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Many neurological disorders in childhood are associated with altered structure and function in other systems. Blood diseases are among the multisystem diseases which have profound implications for the nervous system. Nervous system involvement may be the presenting feature of some hematologic problems. Moreover,neurological complications are the most serious and life threatening sequelae of many blood diseases to the extent that they represent the leading causes of death from these diseases. The prognosis ofuthese diseases usually gets worse when the C.N.S. is involved.

Investigations of an obscure neurological disease in childhood should include searching for an underlying hematologic disorder. On the other hand,nervous system involvement has to be suspected when the pathient,who suffers from a blood disease,develops any neurologic symptom. These investigations should include a plain X-ray on the skull and a fundus examination. It may be necessary in some cases to do an EMG record to detect muscle or nerve lesions,an EEG or a C.S.F examination for meningeal involvement. Angiography of cerebral blood vessls was,untill recently,the procedure of choice to diagnose and localize intracranial hemorrhage or vascular occlusion. Now CAT scanning of the brain,being non-

invasive, less hazardous and more informative, is the diagnostic procedure of choice.

In severe anemia, regardless of its cause, the patient complains of manifestations of brain anoxia in the form of headache, drowziness, vertigo, tinnitus, lack of concentration ability, restlessness and muscle weakness.

Iron deficiency anemia is by far the most common hematologic disease of infancy and childhood. The cause of weakness, irritability, anorexia and impaired intellectual functions that complicate severe cases, is not yet clearly identified. These neurological manifestations are usually reversible. They are corrected few days after institution of iron therapy even before correction of the blood picture.

Pernicious anemia secondary to vitamin B_{12} deficiency is a rare disease of childhood. Neurological manifestations of the disease are detected in up to 90% of untreated cases and are not related to the degree of anemia present. Vitamin B_{12} deficiency results in the syndrome of subacute combined degeneration of the spinal cord. The clinical features are caused by degeneration of the posterior column, pyramidal tract, cranial nerves and less commonly the lateral spinothalamic tract. Parenteral vitamin B_{12} therapy is usually follwed by a good hematological and neurological response in most cases. Complete recovery or at least arrest of progression of the

neurological symptoms is commonly achieved. Folic acid administration,without giving vitamin B_{12},usually aggravate the nervous manifestations of pernicious anemia although it can produce a good hematologic response .

Megaloblastic anemia which results from folic acid deficiency may infrequently affect the nervous system in a simillar way to vitamin B_{12} deficiency. Oral folic acid therapy is usually sufficient to correct the condition.

Sickling hemoglobinopathies,including hemoglobin SS and hemoglobin SC diseases,are known to affect the nervous system in about 25% of cases. The pathological effects on the central nervous system are mostly vascular in the form of thrombosis and occlusion of big and small vessels. Massive intracranial hemorrhage and cerebral embolism are rare but very serious complications. Meningitis,commonly with pneumococcal septicemia,is a common and often lethal complication . No therapy is needed for patients with sickling hemoglobino pathies except during acute episodes or crises. Appropriate antibiotic therapy is given to control bacterial infections. Pneumococcal vaccination and prophylactic pnicillin administration are recommended by many authers. Transfusions of packed red cells or partial exchange transfusions may be needed to reduce the proportion of sickling hemoglobin and sickle cells in severe crisis. Surgical evacuation of intracranial

hematomata may be considered in some cases.

Cooley's anemia,which is the commonest variety of thalassemia,may be infrquently accompanied by some neurologic symptoms. Myopathic syndromes,spinal cord compression by the hematopoietic tissue and pneumoccal meningitis after splenectomy were reported in a few cases. Wasi et al.(1978) have described a syndrome of hypertension,convulsions,cerebral hemorrhage and severe headache in thalassemic patients after repeated blood transfusions.

Polycythemia,particularly the primary type,commonly presents with neurologic symptoms secondary to the decreased C.B.F. and vascular thrombosis. It is a rare problem in children and experience in treatment is too small to allow recommonding specific therapeutic lines other than repeated phlebotomies with replacement of blood by plasma or saline solutions

The frequency of neurological involvement in hemophiliacs ranges from 2-15%. Intracranial and intraspinal hemorrhage were reported with severe deficiency of coagulation factors even without a preceding trauma.

Peripheral nerve lesions,particularly involving the femoral,may occur in hemophilic patients due to traction or compression by a hematoma. Muscle abnormalities were detected in almost all cases of hemophilia after EMG examination. This may be attributed to intramuscular,intraneural or intraarti-

cular hemorrhages with prolonged immobilization. An associating myopathic abnormality in the X-chromosome,adjacent to the locus of hemophilia gene,may be the underlying cause in some cases. Replacement therapy with the deficient factor is the cornerstone in the treatment of bleeding episodes in hemophilic patients. It is recommended to start this type of treatment immediately when bleeding is suspected,not waiting for investigatory documentation. Prompt,agressive and prolonged replacement therapy has to be started before performing any futher investigations for hemophiliacs with a suspected intracranial hemorrhage for fear of their high mortality rate. The necessity to maintain a blood level of the deficient factor of 30-50% for 10-14 days has been stressed by some authors.

Although the complication of intracranial hemorrhage is rare,it must be suspected in patients with idiopathic thrombocytopenic purpura who develop any C.N.S. symptoms because it is the leading cause of death from the disease. Management of these cases should include measures for lowering the intracranial pressure. If the occurence of intracranial hemorrhage is documented by C.T. scanning and there is no rapid spontaneous clinical improvement,emergency splenectomy is indicated. There is usually no need for platelet transfusion except if a neurosugical intervention is necessary and splenectomy fails to raise the platelet count significantly.

Neurologic signs,with variable degrees of severity were noted in about 50% of cases of hemolytic uremic syndrome (H.U.S.). Several mechanisms,including microangiopathic occlusive lesions,were postulated to describe the mechanism of C.N.S. involvement. Treatment is essentially supportive and symptomatic aiming at providing adequate time for spontaneous recovery of renal function and resolution of the microangiopathy. Exchange transfusion and vitamin E therapy were reported to be benifical in some cases.

It is difficult to differentiate clinically between HUS. and TTP. (thrombotic thrombocytopenic purpura). The latter is a rare and serious disease that almost always affects the brain. No proper treatment regimens are yet settled.

Dissiminated intravascular coagulation,on the other hand rarely affect the brain. Treatment is based on correcting the process that has initiated the disease to stop its progression.

The leukemias are the most common form of childhood cancer. Extramedullary leukemia is common in the C.N.S. as well as in the testes during hematologic remissions. The most frequently encountered neurological problems in a leukemic child have been drug toxicity including methotrexate-radiation leucoencephalopathy and viral infections that cause meningoencephalitis. Intra-cranial hemorrhage,leukemic infil-

teration of the C.N.S. and other infections may also occur in a leukemic child. The diagnosis is often difficult because each neurologic complication can result from several causes. Treatment of acute leukemia,particularly the ALL which is the commonest variety,has succeeded in prolonging the survival of affected children,thus complications of therapy,particulary the neurologic ones,become more evident. Treatment regimens should include a C.N.S. prophylaxis after induction of a hematologic remission to erradicate the leukemic cell nests hidden in the C.N.S. away from the reach of systemic drugs; thus minimizing the possibility of later relapses. Intrathecal methotrexate and cranial irradiation are given in some centres for C.N.S. prophylaxis.