Vein of Galen Malformation

A 14 year old male presented for gradual onset progressive diminution of vision right eye and ipsilateral headache of one year duration.

There was no history of seizures, diabetes mellitus or any neurological deficit in the past. No family member had any neurological disease.

Examination revealed no vision in the right eye and 6/6 vision the left eye. Fundoscopy revealed optic atrophy of the right eye. BP was normal, general and systemic examination showed no abnormality. Biochemical parameters were normal. X ray skull lateral view showed thin rim of curvilinear calcification in the supra-sellar region. MRI brain showed a well circumscribed area of flow void due to dilated vein of Galen. MR venography confirmed it to be vein of Galen malformation. MR angiography revealed multiple arterial collaterals in the territory of anterior, middle and posterior cerebral arteries. Hyperintense signals on T2 weighted image and absence of flow void were noted in the region of Torculii Herophilus suggestive of thrombosis. Transcranial Doppler of brain revealed an anechoic lesion in the posterior aspect of brain above the cerebellum.

Vein of Galen malformation is due persistence of embryonic type of circulation. It is a rare form of cavernous malformation of brain and constitutes 1% of all intracranial vascular malformations. Less than 200 cases are reported in world literature. They are more often seen in neonates and young children and cause hydrocephalus, seizures, cognitive dysfunction, focal neurological deficit or cardiac failure.

Our patient had optic nerve involvement on the right side due to either compression by a dilated collateral vessels or possibly due to ischaemia (steal effect).

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