Diseases of the Red Blood cells

The 27th SCOR Global Life Medical Symposium, December 6th 2012, Paris
To share with our clients the latest developments in medicine and their impact on the Life Insurance business.

Meeting devoted to
Chronic anemias and polycythemia, thalassemia and sickle cell disease.

According to the WHO, anaemia, especially iron deficiency anaemias affects approximately 2 billion individuals around the world. Sickle cell disease is now the most common genetic disease in the Paris region and in France generally. Although some forms of anaemia have a relatively moderate impact on health and are reversible with treatment, they are always associated with high morbidity. Other more severe forms are life-threatening. Whether acquired or inherited, they are a major public health issue.

We know that anaemia is the expression of an underlying disease. With Regards to anaemia caused by vitamin B12 deficiency, do the neurological complications always regress with carefully monitored treatment? Is vitamin B12 deficiency more common than we think?

Professor Frédéric GALACTEROS

The neurological complications always regress, but sometimes only partially and may leave minor residual sequelae. The degree of regression depends in fact on the stage of the patient’s illness when treatment is begun. When it is too late, at the senility stage, we simply make a differential diagnosis of degenerative dementia. In Addison’s (Biermer’s) pernicious anaemia, which is a deficiency-related form of erythropoietin insufficiency (selective vitamin B12 deficiency) and which is not uncommonly seen in emergency departments, the regression is generally quite good, particularly the central nervous system signs. On the other hand, the peripheral signs can take a long time to regress, with sensitivity disorders which can cause falls, particularly in the elderly.

It is now known that it is very important to diagnose vitamin B12 deficiency in elderly patients showing signs of disorientation or who have had falls, etc. It is a notion that is now firmly established in geriatric and emergency departments. B12 deficiency does not always mean Biermer’s or Addison’s anaemia. It can be due to changes in absorption in elderly patients or sometimes to a diet without enough animal protein. Most often, it is caused by a combination of dietary problems and absorption difficulties, which can often be explained by the very high frequency of senile gastric atrophy. From the age of 75-80, at least 30% of people suffer from reduced or absent gastric acid secretion, which is an essential factor in the absorption of B12.

Haemolytic anaemia can be a symptom of many diseases, with intrinsic or extrinsic causes. Which drugs are most often involved in non-immune forms such as G6PD deficiency?

These are sulfamides, sulfones (very little used in France) and quinolones (including the 3rd generation ones). Among the sulfamides, the most dangerous (in children) are the antibiotics commonly used for ear infections. Concerning aspirin, it takes exposure to high doses over several days to cause haemolytic anaemia, and it occurs mainly with the most severe forms of deficiency, i.e. the rarest. There are no reactions with aspirin with the more common moderate forms.

It is true that patients on long-term exchange transfusion programmes can develop quite serious secondary haemochromatosis. This problem arises less often in patients with thalassaemia as they start chelation therapy very young and patient compliance tends to be higher than in sickle cell patients. As the classic treatment with deferoxamine administered subcutaneously is quite painful, the availability of oral iron chelators is of great interest to us. However, the drawback is that they are sometimes poorly tolerated, a disadvantage that compounds the problem of poor compliance among sickle cell disease sufferers. When we combine the drugs, we monitor kidney and liver function, to avoid the – rare – problems that could occur. In the final analysis, it can be said that chelators are globally relatively effective.

What advice would you give to a woman with beta-thalassaemia minor wishing to get pregnant?

My advice would be to make an appointment to see a genetic counsellor and to take her partner along so that he can also be tested. Prenatal diagnosis is also offered, if the couple wishes. When both parents carry the gene of beta-thalassaemia, prenatal diagnosis is often requested by the couple, as it is quite a serious disease. In genetic counselling, we also offer to test the siblings (unmarried brothers and sisters in particular) and we advise that the family be informed.

Treating genetic haemoglobin disorders makes you expert in secondary haemochromatosis. How effective are oral iron chelators?

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