

الحمى الروماتيزمية

Rheumatic Fever

ده واحد من اشهر الامراض فى العالم كله و بالذات فى الدول الناميه والغير متقدمه واللى بيمثل واحد من اهم اسباب ال cardiac disease وحاليا اصبح وجوده نادر فى الدول المتقدمه ولا زال يمثل big public health problem فى الدول الفقيره والناميه مثل دول افريقيا ، وسط اسيا وامريكا الجنوبيه .

طب ازاي ال Rheumatic fever بتحصل !؟

باختصار ال Rheumatic fever هو مرض روماتيزمى يعتقد بشده انه نتيجة نشاط مرضى لجهاز المناعه ومهاجمته لل host certain organs

كنوع من انواع ال Autoimmune mechanism

والمسؤول عن تحفيز ال immune system ضد ال host tissues هو وجود بكتيريا تسمى المكورات السبقيه (الميكروب السبحى)

Group A beta- hemolytic streptococci (GABHS)

بعد ما يصاب المريض الذى يتراوح سنه غالبا من بين 5 - 15 سنه ، بعدوى التهاب صديدى باللوزتين بسبب هذه البكتريا ، اذا لم يعالج بطريقه صحيحه او كافيه بعد مرور ايام الى اسابيع قليله بيحدث

Delayed immune- mediated reaction

"Autoimmune Antigen - Antibody reaction : SERUM SICKNESS : triggered by Rheumatogenic strains of GABHS infection in the form of STREPTOCOCCAL PHARYNGITIS mostly in genetically Susceptible individuals ▪

النظريه الاشهر فى تفسير حدوث المرض اسمها

Antigenic Similarity or Molecular Mimicry

ودى معناها وجود تشابه فى التركيب الجزيئى للبروتينات المكونه لهذه البكتريا واللى بتمثل ال Antigens مع التركيب الجزيئى لاعضاء فى الجسم مثل ال JOINTS , HEART , BRAIN AND SKIN

وده بيخلى جهاز المناعه ما يقدرش يميز بين البكتريا الغريبه عن الجسم و ال host tissues اللى بقت شبه البكتريا فى تركيبها و بالتالى بيحصل

Cross reactivity of the humeral immune response to such tissues resulting in

INFLAMMATION + FIBRINOID NECROSIS

✓✓✓✓✓✓

1. Heart:

Inflammation of All layers

Myocarditis + Endocarditis {valvulitis} + Pericarditis ► PANCARDITIS

تحت الميكروسكوب حاجه مميزه اسمها

Aschoff bodies

ودى عباره عن

Granulomatous lesions in the myocardium

الحقيقه ان المشكله الرئيسيه فى التهاب ال Endocardium واللى هو ال Valves ،، وبترتيب اصابة الصمامات من الاكثر اصابه الى الاقل اصابه ✓✓✓✓

MITRAL > AORTIC > TRICUSPID
Valve valve valve

وممكن يبقى اكثر من صمام حصل فيه مشاكل

NB

In the cases of Acute phase of RHEUMATIC FEVER, Acute valvulitis of one or more valves of the above mentioned valves (commonly the MITRAL VALVE) resulting in damage in its apparatus ---> Regurge

After 10 - 20 years of the Acute Rheumatic fever & Rheumatic carditis, Fibrosis occurs which lead to

Either

 Fusion of the Cusps resulting in >>>> Narrowing the valve annulus or orifice (**STENOSIS**)

Calcifications are often present.

E.g: Mitral stenosis (the most common vavular stenotic lesion in the Rheumatic heart disease.

Or

 Shortening of the Chordae tendinea and Papillary muscle dysfunction

Resulting in >>>> Valve Incompetence (**REGUREGE**)

* Chordae tendinea with the help of papillary muscle Control the movements of Valve leaflets.

E.g: Aortic Regurge (the most common valve regure in Rheumatic heart disease).

روماتيزم القلب

هو اخطر مضاعفات الحمى الروماتيزميه واكثرها تأثيرا على صحه المريض بعد مرور سنوات على Acute Rheumatic Fever وده اللى بيخلى المريض يتردد على اطباء القلب والباطنه لسنوات طويله (محتمل طول العمر) و من الممكن ان يؤدى الى مضاعفات خطيره تنتهى فى نهايه المطاف بوفاة المريض.

2. JOINTS :

Arthritis of large joints

Non erosive and heals completely without any residual deformities.

Affected joints ::::> Large joints e.g. knees, elbows, Ankles and wrists

التهاب المفاصل من النوع ده ليه حاجه مميزه جدااااا

MIGRATORY {Fleeting}

وده معناه انه بيصيب مفصل وبعدين يتنقل على المفصل التانى وبيصيب عدد اكثر من او يساوى ٥ عشان كده بنقول عليه

[POLYARTHRITIS]

Arthritis = inflammation of the Joint with

Swelling / Hotness /Redness /tenderness and limitation of movements

بعض المرضى ما بيجلهمش غير الم فقط فى المفاصل بدون علامات التهاب واضحه .

3. Brain:

Affection of the BASAL GANGLIA resulting in ●●●CHOREA●●●

CHOREA: sudden brief involuntary non rhythmic non stereotyped jerky movements like DANCING

CHOREA = derived from Latin word means (dancing)

حركات مفاجئه لا اراديه غير نمطيه تشبه الرقص واكثر حدوثا فى البنات المصابين بهذا المرض ،،

غالبا لا تحدث ال Chorea فى ال acute phase of Rheumatic fever

ولكن بعد عدة اسابيع الى شهور

4. SKIN:

RASH / NODULES

#Erythema Marginatum ::► which is an erythematous skin rash (annular Erythemous macules or Erythematous rings) occurs mostly on the trunk/ limbs . The rings have central pallor.

طفح جلدى احمر على هيئة حلقات مع احمرار اطراف ال lesion ووجود بهتان فى المنتصف [مرفق صوره]

#SCUBUTANEOUS NODULES [مرفق صوره]

Which are firm non-tender lumps located over the extensor surfaces of the bony prominences in relation to tendons {Elbows, Wrists, knees, ankles.

N.B: Subcutaneous nodules are not pathognomonic for Acute Rheumatic Fever, they also occur in other diseases like "RHEUMATOID ARTHRITIS"

Erythema marginatum



Erythema marginatum



Erythema marginatum



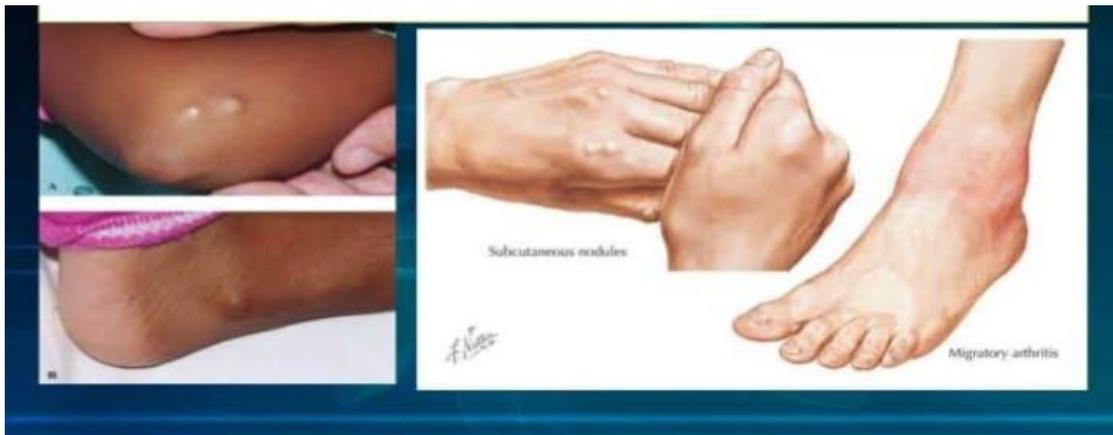
Erythema marginatum (clear centre) typically in Rheumatic Fever



Erythema marginatum



Subcutaneous nodules in rheumatic fever



4) Sydenham's Chorea:

- Sudden brief involuntary jerky non stereotyped movements similar to dancing
- Occur after an interval of months after acute rheumatic fever
- Good prognosis in the majority of cases

5) Erythema Marginatum :

- Macules of erythematous annular rash with central pallor in the middle
- The rash is non-itchy , non-painful
- mostly , they are seen on the trunk and limbs
- Prognosis: good with spontaneous resolution.

6) Subcutaneous nodules:

- Hard or firm subcutaneous lumps over the extensor surfaces of bony prominences in relation to tendons

B) Rheumatic Activity

(Subsequent recurrent attacks of ARF)

- occurs later after the initial attack of acute RF (ARF)
- Occurs in adults at a relatively older age.
- same picture of ARF
- Occurs in the patients who didn't receive the secondary prophylaxis after the initial attack up to the suitable age.

زى الناس اللي جالهم حمى روماتيزميه فى سن صغير ومش بياخدوا بنسيلين طويل المفعول بشكل منتظم.

c) Rheumatic heart disease

ودول المرضى اللي حصلهم chronic rheumatic valvulitis

بعد حدوث ال acute rheumatic fever بحالى ١٠ الى ٢٠ سنة ،، ودول ال acute carditis ترك م

Residual pathology in the previously affected valves

وحصل اما

Fibrosis & fusion of valve cusps --> Narrowing of annulus --> Stenosis

±

Shortening of Chordae tendinae with dysfunction of the leaflets --> valve incompetence --> Regurge

او

Both of Stenosis & Regurge in the same valve

مثال

Double mitral lesion (stenosis & Regurge) Or Double aortic lesion

ملحوظه هاهنا جداه

غالبا ما يكون المرضى المصابون ب RHD ،، عندهم أكثر من valve lesions مع تنوع واختلاف طبيعة ال lesion

زى مثلا mitral stenosis + aortic Regurge

وكمان

Double valve lesions

زى مثلا

Double mitral or double aortic lesions

او كلاهما مع بعض

بالنسبه للاعراض ،، المريض هيبقى باعراض ال

Valve lesion

اللى عنده ،، زى كمان الاعراض ممكن تبقى بسيطه او شديده حسب ال severity of valve lesions

واللى بتختلف من valve للتاتى وتختلف فى ال stenotic lesions عنها فى ال Regurge lesions وطبعا دى ليها معايير فى ال

Clinical examination & ECHO criteria

ايه الفحوصات اللى نعملها للمريض ده !!

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A] For patients with suspected Acute Rheumatic fever or Rheumatic activity >> Do the following

#Labs denoting the evidence of streptococcal infection

- Throat culture or Rapid antigen test
- Or Anti-Streptolysin O titre { ASOT } المتاح فى مصر

ASOT > 200 ---> positive test

Labs denoting the active inflammation

High CRP & ESR (acute phase reactants)

In case of clinical evidence of carditis ± LSHF ---> do ECHO + CXR

In case of negative evidence of carditis & heart failure --> do ECG

- Searching for prolonged P - R interval
-

هل هناك معايير دقيقه وحاسمه للتشخيص!؟

الاجابه : طبعا هناك

International widely used Criteria

Called "JONES CRITERIA"

ودى عباره عن

A) MAJOR CRITERIA (5)

- Arthritis
- Carditis (Clinical/imaging)
- Sydenham's Chorea
- Erythema Marginatum
- Subcutaneous nodules

B) Minor Criteria

- Fever $\geq 38.5 - 39$
- Arthralgia
- Acute phase reactants (positive ESR & CRP)
- ECG showing prolonged P R interval

C) Labs for Evidence of Streptococcal GABHS infection

Anti- streptolysin O titre {ASOT > 200}

عشان تشخص

Rheumatic fever

لازم المريض يكون عنده

2Major or (1 major + 2 minor)

!؟ Acute Rheumatic fever ال مريض ال نعالج

A) ttt of acute attack

- Benzathine penicillin (1, 200,000 IM

- For eradication of infection

Durapen or Pencitard اسمه في السوق

B) for arthritis

Aspirin high dose (4 - 8 gm/d : adult dose)

- RIVO 320 mg (3 × 5 or 6 divided doses per day)

Add PPI

- Alternative : Naproxyn oral

C) for carditis (if present)

- Add oral prednisolone 1 mg/kg/d

D) if heart failure

- Add supportive anti-failure measures

Diuretics + ACEi + BB

■ Then start secondary prophylaxis with

Durapen 1, 200,000 units IM/month

Alternative: Oral phenoxymethyloenicillin (Ospen 250)

If Penicillin allergy

Erythromycin (500 /12 hrs)

■ **Duration of secondary prophylaxis :**

If arthritis alone or carditis without residual lesions detected by subsequent ECHO ---> up to age of 21 years or for 5 years after the last attack

If Carditis + residual cardiac lesions (RHD)

Prophylaxis up to the age of 40 or for 10 years after the last attack

15 years old girl was brought with complaints of fever and joint pains of 5 days duration

She had pain and swelling in the left Knee and this subsided and she developed pain and swelling of the right ankle and elbow

She has a history of Sore throat 3 weeks ago, and it subsided without treatment

On exam :

She had signs of inflammation in the above joints

Heart exam: free

Chest: free

** Rheumatic fever is most likely diagnosis.

Age group 5_ 15

Throat swab to confirm group A beta hemolytic streptococci.

Fleeting arthritis is a major Jones criteria

Echocardiography in a search for carditis (major) which will determine how long the patients will be on a long acting penicillin.

** Clinically:

Erythema marginatum (major)

Subcutaneous nodules (major)

Fever and arthralgia are minor criteria for Jones.

Elevated (CRP .ESR...) minor criteria.

2: major or

1 major And 2: minor...

Jones criteria.