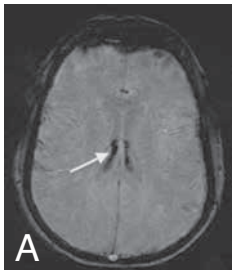
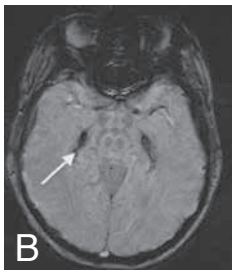
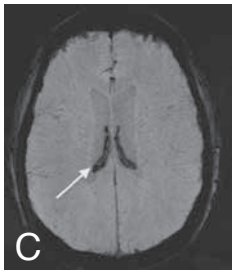
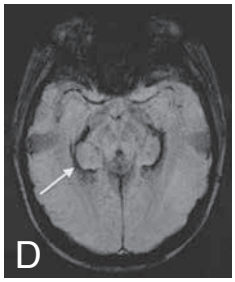
JBR–BTR, 2013, 96: 391.

IMAGES IN CLINICAL RADIOLOGY









Transfusional iron overload presenting as choroid plexus hemosiderosis

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Case 1. A 16-year-old female patient with a history of Diamond-Blackfan Anemia (DBA) treated with repeated transfusions every 3-4 weeks and chelation therapy since infancy presents with transient syncope. Her symptoms progress to include headache, dizziness, and numbness of her right arm, right side of her face, lips, and tongue. MRI of the brain was performed, demonstrating a small frontal subarachnoid hemorrhage (not shown) and dark signal intensity throughout the choroid plexus (Fig. A, B) on susceptibility-weighted imaging (SWI).

Case 2. A previously healthy 22-year-old female was found to have profound anemia and thrombocytopenia. She was transferred to our institution and acute lymphocytic leukemia (ALL) was diagnosed. Regular transfusions were given as part of treatment regimen with the first transfusion 6 months ago. Brain MRI showed "blooming" artifact is visible in the lateral ventricles on SWI sequences, indicating iron deposits of the choroid plexus (Fig. C, D).

Comments

Many clinical conditions include transfusion of red cell concentrates in their treatment with the purpose of alleviating insufficient supply of oxygen to tissues, low concentration of hemoglobin, and/or reduced oxygen carrying capacity and inadequate physiological mechanisms of compensation (1).

The human body has no active mechanisms for the excretion of iron. In each transfusion the patient receives iron in excess (approximately 250 mg) which gradually accumulates in many tissues causing morbidity. When ferritin is trapped in lysosomal membranes it is termed hemosiderin which is an abnormal and insoluble form of iron (2).

Hemosiderosis or iron overload is a pathological condition characterized by deposition of excess iron (hemosiderin) in body tissues. Hemosiderin initially accumulates in the reticuloendothelial system (spleen, bone marrow, and liver) but when these are saturated, deposition occurs in normal body tissues (3). Central nervous system (CNS) iron deposition may involve the choroid plexus, pituitary gland, cortical surfaces, and occasionally the basal ganglia. The choroid plexus is affected first due to its role in brain iron homeostasis and its absence of a blood brain barrier. This combination along with its high blood supply and high surface area increases the uptake of some nutrients such as iron.

CNS iron deposition tends to be asymptomatic except in the pituitary where it results in hypopituitarism. There is no cure for iron overload but early treatment with chelating agents in superficial siderosis is promising (4, 5).

DBA is a rare, genetically heterogeneous, macrocytic anemia presenting early in infancy. Its incidence is approximately 1: 100,000 live births. Laboratory findings include erythroid aplasia, reticulocytopenia, and possible mutation in the ribosomal gene for protein S19. Corticosteroids are first line therapy, although patients refractory to standard treatment necessitate regular blood transfusions as in our case. As a result, many patients with DBA will develop hemosiderosis due to iron overload (4).

ALL is a highly aggressive neoplasm of precursor cells committed to the B-cell or T-cell lineage and represents nearly 25% of all childhood cancers. Chemotherapy is utilized as a first line treatment to reduce the burden of disease as quickly as possible and to restore normal bone marrow function. Packed RCC transfusions are used as needed during treatment (5).

SWI is a high-spatial-resolution 3D gradient-echo MRI sequence which is particularly sensitive to compounds such as blood products, non-heme iron, and calcium that distort the local magnetic field homogeneity producing a "blooming" artifact (7). It is possible that in patients suspected of having CNS hemosiderosis SWI should be the imaging method of choice.

Presence of cerebral hemosiderosis is a rare condition, but may alter systemic treatment of the underlying disease. Therefore it is important to detect it and, if possible, set up therapies focused in avoiding potential CNS complications.

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