**ISSUE** 

February

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OSF NEWSLETTER OF THE QUALITY SLIDE PROGRAM

# Last Month's Slides

# January 2021 Slide **Summaries**

## Slide 1

**Circulating follicular** lymphoma. Monomorphic lymphocytosis consisting of small lymphocytes with a high nucleocytoplasmic ratio and a nucleus sometimes notched "in coffee beans".

# Slide 2

Mantle cell lymphoma

# Slide 3

Follow-up of a hereditary spherocytosis. Anisocytosis (++). Polychromasia (++). Spherocytes (+++). (See article on page 2)

#### Slide 4

Multiple alarms on analyser. Blood blastosis.

Slide 5 Nothing abnormal to report.

Slide 6 Nothing abnormal to report.

# **Monthly Digital Case study** Slide 2

#### Presentation

02 January 2021 Male (76 years old) URGENT: Exploration of a hyperlymphocytosis. Immunophenotyping of circulating lymphocytes in progress.

# **FBC Results**

WBC 27.5 (10^3/mm3) Neutrophils 13 % RBC 3.91 (10^6/mm3) Lymphocytes 8.3 % HGB 10.3 (g/L) Monocytes 0.9 % НСТ 35.0 (%) **Eosinophils** 0.9% MCV 90(fL) **Basophils** 0.0 % MCH 26.3 (pg) MCHC 29.4 (g/dL) PLT 124 (10^3/mm3)

# Slide review

Cytology: Rather large cells, with high nucleocytoplasmic ratio, nucleus with an irregular outline (notches, foliate appearance) and with a rather basophilic cytoplasm.

# Diagnosis

Mantle Cell Lymphoma.

Mantle cell lymphoma (MCL) is a distinct subtype of mature B-cell non-Hodgkin lymphoma (NHL) that accounts for 5-10% of all NHL. MCL aggressive NHL with characteristic morphology and immunophenotype, which typically presents in elderly patients with advanced disease and shows a poor prognosis.

# This issue

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# Monthly Morphology Quiz:



What is the name for the cell in the red box and what condition may you see them in?

# Last month's Quiz:

Who first discovered haemoglobin?



The haemoglobin compound was discovered in 1840 by Friedrich Ludwig Hünefeld.



Explore the future



QSP case 3 slide showing spherocytic red cells

#### Introduction

Hereditary Spherocytosis (HS) is a heterogenous group of disorders characterised by the presence of Spherocytes, Clinical features may include anaemia, jaundice, reticulocytosis and splenomegaly.

#### **Clinical Presentation**

03 January 2020 Female (45 years old) WBC 5.5 (10^3/mm3) RBC 3.37 (10^6/mm3) HGB 10.5 (g/dL) HCT 30.5 (%) MCV 91 (fL) MCH 31.2 (pg) MCMH 34.4 (g/dL) PLT 183 (10^3/mm3) Follow up of Hereditary Spherocytosis

# Hereditary Spherocytosis

An overview of the clinical features, laboratory findings and treatment



QSP case 3 slide showing spherocytic red cells

# Hereditary Spherocytosis (HS)

Hereditary Spherocytosis (HS) was first described in 1871 and the first recorded splenectomy performed soon after. HS shows a wide range of features in regard, severity, mode of inheritance and the actual cause of the protein defect. It is a relatively common disorder with an incidence in North Europe and North America of between 1 in 2000 to 1 in 5000 (depending on method of analysis). Affected patients will show a variable degree of anaemia, jaundice, reticulocytosis and splenomegaly with the clinical severity ranging from symptom free (carrier) to severe haemolysis. Exacerbation of clinical severity can be caused by illnesses such as Infectious Mononucleosis. The majority (75%) of cases will have a family history even if HS has not been formally diagnosed e.g. anaemia, splenectomy, gall stones, jaundice. The diagnosis of HS has been known to be made from birth all the way up to people in their 80's-90's. Sometimes the diagnosis is made serendipitously. HS carriers may not show anaemia, jaundice or splenomegaly and with no spherocytes seen on the blood film.

### **Red Cell Morphology**

Normal red cells are biconcave discs which have a large surface area to volume and have no nucleus, allowing the red cell to contain relatively large amounts of Haemoglobin. Red cells need to be flexible in order to travel through tiny capillaries so that they can absorb and deliver oxygen where needed. A disturbance in the cytoskeleton can have a profound effect on the flexibility and eventual life span of the red cell.



### **Red Cell Morphology Continued**

Red cell membrane integrity is dependent upon several proteins with spectrin being one of the main contributors. Abnormal red cell morphology seen in HS is due a specific genetic mutation leading to a deficiency of, or a dysfunction in spectrin, ankyrin, band 3 and or protein 4.2. The defect means that the red cells can not retain their biconcave shape and become spherocytes.

Spherocytic red cells are removed from circulation by the spleen resulting in splenomegaly, anaemia and an increase in bilirubin.

#### Diagnosis

Spherocytes can be distinguished from normal red cells by the uniform dense colour of the cytoplasm and can be smaller than that of a normal red cell.

As described above, the clinical picture in HS is varied but if Spherocytes are seen in the blood film a Direct Antiglobulin Test (DAT) should be performed to exclude auto haemolysis. A reticulocyte count should also be performed to confirm the degree of excess red cell production.

A family history is essential in the correct diagnosis of HS with note being taken of family members who have had a splenectomy for no obvious reason or gall bladder problems.

Additional confirmatory tests, such as Osmotic Fragility, Acidified Glycerol Lysis time (AGLT) and Pink test ( a modified AGLT) are all based on the fact that spherocytosis are more prone to lysis than normal red cells.

#### **Disease Management**

Folate therapy is recommended in children with moderate and severe HS. The patient should be offered a regular review particularly those with moderate to severe form as splenectomy may be required.

# This Month's Top Morphology Tip

Eosinophils and basophils are myeloid cells, formed during haematopoiesis. Although both are granulocytes, they each have different characteristic which allow you to identify them within a film:



- Eosinophils are 12-17uM in *diameter*
- They have TWO LOBES
- Large acidophilic granules which stain bright red (sometimes connected giving the appearance of an old telephone)
- 1-6% eosinophils in circulating WBC



- Basophil
- Basophils are 10-14uM in diameter
- Bean Shaped Lobes
- Prominent dark blue/purple staining of granules in basic stains
- 0.5-1% of WBC's- basophils are not commonly noted in a normal film

#### **QSP 2.0**

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#### **Bibliography**

Blood Cells A practical guide – Barbara Bain

Guidelines for the diagnosis and management of Hereditary Spherocytosis A BMJ Guideline publication

<u>Guidelines for the</u> <u>diagnosis and</u> <u>management of</u> <u>hereditary spherocytosis,</u> <u>bjh guideline</u>

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