

Figure 1: Plain CT scan image showing hyperdense area in the right lentiform nucleus with surrounding ill-defined hypodense area

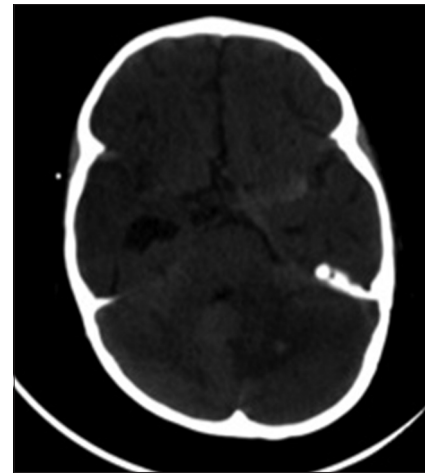


Figure 2: Plain CT scan image showing diffuse ill-defined hypodense area in the left cerebellar region, with hyperdense focus within it, causing compression and displacement of the fourth ventricle. Dilatation of the contralateral temporal horn of the lateral ventricle is noted

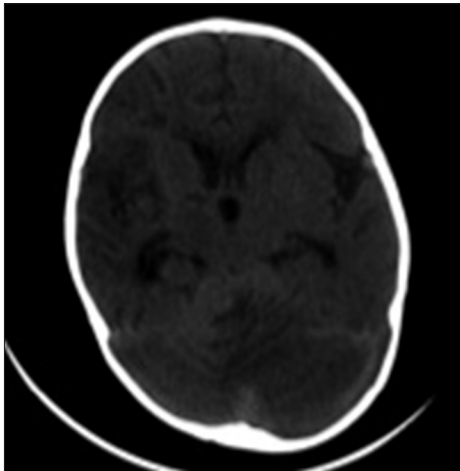


Figure 3: Hyperdensity seen in the region of superior sagittal sinus and left transverse sinus

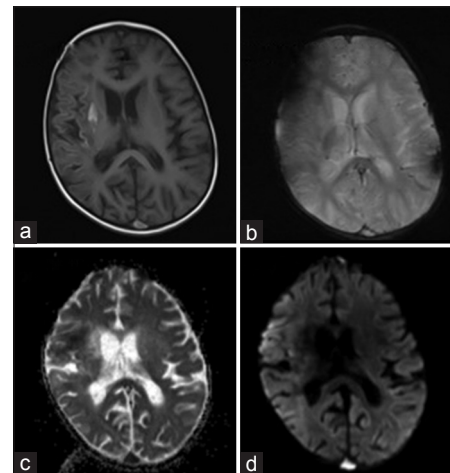


Figure 4: Hemorrhagic infarct in right lentiform nucleus (subacute). (a) Axial T1WI shows a hyperintense area in the right lentiform nucleus. (b) Lesion does not bloom on gradient sequence. (c) On ADC lesion appears bright. (d) No restriction seen on diffusion sequence

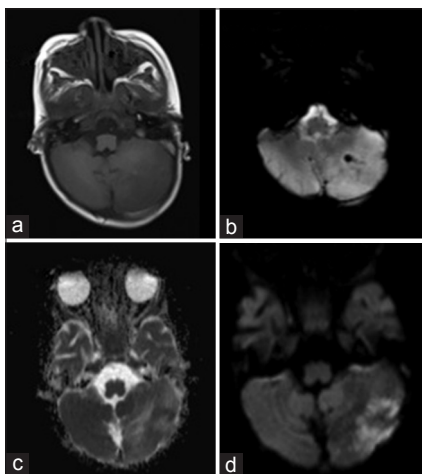


Figure 5: Hemorrhagic infarct in left cerebellar hemisphere. (a) Axial T1WI shows ill-defined hypointense area with few hyperintense foci in the left cerebellar hemisphere. Also hyperintensity is noted in the region of left transverse and sigmoid venous sinus. (b) On gradient sequence, few central areas of blooming are noted. (c) On ADC lesion appears hypointense. (d) Restriction is noted at the corresponding level on diffusion sequence

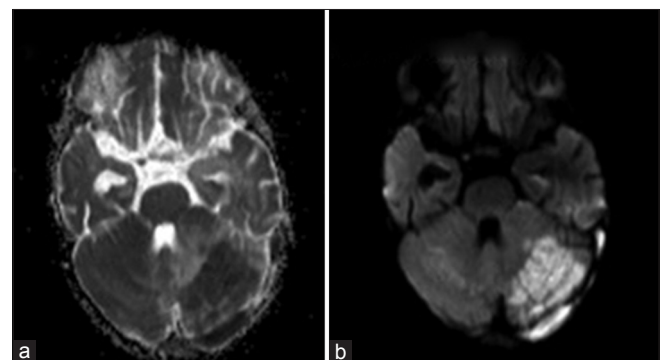


Figure 6: Hemorrhagic infarct in left cerebellar hemisphere. (a) ADC image cranial to previous sections shows diffuse hypointense area in the left cerebellar hemisphere. (b) Restriction is noted at the corresponding level on diffusion sequence

Stroke due to cerebral arterial thrombosis has been attributed to the secondary erythrocytosis of cyanotic heart disease, which is pathologically different from primary erythrocytosis of polycythemia vera. An association between elevated hematocrit and thrombosis has been established in polycythemia vera. However, no association has been established between elevated hematocrit and secondary erythrocytosis in case of cyanotic heart disease.^[7]

Another cause of cerebral infarction in children less than 4 years of age is thrombosis of intracranial dural sinuses or cerebral veins usually in association with iron-deficiency anemia. Superior sagittal sinus, transverse sinus, great vein of Galen, and meningeal veins are the common sites of thrombosis in decreasing order of precedence. Intracranial hypertension may be the only manifestation if the thrombosis is limited to superior sagittal sinus or transverse sinus. Partial or complete recovery is possible even with a severe initial presentation emphasizing the need for early diagnosis and treatment; CT scan is often the first investigation performed on an emergency basis.^[8]

Unenhanced CT scan demonstrates venous thrombosis as linear hyperdensities in the expected locations of the dural venous sinuses and cortical veins. As the thrombus becomes less dense, contrast-enhanced CT scan demonstrates the “empty delta” sign, that is, a filling defect in the posterior part of the sagittal sinus.

Increased attenuation of cerebral veins and sinuses is also noted in cases of polycythemia. New and Aronow^[9] discovered a linear relationship between CT attenuation values and the hematocrit of whole blood preparations. The CT attenuation of hemoglobin was found to be largely due to its protein content. The iron in blood contributed only about 7% of the total attenuation. They also demonstrated that the attenuation of retracted clot was approximately 90%, twice the attenuation of normal intravascular blood (hematocrit level of 43).

Thus, polycythemia is not only a risk factor for development of venous thrombosis, but also a mimicker of the same and if these two conditions coexist, as in the case of Tetralogy of Fallot, the differentiation between the two is only possible by contrast-enhanced CT scan or on MRI.^[5]

In our case, child has extensive venous thrombosis involving the superior sagittal sinus, left-sided venous sinuses, and the left internal jugular vein leading to multiple hemorrhagic infarcts [Figure 7]. Nonvisualization of right internal carotid artery from its cervical portion can be attributed to embolic phenomenon [Figures 8 and 9]. PCV was 46% – mildly elevated level. Hyperdensity in the region of dural venous sinuses was confirmed to be thrombosis on MR venography.

Various options for treatment in children with venous sinus thrombosis include standard or low molecular weight heparin

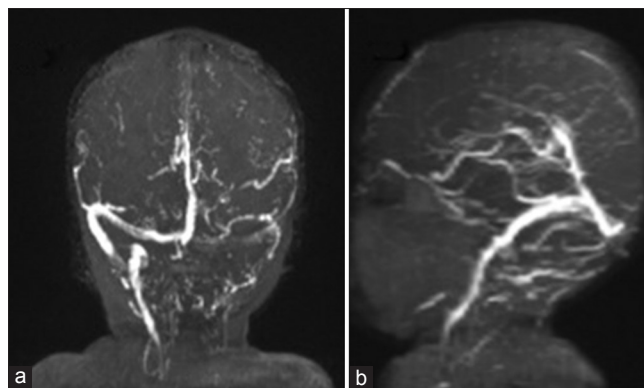


Figure 7: Venous sinus thrombosis. (a) Coronal, (b) Sagittal two-dimensional TOF images demonstrating thrombosis of superior sagittal sinus, left transverse sinus, left sigmoid sinus, and left internal jugular vein

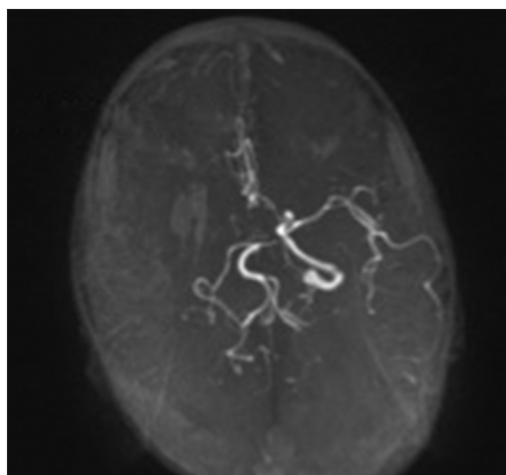


Figure 8: Three-dimensional TOF image depicting nonvisualization of right internal carotid artery and its branches

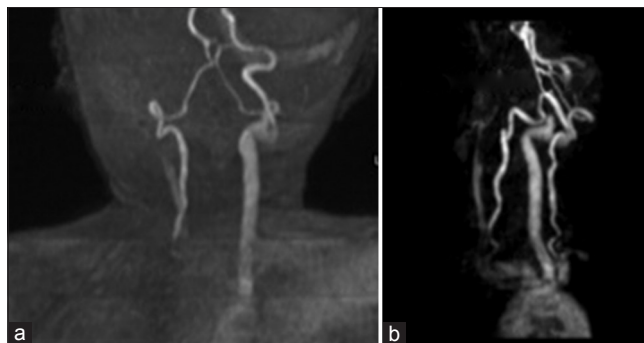


Figure 9: (a) Coronal MIP3D TOF image of neck vessels and (b) three-dimensional TOF image of aortic arch and its branches show nonvisualization of the cervical portion of the right internal carotid artery

for 7-10 days, followed by oral anticoagulants for 3-6 months. Though anticoagulation is controversial in children, treatment with heparin is probably safe and beneficial for these children, even for those with intracranial hemorrhage.^[8]

The clinical practice to phlebotomize patients with polycythemia in order to maintain hematocrit more than

65% has been long followed. Alternative treatment is volume replacement and low-dose iron therapy as repeated phlebotomy can cause iron deficiency leading to microcytic erythrocytes which increases the whole blood viscosity ultimately increasing rather than decreasing the risk for stroke.^[7]

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