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INTRACRANIAL HYPERTENSION WITHOUT CVST IN APLA SYNDROME: AN UNIQUE ASSOCIATION

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BACKGROUND: Antiphospholipid antibody (APLA) syndrome is an autoimmune disorder predisposing to thrombotic complications affecting CNS either by arterial vasooclusion or venous thrombosis. Cerebral venous sinus thrombosis (CVST) secondarily cause raised intracranial pressure (ICP). However intracranial hypertenion without evidence of CVST is rare entity. Here we present two cases of elevated ICP with absence of identifiable CVST.

CASE SUMMARY: Case 1,28 year female had a 2 months history of holocranial headache followed by bilateral painless vision loss reaching lack of light perception over 20 days. CSF opening pressure was elevated . Fundoscopy showed bilateral grade 4 papilledema . MRI revealed partially empty sella with bilateral optic nerve tortuosity Idiopathic intracranial hypertension (IIH) was dignosed. With acetazolamide there was complete resolution of the clinical and radiological abnormalities. 5 months later she presented with acute onset right sided hemiparesis. MRI was suggestive of acute left MCA infarct.MR venogram was normal. APLA came positive with high titres of Anti cardiolipin and Beta 2 glycoprotein both IgG and IgM. Case 2, 23 year female presented with headache and diplopia of 2 months duration. CSF pressure was elevated and Grade 3 papilledema was seen. MRI showed bilateral optic nerve hyperintensities with nerve head protrusion with normal MRV. APLA profile showed elevated beta 2 glycoprotein IgG and IgA.

CONCLUSION: This is an important non thrombotic complication of APLA syndrome and requires further large scale study for insight into the pathogenesis and early recognition to avoid future complications.