SURGERY IN SICKLE CELL ANEMIA

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SUMMARY :

Sickle cell anemia is a hemoglobin disorder with a wide range of clinical manifestations and complications. Medical treatment is the mainstay of management but surgery is indicated in some cases. The authors review the main surgical indications in sickle cell anemia as frequently encountered and review treatment guidelines.

KEY WORDS: – Sickle cell anemia – Surgery – Indications.

RESUME :

La drépanocytose est une hémoglobinopathie avec des multiples manifestations et complications. La prise en charge est surtout médicale mais cependant, la chirurgie est indiquée dans certains cas précis. Les auteurs passent en revue les indications chirurgicales fréquemment rencontrées et font le point des traitements chirurgicaux.

MOTS CLÉS : Drépanocytose – Chirurgie – Indications.

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Vaso-occulsion resulting from the polymerisation of sickled erythrocytes in small and sometimes large vessels cause vascular injury leading to multiple organ dysfunction and complications. Treatment is rapidly evolving and has been reviewed by several authors in this Journal. In this chapter, major surgical indications in sickle cell anemia (SCA) directed at disease complications are discussed.

A- BONE AND OSTEOARTICULAR COMPLICATIONS

1- OSTEOMYELITIS

This is frequent after 6 months and rarely before 3 months [1]. Extremities are mostly affected in infants, and long bones in older children [1]. In sicklers, bone lesions are multifocal, bilateral and symmetrical and affects mostly the distal part of long bones [1]. The main frequent germ is salmonella [1-2]. In some studies in Nigeria Staphylococcus aureus is most frequent [3].

Factors which explain the osteomyelitis in sickle cell patients are bone infarction, multiple hospital admissions (which increase the risk of contact with germs), disorders of leucocyte phagocytosis due to hypoxia, functional asplenia and abnormalities of the alternate pathway of complement activation [1,4]. It has also been demonstrated that in sicklers, salmonella frequently cross the digestive mucosa into mesenteric lymph nodes, blood and bone marrow, avoiding phagocytosis because of the inability to use the alternate complement pathway [1]. Marrow congestion and formation of vascular thrombosis produces zones of infarction and necrosis which protects the germs from the action of antibodies and antibiotics. Finally micro-abscesses eventually form, resulting in osteomyelitis[1]. Antibiotics and plaster immobilization are necessary, to prevent pathologic fractures. Surgery is indicated to extract bone sequesters and abscesses [1].

2- SEPTIC OSTEOARTRITIS

These are less frequent than osteomyelitis. The most frequent germs are Staphylococcus aureus, Escherichia coli, and Salmonella [4]. Following thickening of the articular cavity and soft tissues, the articular space reduces with subsequent osteoporosis and bone destruction. Diagnosis is made by tapping and analysis of the articular fluid. Treatment is by arthrostomy and irrigation drainage with antibiotherapy and immobilisation of the joint.

3- BONE INFARCTION

Falciformation of erythrocytes produces vaso-occulsion with cellular sequestration and infarction as the ultimate consequence. Stasis and capillary congestion produces more anoxic conditions which aggravate falciformation. Clinically, there are bone pains, soft tissue tumefaction, fever, leucocytosis and increased erythrocyte sedimentation rate. Long bones (especially at the diaphyso-mataphysary junction) are affected in the older sickler child and adult. Preferential localisations by order of frequency are, proximal parts of the femur and humerus, distal zones of the femur and proximal zones of the tibia. The diaphysis of other bones could also be affected [4]. Epiphysary infarction is also frequently observed in the homozygote adults than in children. This is because, the cartilaginous part receives its nutrition from the synovial fluid than from the arterial ciculation. Epiphysary infarction is often bilateral, with preference to the femoral and humeral heads. Sometimes distal epiphysis of the femur, proximal zones of the tibia and distal zones of the humerus [4]. Surgery is indicated to treat and prevent pathologic fractures which often complicate bone infarction. Careful cleaning of the joint to the cartilage level could be useful in some cases.

4- ASEPTIC BONE NECROSIS

It occurs in 10 to 50% of adults with SCA [2]. This complication occurs most commonly in late adolescence and early adult life although it can be seen as early as 8-10 years [5]. The pathogenesis is not clear, but may be related to the fact that, these structures are essentially enclosed structures with a single incoming artery and outgoing vein. Increased viscosity of blood retards the venous outflow, resulting in an increase in tissue pressure within the enclosed input with resultant hypoxia of the marrow and bone. Aseptic necrosis of the proximal epiphyseal segments of the humeri and especially femora is a relatively common event in sickle cell disease [6]. Continued weight bearing on a softened femoral head may lead to damage, irregularity of the joint surface and painful limitation of movements.

Clinically, there is persistent pain on walking, localised to one hip and accentuated on climbing steps. Examination shows painful limitation of passive movements of the affected hip, especially on internal and external rotation. Early diagnosis and prevention of further weight bearing, may limit destruction of the femoral head. Occasionally traction in the hospital may be necessary [5]. Symptomatic osteonecrosis of the hip in SCA has a likelihood of leading to femoral head collapse, necessitating surgical intervention. When osteonecrosis develops, deterioration is rapid and in most patients, surgery is necessary because of intractable pain [7].

If permanent damage occurs, limited remodelling functional limitations may require total hip replacement. This is rarely indicated before young adult life [5].

5- OTHERS [4]

Fractures are frequent in sickle cell patients affecting mostly long bones or vertebral. Osteoporosis, infarction and osteomyelitis are contributing factors. Hemarthrosis could also complicate epiphyseal necrosis. Non hemorrhagic articular effusions have also been described, and are localised on the knees and elbows and last a few days.

6- LEG ULCERS

They occur in 20% of adults with SCA [2]. They develop most frequently between 15-20 years. These ulcers occur spontaneously or as a result of local trauma with subsequent infection and skin necrosis but no specific organisms have been incriminated. Other contributing factors are vessel occlusion by sickled cells, increased venous pressure and decreased oxygen carrying capacity with resultant tissue hypoxia. The majority of ulcers occur at the lower third of the leg above the ankle, less commonly on the dorsum and rarely on the sole of the feet. The left leg is involved in 30% of the cases, right leg in 20% and both legs in 49% [8]. Healing is slow and ulcers are prone to spontaneous relapse. Treatment includes debridement with proteolytic enzymes, regular dressing at home twice daily with mild antiseptics. Oral zinc sulphate (200 mg three times daily, significantly improves healing. Skin grafts may be used in clean vascular ulcers, but walking before complete healing commonly leads to failure of the pinch grafts [5].

B- SOFT TISSUE COMPLICATIONS

1- ADBOMINAL PAINS

Episodes of abdominal pains sometimes mimicking a surgical abdomen are frequent in children with SCA and may result from a variety of mechanisms [5,8]. The pain can be attributed to marrow infarction of the vertebral bodies with subsequent pressure on nerve roots or enlarged mesenteric and retroperitoneal lymph nodes. Other differentials include splenic diseases (acute splenic sequestration crisis, splenitis, splenic infarction and abscesses), hepatobiliary diseases (biliary colic, acute cholecystitis, hepatic crisis, hepatitis and liver abscesses), peptic ulcer disease and ischemic colitis. Abdominal tuberculosis, fatal venous thrombosis of the hepatic, postal, superior mesenteric and splenic veins and acute pancreatitis should be considered [8]. Careful evaluation of each patient, based on history, clinical examination, laboratory and radiologic investigations is essential to avoid unnecessary surgical interventions with their inherent risks [8]. The pain in non surgical conditions is relieved with hydration and oxygen in 97% of cases within 48 hours [9].

2- SPLENIC DISORDERS

Due to repeated splenic infarction, the spleen undergoes fibrosis and becomes a fibrosiderotic nodule (autosplenectomy). Repeated acute sequestration crisis or infection of an infracted area with abcess formation often occurs [8]. Because splenectony is associated with a high risk of post-splenectomy sepsis and the tendency towards splenic atrophy in adulthood in SCA, it is reserved only for patients with recurrent acute splenic sequestration crisis, splenic abcesses and less commonly persistent massive splenomegaly. Patients should receive polyvalent pneumococcal and Hemophilus influenzae vaccines two weeks before elective splenectomy. In case of emergency splenectomy, the vaccines should be given immediately post operatively. Long-term penicillin prophylaxis is indicated, until adulthood or at least two years following splenectomy to reduce the incidence of post splenectomy sepsis [8].

Acute splenic sequestration crisis (ASSC) is a life threatening complication of SCA. It is rare in adults due to progressive splenic fibrosis as a result of repeated infarctions and occurs mostly in infants and young children less than 8 years. The exact pathogenesis is unknown. Splenic outflow obstruction with sequestration of red cells and platlets is a triggering factor. Splenectomy is reserved for selected patients with recurrent episodes of ASSC or who may develop red cell alloantibodies which may hamper future transfusions [8].

3-CHOLELITHIASIS

Patients with SCA are at high risk of developing pigmented gallstones due tot chronic hemolysis. These form as early as 3-4 years and reach an incidence of 40% in SS patients by the age of 20 years [5].

They are usually small and multiple and cause symptoms by obstructing the cystic duct and common bile duct or because of acute or chronic cholecystitis. Elective laparascopic cholecystectomy is recommended for pediatric patients with SCA to prevent the risk of an emergency cholecystectomy procedure.

The primary benefits of this approach is a shorted hospital stay after surgery, decreased post-operative discomfort, decreased risk of complications and a greater return to normal activities [10].

4- GENITO-URINARY DISORDERS

Priapism is a painful erection of the penis and affects 30-40% of post-pubertal Jamaican male patients with SS disease [5]. There are two clinical patterns : stuttering episodes lasting 3-4 hours and does not impair normal sexual function and major attacks lasting more than 24 hours with extreme pain, often penile oedema and usually followed by irreversible damage to the vascular erectile system and impotence [5,11]. For stuttering priapism new antisickling agents, calcium channel blockers (diltiazem) and vasodilators are safe and more appropriate than diethylstilbestrol which was formerly widely used.

Major priapism crisis required emergency exchange transfusion, verapamil with good hydration and major analgesics. If it does not resolve in less than 48 hours, surgical intervention to decompress the penis is warranted, preferably through a shunting procedure that has the best chance to keep potency intact (Winter shunt) and possibly using intracorporeal pressure and blood gas monitoring [11]. Other genito-urinary disorders in SCA are renal medullary carcinomas and testicular infarction [12].

CONCLUSION

Although the main management of sickle cell anemia is medical, affected individuals have multiple surgical problems which pose a formidable diagnostic challenge. Careful evaluation of patients is extremely important to select cases requiring surgery.

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