



Government of Western Australia
WA Country Health Service



Case Presentation: The Master of Disguise

Date: 16.11.2024

Presenter: Yiko Wong

Region: Goldfields –
Kalgoorlie Health Campus



Acknowledgement of Country

The WA Country Health Service acknowledges the traditional custodians throughout Western Australia and their continuing connection to the land, waters and community. We pay our respects to all members of the Aboriginal communities and their cultures; and to Elders both past and present and emerging.



CASE STUDY

- 63-year-old gentleman, antique cars painter and restorer
- Background of iron deficiency anaemia, refused endoscopy in August 2023
- Otherwise generally fit and active, non-smoker, nil alcohol

- Referred from GP in August 2023 for chronic musculoskeletal pain for 9-12 months
- Pain and stiffness over bilateral feet, knees, hips, back, shoulders, and hands
- Not improving with analgesics, anti-inflammatories or chiropractic therapies



CASE STUDY

On objective assessment:

- Limited ROM due to pain/stiffness
- Function preserved
- Nil obvious deformities

- Significant loss of weight (94 – 84kgs)
- Intermittent subjective fevers with night sweats

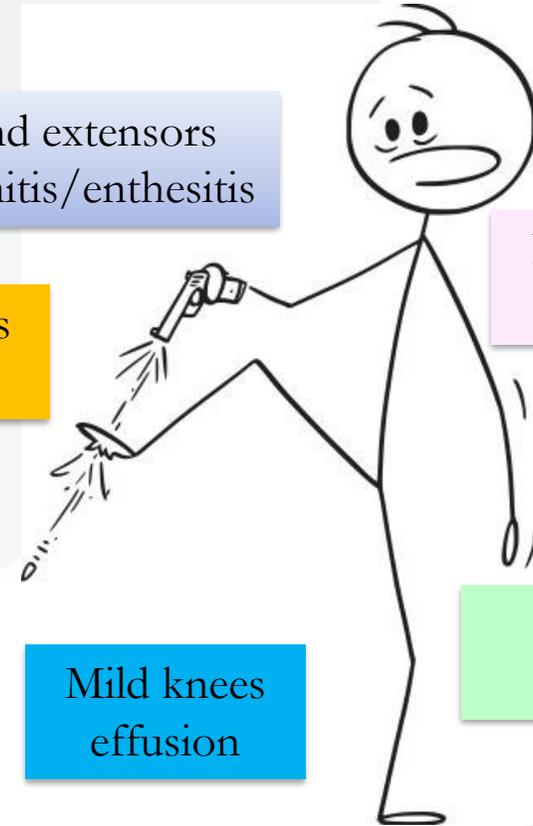
Bilateral plantar fasciitis
(USS proven)

Hand extensors
tendinitis/enthesitis

Bilateral rotator
cuffs tendinitis

Mild knees
effusion

Left tennis &
golfer's elbow





INITIAL INVESTIGATIONS

- Raised inflammatory markers - ESR 71, CRP 80, Ferritin 1000
- Hb 115
- WCC 7.82 (Neutrophils 3.14, Lymphocytes 0.81, Monocytes 3.69)
- Platelet 83

- ANA 160, LDH 271
- ENA, RF, anti-CCP, HLA B27 negative
- Tumour markers negative



IMAGING





DIFFERENTIAL DIAGNOSES

- Spondyloarthropathy with enthesitis, tendinitis, and bilateral plantar fasciitis
- To rule out malignancy or paraneoplastic syndrome (94 – 84 – 73 – 66kgs)



PROGRESS

- Further investigations:
- Pan-CT scan revealed mild hepatomegaly from fatty liver, moderate splenomegaly with splenic varices, mildly enlarged pelvic lymph nodes < 2cm
- Faecal occult blood was negative
- Endoscopy done in January 2024 (after much persuasion) showed mild diverticulum
- Inguinal lymph node biopsy was in favour of a reactive process

- Treatment flow:
Celecoxib and Sulphasalazine (September 2023)
but unable to tolerate
↓
Hydroxychloroquine (January 2024)
↓
Low dose Prednisolone (February 2024) for
symptoms control
↓
Methotrexate (April 2024) by Visiting
Rheumatologist



PROGRESS

- First peripheral blood film (technician report) showed monocytosis, which could be reactive or inflammatory, suggested to repeat in 3 months
- Repeated peripheral blood film showed a **striking monocytosis**, abnormal neutrophilic nuclear segmentation without blasts, suspicious of primary bone marrow disorder such as **chronic myelomonocytic leukaemia (CMML)**
- **CMML proven** by bone marrow trephine biopsy in April 2024



CASE STUDY IN A NUTSHELL

- 63-year-old gentleman, with long-standing seronegative tendinitis, and minimal improvement in symptoms control with DMARDs
- Significant weight loss, chronic monocytosis and chronic myelomonocytic leukaemia (CMML) – 2% blasts, 11% monocytes, cytogenetic studies demonstrated multiple poor prognostic mutations including NRAS, ASXL1, IDH2, RUNX1, TET2, and SRSF2
- Not keen for any treatment
- On regular surveillance under Haematologist
- 48% cumulative incidence risks of AML at 48 months, median survival of 18 months



PARANEOPLASTIC SYNDROMES IN RHEUMATOLOGY

Chronic Myelomonocytic Leukemia and Systemic Autoimmune Disorders: Experience in Argentina

[Maria Perusini](#)¹ · [Dorotea Fantl](#)² · [Natalia Schutz](#)¹ · [Ana Basquiera](#)¹ · [Jorge Arbelbide](#)¹

for Rheumatoid Arthritis

[Anthony Kunnumpurath](#), MBBS,¹ [Sai Prasad Desikan](#), MD,¹ [Charles McClain](#), MD,¹ and [Raman Desikan](#), MD¹

Affiliations + expand

PMID: 8507228 DOI: [10.1002/art.1780360617](https://doi.org/10.1002/art.1780360617)

Affiliations + expand

PMID: 28239882 DOI: [10.1111/ced.13071](https://doi.org/10.1111/ced.13071)

ovarian cancer



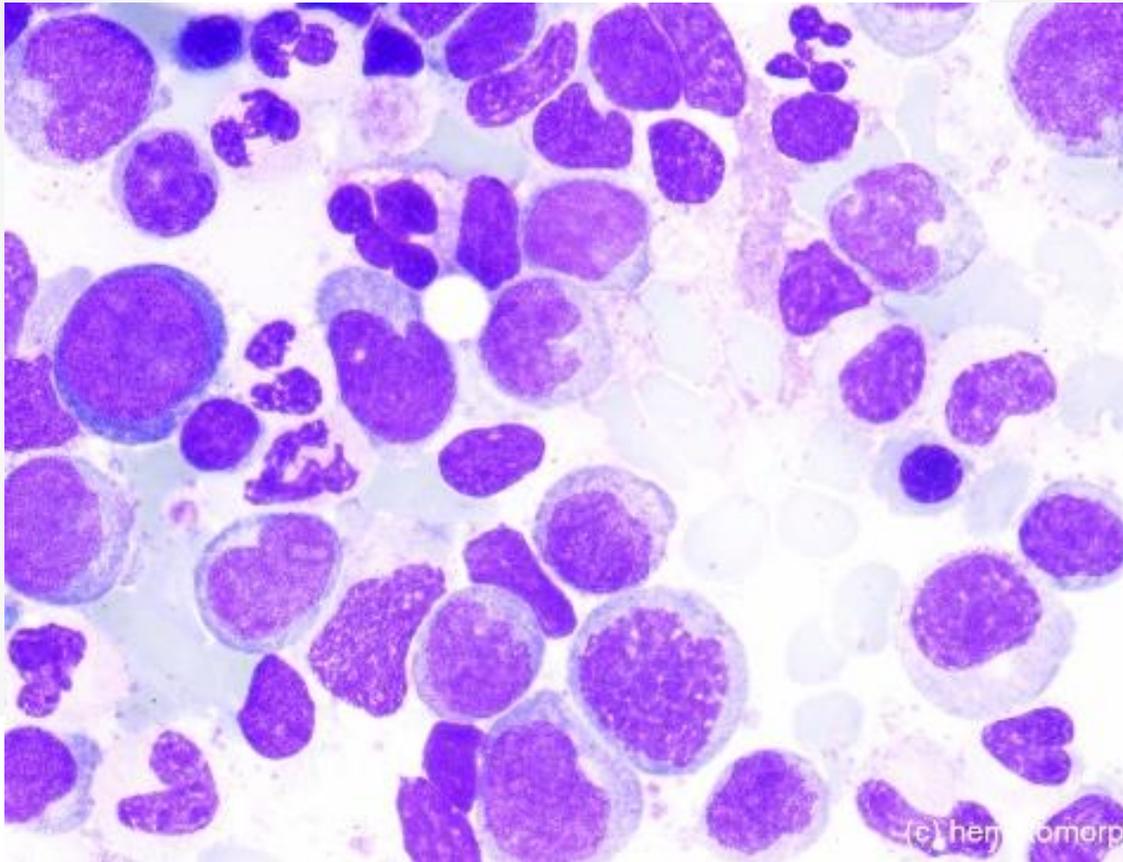
PARANEOPLASTIC SYNDROMES IN RHEUMATOLOGY

- **PFPAS** - Palmar Fasciitis and Polyarthritits Syndrome is characterized by flexion contractures of distal phalanges, fasciitis, and polyarthritits, usually seronegative
- It occurs in ovarian cancer, endometrial cancer, stomach cancer, pancreatic cancer, prostate cancer, cervical cancer, and in Hodgkin disease
- Fibrogenic and connective tissue growth factors with autoimmune mechanisms produced by the neoplasm

- Systemic Autoimmune Inflammatory Disorders (**SAID**) and polyserositis have been associated with CMML
- 10% to 30% association
- Chronic inflammation is thought to lead to oncogenesis and development of myeloid malignancies
- A significant association is shown between CMML and systemic vasculitis



CMML



- Mixed myelodysplastic myeloproliferative neoplasm, often characterised by sustained, peripheral monocytosis ($\geq 0.5 \times 10^9/L$), or monocyte count of $\geq 10\%$ of WCC
- Affects 2 in 100,000 people in Australia annually, more common in men older than 70 years
- Related to genetic mutations of TET2, SRSF2, ASXL1 and RAS
- Median overall survival of 30 months



TREATMENT

- Depends on factors including stage of disease, extent of symptoms, age, and general overall health
- Higher risks of AML transformation (15-20% over 3-5 years) with higher blasts count
- Blood products transfusion
- Cytotoxic chemotherapy – Azacitidine (For those that resemble MDS)
- Allogeneic stem cell transplant based on risk stratification by the Mayo Molecular Model (MMM) – The only modality that secures potential cure or long-term survival
- PREACH-M trial



TAKE HOME MESSAGES

- 1 Do not underestimate atypical musculoskeletal presentation in patients above the age of 55 - Suspect occult malignancy or paraneoplastic syndrome
- 2 A significant unintentional weight loss ($>5\%$ in 6-12 months) warrants investigations for malignancy
- 3 Always look at the differential counts in full blood count/order blood film

123



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QUESTIONS

COMMUNITY | COMPASSION | QUALITY | INTEGRITY | EQUITY | CURIOSITY