LETTER TO EDITOR

BILATERAL HEMORRHAGIC CEREBELLAR INFARCT IN PRIMARY ANTIPHOSPHOLIPID SYNDROME

Ischemic strokes in children are rare[1,2] and posterior circulation strokes are rarer.[3] Hemorrhagic infarcts of the brain have not been reported in primary antiphospholipid syndrome (APLA). We document the rare instance of bilateral hemorrhagic cerebellar infarction with pontine infarction due to APLA syndrome.

A 13-year-old boy presented with bitemporal headache and developed projectile vomiting, became drowsy, unresponsive and admitted to the Intensive Care within 2 h.

Neurological examination revealed a Glasgow Coma Scale of 4/15. Pupils were equal and reactive to light. Fundi were normal. Plantars were flexor with normal tendon reflexes. The child developed decerebrate rigidity and respiratory arrest requiring endotracheal intubation.

Computerized Tomography (CT) of the Brain showed bilateral hypodensities in the cerebellar hemispheres with central hyperdensity, left more than right [Figure 1].

T1W magnetic resonance imaging (MRI) showed areas of hyperintensities in the posterior cerebellar hemispheres bilaterally suggestive of hemorrhage [Figure 2]. T2W image showed hyperintensity in both the cerebellar and pontine regions suggestive of infarction [Figure 3]. The fluid attenuation inversion recovery (FLAIR) sequence also showed hyperintensities in the above regions suggestive of infarction [Figure 4].

MR Angiography showed enlargement of the proximal portion of the basilar artery with high signal intensity on T1 weighted image suggestive of basilar artery thrombosis with normally paired posterior inferior cerebellar arteries and no evidence of dissection.

Cardiac work up including Transesophageal echocardiography, X-ray of cervical spine and metabolic workup were normal. Hematologic work up including Homocysteine, Antithrombin III, Protein C, Protein S, Factor V Leiden, sticky platelet syndrome were normal and Rheumatologic work up including ANA and Lupus Anticoagulant (LAC) were negative. Anticardiolipin antibody IgG was 11.4 GPL units/ml (Normal less than ten) and IgM was 38.1 MPL units/ml (Normal less than ten). This was repeated 6 weeks after the episode that was positive in rising titers, IgM was 51.1 MPL units/ml and IgG was 14.2 GPL units/ml.

In children, ischemic stroke is most commonly reported in the distribution of the middle cerebral artery. Posterior cerebral and vertebrobasilar involvement have been distinctly rare.[3] Children unlike adults have some unusually common risk factors for ischemic stroke like infection, trauma, cardiac disease, migraine, sickle cell anemia, and prothrombotic disorders.[4]

Our patient was evaluated for all the above said risk factors and based on the clinical and laboratory findings a definite diagnosis of APLA syndrome was made in our patient.[5] Aspirin was withheld in the initial 2 weeks because of the hemorrhagic nature of the infarct, but after 2 weeks the patient was started on low dose aspirin. At discharge, the boy had intact higher functions with minimal cerebellar signs.

Prolonged anticoagulation is mandatory in such patients. Warfarin and aspirin have been tried. However, differences of opinion exist on the duration of therapy. Some workers recommend till APLA has been negative for 4–6 months while others suggest life-long therapy. Although rare, this case report highlights the need for emergency room physicians to consider the possibility of Posterior circulation stroke as a cause for
unconsciousness in selected subgroups of children.

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REFERENCES

MYXOID ADRENAL CORTICAL CARCINOMA – A RARE VARIANT OF ADRENOCORTICAL CARCINOMA

Dear Sir,

Adrenocortical carcinomas are malignant neoplasms of the adrenal cortex generally affecting patients in their fourth and fifth decades of life. Myxoid change is a very rare phenomenon in adrenocortical carcinoma, and only 11 similar cases have been reported to date. Myxoid changes in adrenocortical neoplasm can be present in adrenocortical adenomas also.[1]

A 49-year-old male while being investigated for hypertension was found to have right adrenal tumour on ultrasonography. Physical examination was negative. There was no hypokalemia and the metabolic workup for pheochromocytoma was negative. Computerized tomography scan revealed heterodense right adrenal tumour of size 6 x 5 cm. There was no evidence of invasion to adjacent structures. Right adrenalectomy was performed. The tumour was well encapsulated and resected without any difficulty. Cut section demonstrated circumscribed tumour with uniform myxoid appearance [Figure 1]. On histopathology examination there were features of malignancy like necrosis, vascular invasion, 4–5 mitotic figures/high power field. The tumour cells showed a pseudo glandular pattern with myxoid material inside [Figure 2]. Staining done with mucicarmine and Per-iodic acid Schiff (PAS) showed focal staining for myxoid matrix [Figure 3]. The patient is alive after a follow up period of 1 year with out any evidence of local recurrence and metastasis. Myxoid adrenocortical carcinoma is a rare variant of adrenocortical carcinoma. The presence of myxoid changes in adrenocortical neoplasms usually raises the possibility of malignancy.[1] Tang et al first described this variety in 1979.[2] Myxoid changes have also been reported with adrenal adenoma and these were mostly metabolically normal.[3] The differentiation of benign and malignant tumours can be made by presence of necrosis, vascular invasion, capsular invasion and greater than three mitosis per high power field.[3] The recent literature is replete with articles evaluating the potential role of growth factors, markers of proliferation (Ki 67 and MIB), tumour suppressor genes (p53 Rb-1 and p27) and apoptotic regulators (bcl-2) in differentiating adenoma and carcinoma.[4] Some of these may have prognostic value also.[4] The histochemical stains done are Alcian Blue, PAS, Mucicarmine. The histochemical profile of the myxoid material in our study is consistent with that of previous reports.[2,5] In vast majority of the cases immunohistochemical staining shows vimentin, synaptophysin and inhibin positivity, which is typical of adrenocortical neoplasms.[1] The differential diagnosis of myxoid tumours in retroperitoneum includes chordoma, myxoma, lipoma, liposarcoma, benign and malignant nerve sheath tumours.[2,5] The 5 years survival rate for malignant myxoid adrenocortical tumour is 50% while that for the adenomas it is 100%.[1] The common sites of metastasis are liver and lungs. Local recurrences have also been reported in 2/10 cases reported previously.[1]

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