LITERATURE REVIEW

Two topics

- 1) Autoimmune haemolytic anaemia / Evans syndrome as the presenting manifestation of SLE

- 2) Dengue diagnosis in SLE patients

Auto Immune Haemolytic Anemia (AIHA)

- Haematological disturbances are common in systemic lupus erythematous (SLE) (10-83%).
- Auto-immune haemolytic anaemia (4-10%) is an uncommon disorder characterised by autoantibodies targeting RBCs
- AIHA may manifest in SLE patients at the time of diagnosis or within the first year of the disease.
- AHA is often associated with thrombocytopenia, lupus nephritis, and central nervous system activity

AIHA

- AIHA is more common in cSLE (up to 20%) than in aSLE
- AIHA is more severe in paediatric compared to adult populations
- Anti-dsDNA was the most frequent specific antibody detected in aSLE and cSLE patients, reinforcing the concept that AIHA occurs in the context of active systemic disease.

AIHA & EVANS SYNDROME

- **AIHA** is defined as presence of anemia (hemoglobin <10g/dL) and evidence of hemolysis (reticulocytosis and/or positive direct antiglobulin test (DAT)/Coombs test) at SLE diagnosis

- **Evans syndrome (ES)** is defined by the simultaneous or sequential presence of immune thrombocytopenia and AIHA, and thrombocytopenia according to platelet count <100,000/cumm in the absence of drugs.

Evans syndrome

- Evans syndrome is a rare syndrome .
- It was first described by Evan and Duane in 1951.
- It is one of the rare presenting features of autoimmune disorders, especially systemic lupus erythematosus (SLE), and sometimes may even precede the onset of disease.
- Primary Evans syndrome with no cause is very rare and is seen in children.
- It can be associated with severe haemolytic anaemia & lupus nephritis

Evans syndrome

- ES can be associated with an underlying disorder such as lymphoproliferative disorders and common variable immunodeficiency

- Although there are only few reports of an association between ES and connective tissue diseases, both diseases are caused by cellular and humoral immune abnormalities.

- Therefore, it is not surprising that systemic lupus erythematosus (SLE) is the main condition associated with ES in young adults

Association with other autoimmune disease and cancer in 26 patients with systemic lupus erythematosus (SLE) and Evans Syndrome (ES).

Other autoimmune diseases	n	Percentage
Antiphospholipid Syndrome	06	23%
Systemic sclerosis	01	3.8%
Graves disease	01	3.8%
Hashimoto Thyroiditis	02	7.6%
Sjögren syndrome	01	3.8%
Autoimmune Hepatitis	01	3.8%
Takayasu's Arteritis	01	3.8%
Malignancy	02	7.6%

Treatment

- Glucocorticoid therapy is the first-line treatment for AIHA at diagnosis, followed by the concomitant use of immunosuppressive agents

Dengue & SLE

Dengue & SLE - published literature

- **Dengue and lupus: The problem of co-occurrence - Asian Pacific Journal of Tropical Disease** December 2015.
- **Dengue fever evolving into systemic lupus erythematosus and lupus nephritis: a case report -**Lupus. 2012 Aug.
- **Dengue fever triggering systemic lupus erythematosus and lupus nephritis: a case report -** Int Med Case Rep J. 2013;

Dengue & SLE - very much like our case

Delayed diagnosis of systemic lupus erythematosus due to misinterpretation of dengue serology.

Scand J Rheumatol. 2012 Feb

Santosa A, Poh Z, Teng GG.

Systemic lupus erythematosus (SLE) is a multisystem disease with protean manifestations that **often mimic infections**, which are **well-recognized triggers** of the onset or exacerbation of SLE.

The distinction between lupus activity and infection **is complicated** by the tendency of false-positive serologies, such as monospot, human immunodeficiency virus (HIV), hepatitis C virus, and cytomegalovirus , owing to lupus autoantibodies.

- Dengue infection is prevalent in our country , leading to a high index of suspicion for DF and habitual serology testing in face of acute febrile illness
- However, fever, rash, and polyarthralgia are among the most common presenting features of SLE .
- Indeed, 4% of patients with PUO are later attributed to SLE

- Upon persistence of symptoms it was prudent to re-examine the entire diagnostic process, including re-taking the history.

- False positivity of the initial dengue IgM was supported by the lack of seroconversion of paired sera 3 weeks later.

- In addition, dengue IgM should remain positive for 8-12 weeks in a true infection

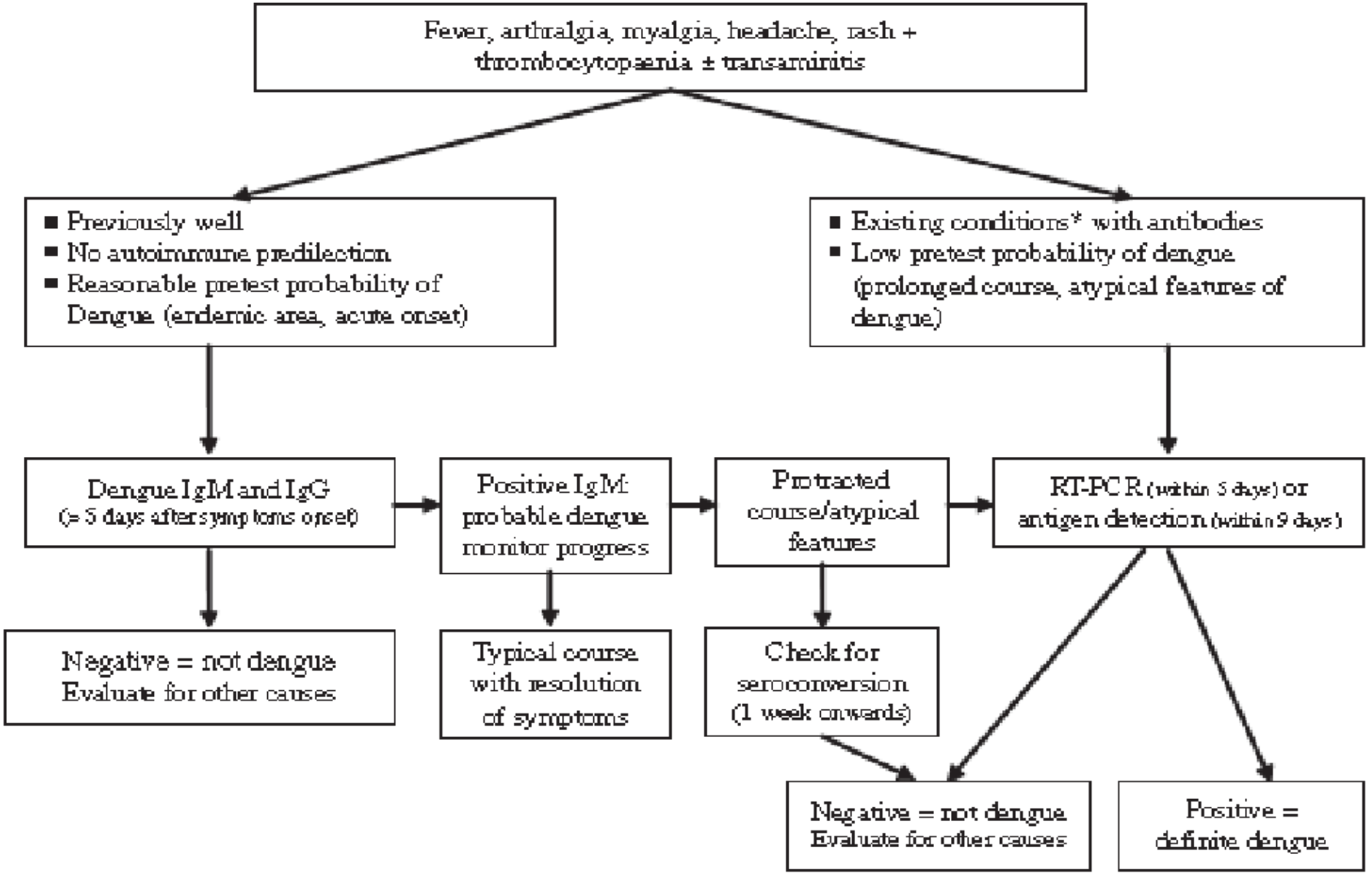
False positive results

Approximately 120 autoantibodies, arising from polyclonal B-cell activation, with mixed isotype, affinity, and avidity have been catalogued in SLE.

Antibodies with low affinity (e.g. IgM subtype) are more likely to bind non-specifically, potentially crossbinding with other antigens.

Increased numbers of autoantibodies increase the likelihood of false positive serological tests of many infectious diseases.

Approach to patients with dengue




Thank
you!!