

Symbolic and Indicatory-Adult Onset Still's Disease

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Received: September 30, 2026; **Published:** October 05, 2026

Adult onset Still's disease emerges as an infrequently discerned, systemic inflammatory disease classically delineating a triad of clinical symptoms as pyrexia with spikes, maculopapular exanthema and arthralgia. The systemic disease demonstrating generalized lymphadenopathy frequently expounds paracortical hyperplasia and immunoblastic reaction.

The exceptionally encountered disorder commonly implicates young adults and exemplifies a female predilection [1,2].

Of obscure pathophysiology, adult onset Still's disease is associated with secretion of interleukins, macrophage colony stimulating factor (M-CSF), interferon gamma and tumour necrosis factor alpha. Besides, genetic factors, immune dysfunction and diverse infections may engender the disorder [3,4].

Adult onset Still's disease emerges as a systemic inflammatory disease expounding clinical symptoms as pyrexia with spikes, sore throat, arthralgia, cutaneous rash and hepatosplenomegaly. Additionally, reactive haemophagocytic syndrome may be observed [3,4].

Grossly, the condition is accompanied by a nonspecific enlargement of involved, miniature lymph nodes [4,5].

Upon microscopy, paracortical region expresses predominant hyperplasia. Alternatively, a 'mixed' pattern with paracortical and diffuse or paracortical, follicular and diffuse hyperplasia of lymphoid architecture is observed. Sinus histiocytosis may or may not concur. Vascular proliferation appears confined to inter-follicular areas.

Frequently, immunoblastic reaction and mixed inflammatory cell infiltration may be observed. Exceptionally, pericapsular endarteritis or features of haemophagocytosis may appear [4,5].

Adult onset Still's disease demonstrates immune reactivity to CD20 and CD3, especially within follicular and inter-follicular areas. Inter-follicular T cells appear preponderantly immune reactive to CD4, in contrast to CD8. Constituent immunoblasts delineate a B cell phenotype with immune-expression of CD30. Plasma cells expound a polyclonal light chain expression.

Adult onset Still's disease demonstrates a polyclonal profile with B and T cell genetic rearrangements [8,9].

Adult onset Still's disease requires segregation from neoplasms as peripheral T cell lymphoma (PTCL), angioimmunoblastic T cell lymphoma (AITL) and T zone variant of peripheral T cell lymphoma, not otherwise specified (PTCL-NOS) [8,9].

Adult onset Still's disease may be appropriately ascertained by cogent clinical, haematological and biochemical parameters. Concurrency with adjunct imaging features appear beneficial [8,9].

• AT-rich interactive domain-containing protein 1A (ARID1A) • B-raf proto-oncogene, serine/threonine kinase (BRAF) • Bromodomain-9 (BRD9) • Cancer testis (CT) • Caspase recruitment domain (CARD11) • C-C chemokine receptor-4 (CCR-4) • Chromodomain-helicase-DNA-binding protein (CHD)3 • CREB-binding protein (CREBBP) • Cyclin dependent kinase-2 (CDKN-2) • Cyclooxygenase-2 (COX-2) • Dynamin-3 (DNM-3) • Embryonic stem cell regulators • Eph receptor A4 (EPHA4) • Forkhead box P3 (FOX-P3) • GATA-binding protein-3 (GATA-3) • Histone deacetylase-6 (HDAC-6) • Histone-lysine N-methyl transferase (KMT)-2D or myeloid/lymphoid or mixed lineage leukaemia protein-3 (MLL-3) • MYC associated factor X (MAX) • Metallothionines I and II (MTI/II) • Methylthioadenosine-phosphorylase (MTAP) • Mitogen-activated protein kinase 1 (MAPK1) • MYC-binding protein (MYCBP) • NOTCH1 • Nuclear factor kappa-light-chain-enhancer of activated B cells (NFκB) • Nuclear factor of activated T cells (NFAT) • Phospholipase C gamma1 (PLCG1) • P21 • P53 • Phosphatase and tensin homolog (PTEN) • Plastin-3 (PLS-3) • Protein kinase, CGMP-dependent (PRKG-1) • Receptor tyrosine kinase (RTK) • Retinoblastoma-1 (Rb-1) • Ribosomal proteinS6 kinase-1 (RPS6KA-1) • Runt-related transcription factor-3 (RUNX-3) • Src homology region 2 domain-containing phosphatase-1 (SHP1) • Special AT-rich sequence-binding protein1 (SATB1) • SRC proto-oncogene, non-receptor tyrosine kinase (Src kinases) • Suppressors of cytokine signalling-3 (SOCS3) • SW1/SNF-related, matrix-associated, actin-dependent regulator of chromatin-A4 (SMARC-A4) • Tet methylcytosine dioxygenase 2 (TET2) • Thymocyte-selection-associated high-mobility group box (TOX) • Tumour protein p53 (TP53) • Tumour-necrosis factor receptor superfamily-1B (TNFRSF-1B) • Twist • Zinc finger E-box-binding homeobox 1 (ZEB1)

Table 1: Genetic dysregulation and signalling pathways in cutaneous T cell lymphomas [6,7].

Subjects depict anaemia, neutrophilic leucocytosis and elevated erythrocyte sedimentation rate (ESR). Besides, elevated serum levels of ferritin and C reactive protein (CRP) are discerned. Liver function tests are abnormal with augmented levels of serum aspartate aminotransferase (AST) and alanine aminotransferase (ALT) [9,10].

Upon radiography, involved joints delineate progressive bony erosion and narrowing of joint spaces. Frequently, the condition concurs with hepatosplenomegaly [9,10].

Adult onset Still's disease may be suitably managed with anti-inflammatory drugs or non steroidal anti-inflammatory drugs (NSAIDs), corticosteroids or antirheumatic agents [10,11].

Adult onset Still's disease is a self limiting condition wherein prognostic outcomes are contingent to specific disease pattern. Generally, subjects with localized disease display a superior prognosis, in contrast to individuals with disseminated disabilities or severe complications. The disease can be self limiting and monocyclic or polycyclic wherein clinical symptoms appear to reoccur or demonstrate a chronic, articular pattern [10,11].

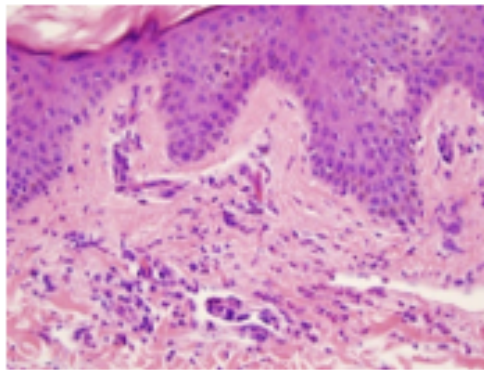


Figure 1: Adult onset Still's disease delineating a mixed inflammatory cell infiltrate comprised of neutrophils, lymphocytes and macrophages confined to the upper dermis. Fibrin is accumulated within the dermo-epidermal junction. Epidermis shows mild parakeratosis and isolated necrotic keratinocytes [12].

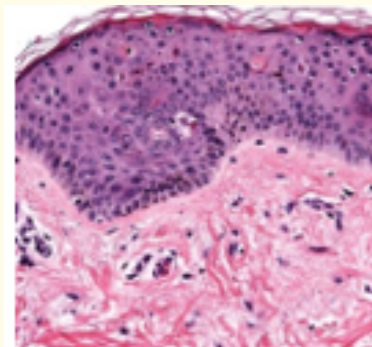


Figure 2: Adult onset Still's disease demonstrating a mixed inflammatory cell infiltrate comprised of neutrophils, lymphocytes and macrophages confined to the upper dermis. Superimposed epidermis shows parakeratosis and isolated keratinocytes [13].

Bibliography

1. Bhargava J and Panginikkod S. "Still disease". Stat Pearls International. Treasure Island, Florida (2026).
2. Cognacq G and Stratton R. "Adult-onset still's disease". *New England Journal of Medicine* 394.15 (2026): e29.
3. Ottu Para NK., *et al.* "Adult-onset still's disease: A diagnostic odyssey and therapeutic triumph". *Cureus* 18.1 (2026): e101399.
4. Rivera K and Kesari C. "Diagnostic complexity in systemic inflammation: Adult-onset still's disease". *Cureus* 18.1 (2026): e101864.
5. Aswani A., *et al.* "A diagnosis of exclusion: Unraveling adult-onset still's disease". *Cureus* 18.4 (2026): e106732.
6. Bagherani N and Smoller BR. "An overview of cutaneous T cell lymphomas". *F1000Research* 5 (2016): F1000 Faculty Rev-1882.
7. Dutra E., *et al.* "Primary cutaneous anaplastic large cell lymphoma with 6p25.3 rearrangement and epidermotropism". *American Journal of Dermatopathology* 44.3 (2022): 196-197.
8. Sheridan K., *et al.* "Delayed diagnosis: Adult-onset still's disease initially mistaken for tuberculosis in a Nigerian man". *Cureus* 18.6 (2026): e110192.
9. Saleh C. "Adult-onset Still's disease". *Medicine* 53.11 (2025): 757-761.
10. Mitrovic S., *et al.* "Periodic and non-periodic fevers". Cham: Springer. Adult-onset Still's disease (2019): 93-132.
11. Rao S., *et al.* "Adult-onset Still's disease: A disease at the crossroad of innate immunity and autoimmunity". *Frontiers in Medicine (Lausanne)* 9 (2022): 881431.
12. Image 1 Courtesy: Dermatopathology.
13. Image 2 Courtesy: Wiley Online Library.

Volume 22 Issue 10 October 2026

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