Case report

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Medical history

- A 44 year-old woman
- First consultation for Sickle cell disease:
  No functional sign

Antecedent:

SC sickle cell disease
Arthralgia
Clinical examination

- BCVA:
  RE/LE 20/25 P2

Slit lamp: normal
Intraocular pressure: normal
Fundus examination revealed:
How would you manage this patient?

- Photocoagulation?: PPR / retinal peripheral scatter coagulation? Both eyes?

- Absention and monitoring?

- Hemodilution?
Treatment

- Retinal peripheral scatter coagulation in both eyes

- Systemic management
Sickle cell disease retinopathy
• First genetic ocular disease

• Prevalence mondial : 2.3% of the world population.

• Haemoglobinopathy :
  Mutant haemoglobins S or C are inherited as alleles of normal haemoglobin A :
  AS, SS, SC, Sthalassemie

Severe form are associated with SC and Sthal forms .10-12% of eyes.

AS : 8% of African American
SS : severe systemic complications, 2% of African American
SC: 0.2% African American
Sthal : Severe ocular complication

Proliferative retinopathy (PSR)

Stage 1: Peripheral arteriolar occlusion
Stage 2: Peripheral arteriovenous anastomoses
Stage 3: Sea Fan
Stage 4: Vitreous Haemorrhage
Stage 5: Retinal Detachment

Leveziel N, Lalloum F, Bastuji-Garin S, Binaghi M, Bachir D, Galacteros F, Souied E. Sickle-cell retinopathy: Retrospective study of 730 patients followed in a referral center

• PPR versus retinal peripheral photocoagulation versus abstention ???

Spontaneous autoinfarction and regression of neovascularisation is well documented in PSR.


Nagpal KC, Patrianakos D, Asdourian GK, Goldberg MF, Rabb M, Jampol LM. Spontaneous regression (autoinfarction

Downes et al found that spontaneous regression occurred in 32% of PSR.

Scatter laser photocoagulation to natural course of PSR by analyzing different sea fan characteristics.

**Table 1 - New Grading of Stage III Proliferative Sickle Retinopathy**

<table>
<thead>
<tr>
<th>Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>A</td>
<td>Sea fan flat with leakage &lt; 1 MPS disc area</td>
</tr>
<tr>
<td>B</td>
<td>Elevated sea fan with hemorrhage</td>
</tr>
<tr>
<td>C</td>
<td>Elevated sea fan with partial fibrosis</td>
</tr>
<tr>
<td>D</td>
<td>Complete sea fan fibrosis without well demarcated vessels</td>
</tr>
<tr>
<td>E</td>
<td>Complete sea fan fibrosis with well demarcated vessels</td>
</tr>
</tbody>
</table>

Retinal photocoagulation for proliferative sickle cell retinopathy: a prospective clinical trial with new sea fan classification.
Results:
Spontaneous regression occurred in 10 out of 35 patients: 32% in the untreated group.

However analysis of subtype:
Grade A AND C regress in the same proportion in both groups.

Retinal photocoagulation for proliferative sickle cell retinopathy: a prospective clinical trial with new sea fan classification.
TABLE III - OUTCOMES OF NEW GRADE STAGE III PROLIFERATIVE SICKLE RETINOPATHY

<table>
<thead>
<tr>
<th>Grade</th>
<th>Evolution</th>
<th>Treated (n=38)</th>
<th>Untreated (n=35)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>A (n=20)</td>
<td>Progression</td>
<td>1 (SC)</td>
<td>2 (1 SC, 1 SS)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>Regression</td>
<td>10 (4 SC, 6 SS)</td>
<td>6 (3 SC, 3 SS)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>No change</td>
<td>1 (SS)</td>
<td>0</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>Complications</td>
<td>0</td>
<td>0</td>
<td>NS</td>
</tr>
<tr>
<td>B (n=20)</td>
<td>Progression</td>
<td>1 (SC)</td>
<td>6 (4 SC, 2 SS)</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td></td>
<td>Regression</td>
<td>9 (3 SC, 6 SS)</td>
<td>2 (1 SC, 1 SS)</td>
<td>0.02</td>
</tr>
<tr>
<td></td>
<td>No change</td>
<td>1 (SS)</td>
<td>1 (SS)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>Complications</td>
<td>0</td>
<td>5 (4 SC, 1 SS)</td>
<td>0.04</td>
</tr>
<tr>
<td>C (n=11)</td>
<td>Progression</td>
<td>2 (1 SC, 1 SS)</td>
<td>1 (SS)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>Regression</td>
<td>2 (SS)</td>
<td>2 (1 SC, 1 SS)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>No change</td>
<td>2 (SC)</td>
<td>2 (1 SC, 1 SS)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>Complications</td>
<td>0</td>
<td>0</td>
<td>NS</td>
</tr>
<tr>
<td>D (n=8)</td>
<td>Progression</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Regression</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>No change</td>
<td>5 (2 SC, 3 SS)</td>
<td>3 (1 SC, 2 SS)</td>
<td>NC</td>
</tr>
<tr>
<td></td>
<td>Complications</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>E (n=8)</td>
<td>Progression</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Regression</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>No change</td>
<td>4 (2 SC, 2 SS)</td>
<td>1 (SS)</td>
<td>NS</td>
</tr>
<tr>
<td></td>
<td>Complications</td>
<td>0</td>
<td>4 (3 SC, 1 SS)</td>
<td>&lt;0.05</td>
</tr>
</tbody>
</table>

SC = Homozygous; SS = Heterozygous; NS = Not significant; NC = Not calculated
Conclusion:
No treatment for grade A and C Sea fan: abstention and monitoring.

Retinal photocoagulation for proliferative sickle cell retinopathy: a prospective clinical trial with new sea fan classification.
Sayag D, Binaghi M, Souied EH, Querques G, Galacteros F, Coscas G, Soubrane G.
Posterior complications
Choroidal affection:

- Thinning
- Occlusion of the long posterior ciliar artery?
- May occurred in children?
• **Ischemic maculopathy:**

- Chronology with the retinopathy? Vascular anatomic role?
- Correlation with the systemic gravity? The hemodilution?
- Those changes may or may not be associated with peripheral vascular occlusion and can occur in children
PSR and Maculopathy

- Case 1: 21-year-old African American man (SS variant) early neovascularization bilaterally. SD-OCT scan revealed thinning of the temporal macula, with selective loss of the retinal ganglion cell and nerve fiber layer.

- Case 2: 42-year-old African American man (SS variant) OCT: loss of the inner layers of the temporal macula.

- Case 3: A 22-year-old African American man with sickle cell disease (SS variant) temporal peripheral sclerosed vessels with early neovascularization. SD-OCT scan revealed a similar configuration of the temporal macula, with loss of the inner retinal layers.

**Ischemic maculopathy:**

bearing the SC genotype, the risk of developing proliferative sickle-cell retinopathy (PSR) is highest between the ages of 15-24 for males and 25-39 for females; for those with the SS genotype, it is higher between the ages of 25-39 for both sexes.


Age for ischemic maculopathy?


Correlation with the retinopathy ? Th systemic gravity ?

Children in CHIC hospital ??
The emerging treatment options that can be employed to improve tissue perfusion in these patients include:

I. Increased HbF: hydroxyurea, Omega-3, erythropoietin, 2-deoxy-5-azacytidine.
II. Erythrocyte hydration: clotrimazole, magnesium pidololate.
IV. Antioxidant therapy: glutamine, deferiprone.
V. Antithrombotic agents: heparin, ticlopidine, warfarin.
VI. Vasodilation: nitric oxide, arginine, Flocor.
VII. Decrease in HbS cells: transfusion, apheresis.
VIII. Hematopoietic stem cell transplantation and gene therapy.
THANKS FOR YOUR ATTENTION