: 23 May, 2024 Acceptance Date: 25 June, 2024

ABSTRACT
Connective tissue or Systemic autoimmune Rheumatic diseases are characterized by autoantibodies that can affect tissues and organs throughout the body. There are more than 200 types of connective tissue diseases. This case series presents management of Abnormal uteruine bleeding, chronic pelvic pain in 3 cases, 2 cases of Mixed connective tissue disorder and 1 case of Polymyositis.Hysterectomy in a patient with Connective tissue disorder for gynaecological indications example severe menstrual bleeding, fibroids, adenomyosis requires careful preoperative planning and coordination among various medical & surgical specialties. Surgical Management of these patients require availability of HDU,ICU and multi disciplinary team approach.

Key words: Surgical, Gynaecological, Connective Tissue Disorders

INTRODUCTION

Connective tissue or Systemic autoimmune Rheumatic diseases are characterised by autoantibodies that can affect tissues and organs throughout the body.

There are more than 200 types of connective tissue diseases [1]. They may be inherited, caused by environmental factors, autoimmune triggers or most often, cause is idiopathic. These include RA, scleroderma, SLE, Polymyositios, MCTD, UCTDs etc. Polymyositis is a rare disease with incidence of 0.5 to 8.4 per 100,000 persons worldwide.

These patients may suffer from various gynaecological issues pertaining to disease itself or as a separate manifestation.

Abnormal uterine bleeding can be attributed to various underlying causes, including endometrial pathology, hormonal imbalances, and systemic diseases.

This case series presents management of AUB in 3 cases, 2 cases of MCTD and 1 case of Polymyositis . It highlights the challenges faced in managing abnormal uterine bleeding in a patient with MCTD and hypertension. MCTD typically manifests with a distinct set of clinical manifestations, serological markers, and characteristic autoantibodies, such as anti-U1 ribonucleoprotein (anti-U1 RNP). This case series aims to present a comprehensive clinical profile, diagnostic challenges, and management strategies for a patient diagnosed with MCTD and Polymyositis. By understanding the unique aspects of these diseases, healthcare professionals can improve diagnostic accuracy, optimize therapeutic approaches, and enhance patient care for individuals affected by this complex connective tissue disorder.

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CASE 1

A 46-year-old female, para 4, live 4, presented in emergency with a four-month history of heavy menstrual bleeding, passing clots, and dysmenorrhea. She also complained of easy fatiguability. The patient had a medical history of MCTD, hypertension, and a surgical history of previous tubal ligation. Laboratory investigations revealed a positive ANA (nuclear, speckled) and elevated anti-CCP (33 U/mL) and ESR (28 mm/hr) levels. Her Hb was 6 gram %. She was on prednisolone 5 mg OD, hydroxychloroquine 200 mg OD, and amlodipine 5 mg OD.She was advised an ultrasound abdomen which showed thickened endometrium ET 20 mm.

Investigations and Diagnosis:

An endometrial biopsy was performed, which confirmed the presence of a benign endometrial polyp. Additionally, focal adenomyosis and deep infiltrating endometriosis were identified on CEMRI.

Treatment and Procedure:

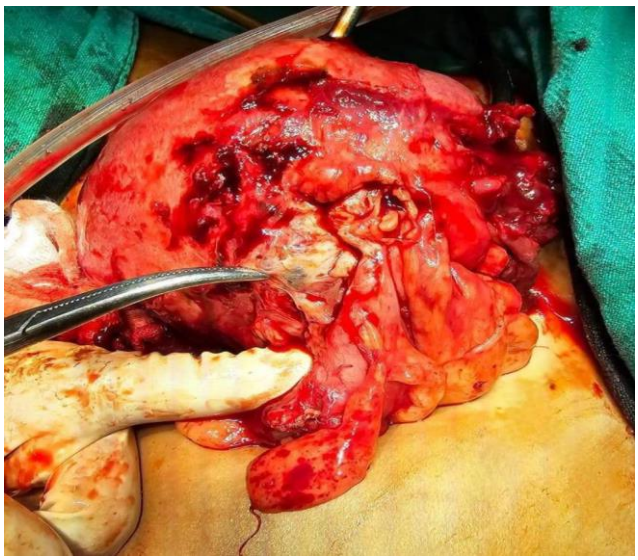
Patient was posted for TLH and was admitted one day prior to surgery. Anaesthesia fitness was taken. Prednisolone and hydroxychloroquine was given till one day prior to surgery. General anesthesia was administered.

A 10 mm infraumbilical port was inserted, pneumoperitoneaum created followed by insertion of camera and abdominal cavity visualized.

Large globular uterus visualized. Bilateral ovaries were cystic looking. Posteriorly uterus was densely adhered to rectum and sigmoid. Anterioriorlyomentum was adhered to anterior abdominal wall

Due to the patient's extensive pelvic adhesions and bowel involvement observed during the surgery, a decision was made to convert to an abdominal hysterectomy. (As shown in Pic 1)

Per operatively Uterus was buried deep in pelvis. Bowel adhered to UV fold reflection in lower uterine segment . The TAH with bilateral salpingectomy was performed successfully, and the patient was transferred to the recovery unit in stable condition.(depicted in Pic 2)



Pic 1- Dense bowel adhesions on posterior wall of uterus extending from fundus to cervix

Pic 2- Globular adenomyotic uterus with bluish spots on posterior wall

DOI: 10.69605/ijlbpr_13.7.2024.14

Postoperative Management

Postoperatively, The patient resumed prednisolone and hydroxychloroquine on the first postoperative day. Day 3 cbc done after surgery revealed severe thrombocytopenia platelets 20000, she was started on higher dose of oral steroid.

She had an uneventful recovery and was discharged with appropriate postoperative instructions and follow-up plans. 1 week follow up revealed improvement in thrombocytopenia. Patient was referred to Rheumatologist for follow up. Her histopathology showed adenomyosis uterus.

CASE 2

A case of a 39 year old female P2L2 with a history of mixed connective tissue disorder (MCTD)with pulmonary fibrosis who presented with heavy menstrual bleeding and Dysmenorrhea for 1 year not responding to medical management.

A 39 year old female para 2, live 2 presented with a 2 month history of heavy menstrual bleeding , passing clots , and dysmenorrhea. She also complaioned of joint pains.The patient had a medical history of MCTD with pulmonary fibrosis and a surgical history of previous tubal ligation.

Labortary investigations revealed a positive ANA , anti U1 SNRP positive and elevated serum IgE (131IU/ml) and normal value of CPK-MB. She was on hydrochloroquine 300mg OD and used inhaler for pulmonary fibrosis.

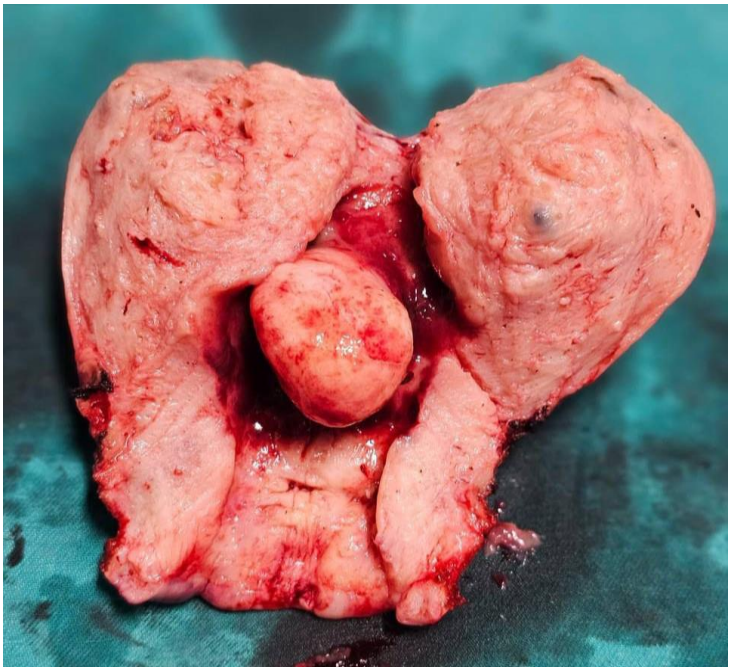
Investigations and Diagnosis

TVS pelvis showed multiple fibroids with one submucosal fibroid (approx 3x 2 cm size) and diffuse adenomyosis. CECT chest was also done as advised by pulmonologist, part of preoperative workup, showed mild pulmonary fibrosis. Spirometry was done- within normal limits. CPK-MB- 1.07(normal range)

Treatment and Procedure

Patient was posted for TAH as she was high risk for general anaesthesia pertaining to pulmonary fibrosis and was admitted one day prior to surgery. Anaesthesia fitness was taken. Hydrochloroquine was given one day prior to surgery. Spinal anaesthesia was given. Abdomen opened in layers. Uterus was 10 to 12 weeks size with multiple fibroids of variable sizes. TAH with bilateral salpingectomy and left oophorectomy was done as left ovary was cystic looking. & on cut section a submucosal fibroid of 2*3 cm present on anterior wall of uterus of uterus(FIGO type 1) and one fundal adenomyoma was seen of size approx 4 cm . (refer to Pic 3)

The TAH was performed successfully and the patient was transferred to the recovery unit in stable condition.



Pic 3 - Submucosal fibroid with focal and diffuse adenomyosis, the focal adenomyosis showed bluish spot

Postoperative Management

Postoperatively, the patient resumed hydroxychloroquine on the first postoperative day. She had an uneventful recovery and was discharged with appropriate postoperative instructions and follow-up plans. histopathology report showed focal adenomyoma & leiomyoma.

CASE 3

Total Abdominal Hysterectomy in view of abnormal uterine bleeding & continuous pelvic pressure and chronic pelvic pain in a Patient with polymyositis.

The patient, a 39-year-old nulligravida, Known case of Polymyositis, complained of heavy menstrual bleeding over the past five months, with excessive passage of clots and severe dysmenorrhea, chronic

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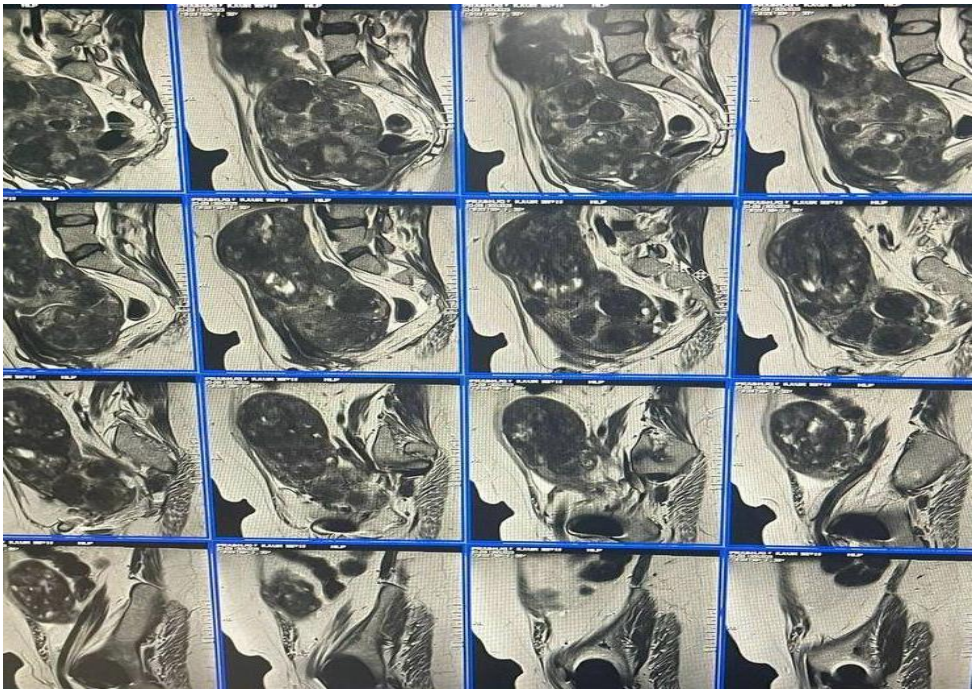
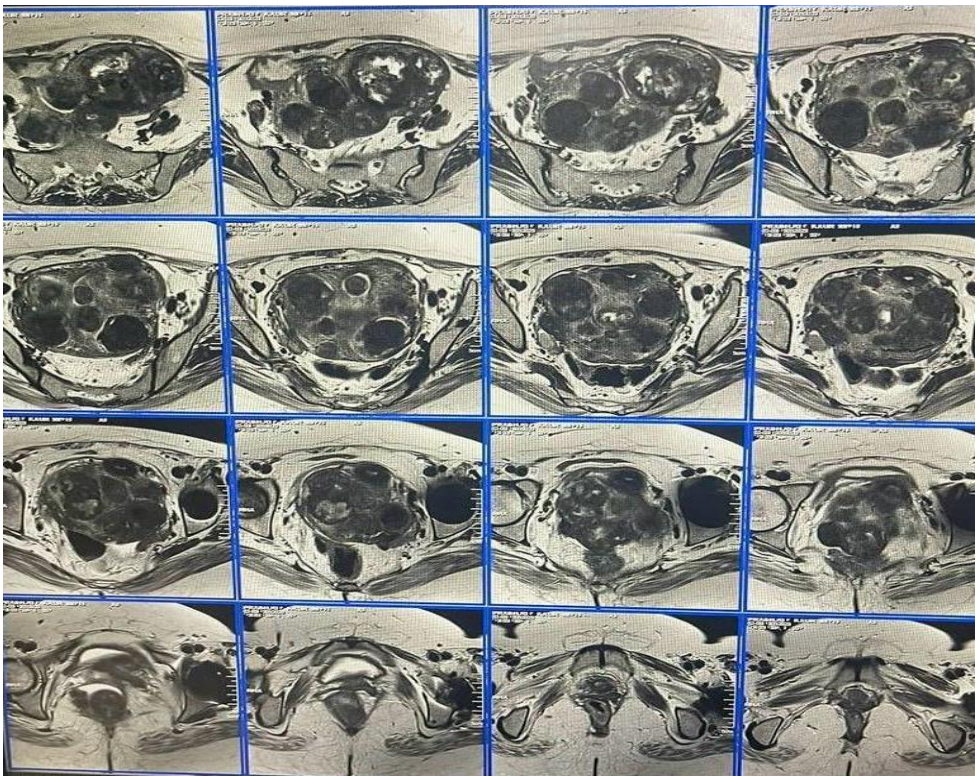
pelvic pain for 1 year with moderate to severe pelvic pressure pain off and on for 4 to 5 months. An MRI abdomen revealed an enlarged uterus with multiple submucosal and intramural fibroids. (Refer to Pic 4 and 5). Additionally, multiple cervical polyp were identified protruding into the posterior fornix. the polyp was biopsied in opd and histopath showed benign cervical polyp. The patient had a history of polymyositis, confirmed by biopsy, and was receiving treatment with methotrexate, rituximab, tacrolimus, and methylprednisolone. She had a past surgical history of left total hip replacement 8 years back and myomectomy 6 to 7 years back which was done under CSE in UK. She also had history of 2 failed attempts of IVF/ART. She had dyspnea on exertion and was limited to wheelchair during acute exacerbation of the disease.



Pic 4 - MRI Sagittal view showing multiple fibroids in patient with polymyositis



Pic 5 - MRI axial view showing multiple fibroids in uterus

DOI: 10.69605/ijlbpr_13.7.2024.14

Perioperative Management

Given the severity of the patient's symptoms and the findings on imaging, a decision was made to perform a total abdominal hysterectomy with bilateral salpingectomy under combined spinal and epidural anaesthesia . Preoperative optimization was conducted, including the discontinuation of methotrexate, tacrolimus, and rituximab in consultation with the rheumatology team. the oral prednisolone was continued till the day of the surgery(replaced by inj hydrocortisone given in immediate perioperative period) , inj rituximab was stopped 3 months prior to surgery, tablet methotrexate was stopped 1 week prior to surgery.

The patient's hemoglobin levels were optimized from 8.5 g/dL to 10 g/dL. the abdomen was opened in layers by pfannensteil incision.

Intraoperative Findings

During surgery, an irregularly large uterus was observed with distorted anatomy, approximating a size of 22-24 weeks of gestation.(refer to Pic 6) The most prominent fibroid, measuring 10-12 cm, was located on the anterior fundal region. Bilateral ovaries and tubes were found to be adherent. Adhesiolysis performed. A right ovarian cystectomy was performed due to endometriosis. Sigmoid adhesions to the posterior surface of the uterus was also present, but there was no bowel nodule. Total duration of surgery was approximately 1 hour 30 min.

Pic 6 - Hysterectomy specimen with Mutiple fibroids

Postoperative Care & Followup

Following the surgery, patient was kept in HDU for 48 hrs, she received intravenous hydrocortisone 100 mg three times daily, which was subsequently switched to oral prednisolone 10 mg on postoperative day 3 in addition to antibiotic prophylaxis. The postoperative period was uneventful, and the patient was discharged on postoperative day 5. 10 months followup of the patient post surgery is unremarkable.

DISCUSSION

Patients with Connective tissue disorders have variable gynaecological manifestations, which could be pertaining to disease per se or irrespective of the underlying systemic disorder. Patients may present with various gynaecological clinical presentations, such as heavy menstrual bleeding , dysmenorrhea, mass lesions, chronic pelvic pain, etc.

A Study was conducted by Martina Orlandi et al in women with rheumatic diseases (RDs) menstruation-

DOI: 10.69605/ijlbpr_13.7.2024.14

related disorders. The aim of this study was to evaluate gynecological symptoms/disorders in fertile age women with RDs. All patients (n = 200) filled up a self-administered questionnaire on their gynecological history, menstrual cycle pattern, menstrual-related symptoms, and quality of life (QoL). The RD group was then compared to a control group of 305 age matched fertile age women.

Among patients with RDs, 58% had arthritis, 40% connective tissue diseases (CTDs), and 1.5% systemic vasculitis. When compared to controls, women with RDs reported more frequent heavy menstrual bleeding (HMB) during adolescence (51.7 and 25.4%, respectively; p= .0001) and adult life (37.7 and 25.9%, respectively; p = .0065). Also, dysmenorrhea in adolescence was significantly more common among cases (55.6 and 45.4%, respectively; p = .0338). Gynecological pain (dysmenorrhea, non-menstrual pelvic pain, dyspareunia, dysuria, and dyschezia) in patients with RDs was more frequent than in controls (p= .0001, .0001, .0001, .0001, .0002, respectively). Considering women who reported moderate and severe symptoms in RDs, dysmenorrhea and dyspareunia remain significantly more frequent in women with RDs than in controls (p = .0001; p = .0022; respectively). QoL scores were significantly reduced in women with RDs, either in physical (p = .0001) and mental domains (p = .0014) of short-form 12. They concluded women affected by RDs frequently presented menstruation-related disorders; thus, female patients with RDs should be questioned about gynecological symptoms and referred to the gynecologist for an accurate evaluation.[2]

Nina Shigesi [3] et al conducted a meta analysis& found a potential coexistence of endometriosis & autoimmune diseases. Their Study focused on peer-reviewed published articles that reported an association between endometriosis and autoimmune diseases, excluding case reports/series, review papers, meta-analyses, organizational guidelines, editorial letters, expert opinions, and conference abstracts.

A total of 26 published population-based cross-sectional, case–control, and cohort studies

They observed associations between endometriosis and autoimmune diseases suggest that clinicians need to be aware of the potential coexistence of endometriosis and autoimmune diseases when either is diagnosed. All 3 cases in our case series had associated endometriosis or Adenomyosis, 2 patients had associated endometriosis. 3rds case, was a known case of polymyositis , had multiple fibroid uterus, & endometriosis. she underwent TAH with left salpingoophorectomy and right salpingectomy.

the 2nd case was 44 yrs old patient with AUB, with adenomyosis & deep infiltrating endometriosis, underwent TAH, left oophorectomy. Bilateral opportunistic salpingectomy was also done.

Patients with connective tissue disorders presenting with significant gynaecological complaints pose remarkable challenge in management. These patients are at risk of increased complications associated with surgical management, pertaining to their illness and drug induced immunosuppression.

As these patients are on drugs causing chronic immunosuppression, Considering surgical management in these patients pose them at high risk of surgical site infections[4], postoperative infections & other associated morbidities.

Coccolini F conducted a systematic review of literature on immunocompromised patients & Concluded that Strict adherence to SSI infection preventing bundles must be implemented worldwide especially in immunocompromised patients .

Patients with connective tissue disorders are on immunosuppressants hence are at high risk of surgical site infections. Considering all our 3 cases on immunosuppresants drugs considered as high risk for SSI, we followed all the strict surgical asepsis, intravenous antibiotic administration 1 hour prior to skin incision, we had taken these cases as 1st case in their respective OT schedules, prior to the OT fumigation of the OT was done in the morning, experienced & best surgical team scrubbed. In these patients with AUB, though medical management is usually considered as first line treatment . Surgical management is warranted in cases where Medical management fails or is not beneficial.

Apart from surgical site infections some of the connective tissue diseases pose anaesthesia risk also.

Radeka JZ [5] et al, concluded in their article that neuromuscular disorders pose anaesthetic challenges both during the preparation for surgery itself, the operative procedure and postoperative recovery in intensive care units. In these patients requiring surgery for respective indications, the mode of Anaesthesia should be individualized.

Our first case was kco MCTD, without lung involvement, so she was taken up for total laparoscopic hysterectomy under GA. But due to dense gut adhesions hysterectomy was completed by converting to laparotomy.

In our second case, Spinal anaesthesia was preferred for hysterectomy because she had moderate interstitial lung disease and administration of GA posed to be high risk considering her lung condition.

Our third case patient with polymyositis was given combined epidural and spinal pertaining to life threatening risks associated with administration of General Anaesthesia

Problems of anesthesia in patients with NMDs arise from the nature of the condition itself and the interaction of anesthetics and muscle relaxants with anticholinesterase drugs used in therapy.

similar conclusion was derived by Radkowski P [4] in their review article,they also stressed on the importance of preoperative evaluation, medical-history taking, risk of difficult intubation or cardiac incidents, respiratory insufficiency, and frequent pulmonary infections. It is also necessary to keep in mind that these patients are at risk of prolonged

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paralysis, hyperkalemia, rigidity, malignant hyperthermia, cardiac arrest, rhabdomyolysis, or even death.

Radkowski P[6] et al advised on assessment of individual risks of each patient before anesthesia. Therefore, it is important (and even necessary before major surgery) to perform a thorough preoperative examination to not only determine perioperative risk but also to ensure optimal perioperative follow-up.

Radkowski P et al[6] also mentined in their paper review that patients suffering from chronic connective tissue disease are a highly challenging population in the perioperative context. pertaining to protean organ related consequences in these patients.

Management of patients with AUB and coexisting rheumatic disorders is complex, requires multi disciplinary team approach.[7] The preoperative optimisation of the patient's medication regimen, particularly the immunosuppressive agents, is crucial to mitigate potential complications. The successful outcome of the surgery and uneventful postoperative period highlight the importance of a multidisciplinary approach to patient care.

The above case series depicts the safety of surgery in these patients in a well equiped tertiary care center with availability of adequate surgical expertise & multidisciplinary team. In such setups,surgery can be performed as and when indicated.

CONCLUSION

Hysterectomy in a patient with Connective tissue disorder for gynaecological indications eg severe menstrual bleeding, fibroids, adenomyosis requires careful preoperative planning and coordination among various medical & surgical specialties. Surgical Management of these patients require availability of HDU,ICU and multi disciplinary team approach. This case series emphasizes the significance of optimizing medication regimens, conducting thorough preoperative assessments, and addressing potential complications in achieving favorable surgical outcomes

Conflict of Interest - Author declares no conflict of interest

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