Brief Communication

Microprolactinoma with visual field defect: An unsuspected etiology

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ABSTRACT

Microprolactinomas present usually with menstrual disturbance and galactorrhoea. The presence of visual field defects is not expected in these patients as the tumor does not involve structures outside the sella. Visual field defects in a case of microprolactinoma confuse the clinician and warrant extensive search for an alternate etiology. We present a young lady with microprolactinoma and visual field defects. Etiological work-up revealed a diagnosis of idiopathic intracranial hypertension (IIH) associated with microprolactinoma. Treatment with Acetazolamide and Cabergoline completely resolved the clinical symptoms and visual defects. The unusual occurrence of IIH in a case of microprolactinoma as the cause of visual field defect is highlighted in our case report.

Key words: Diopathic intracranial hypertension, microprolactinoma, visual field defect

Introduction

Prolactin-secreting pituitary adenoma (prolactinoma) usually presents with local and systemic manifestations. The local features include features of raised intra-cranial tension and visual field defects in patients with macroprolactinoma.[1] Microprolactinoma usually presents with galactorrhoea and menstrual disturbances in females and sub-fertility in males. Microprolactinoma does not lead to visual field defects as the optic chiasm is located outside the sella. Idiopathic intracranial hypertension (IIH), previously known as pseudotumor cerebri or benign intra-cranial hypertension, is a condition characterized by increased intra-cranial pressure without a mass lesion or hydrocephalus.[2] These patients have normal composition of cerebrospinal fluid (CSF) and present with headache and visual field defects. IIH is reported to be associated with obesity and a variety of diseases and drugs. The association of an IIH with microprolactinoma has not been reported widely in published literature.[3] We report a young female patient with a microprolactinoma who had a persistent headache and visual deterioration despite adequate management of the prolactinoma. Further detailed work-up revealed the association of IIH and appropriate therapy of the same resulted in complete improvement of the vision.

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Case Report

A 32-year-old female patient presented to our department with history of headache, oligomenorrhea, and galactorrhoea for 2 years duration. The headache was located in the frontotemporal region with maximal intensity during the morning hours. She denied a history of vomiting, diplopia, or blurring of vision. She gave birth to 2 children with last childbirth about 7 years back. There was no history to suggest thyroid dysfunction, systemic or psychiatric ailment, and drug intake. Examination revealed normal vital parameters with a body mass index of 21.2 kg/m^2 and there was no evidence of goiter. Her visual field examination by confrontation tests was apparently normal and she had no neurological deficit. Fundus examination showed mild pallor and was considered normal. Hormonal profile revealed hyperprolactinemia (serum prolactin – 1228 ng/mL) with

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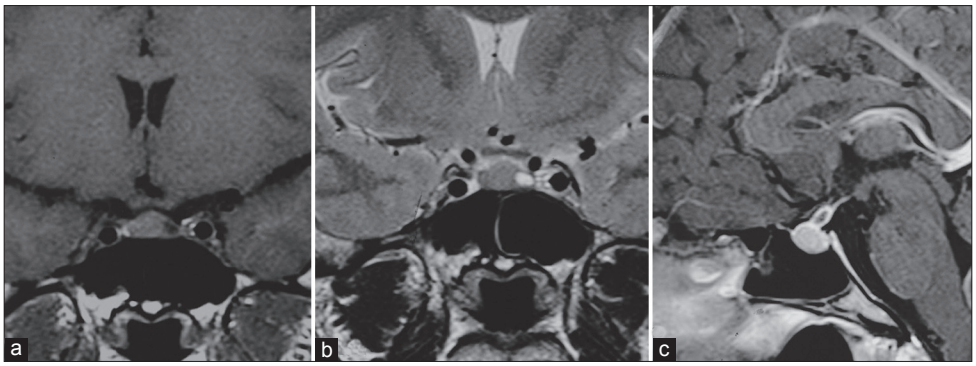

Figure 1: MRI showing pituitary microadenoma (a, b) and stalk thickening (c)

normal thyroid and gonadal axes evaluation. Dynamic and contrast enhanced MRI study of the sella and pituitary gland showed well-defined intra-sellar mass of 0.8 × 0.4 × 0.6 cm^3 over the left side of the pituitary gland without any supra-sellar extension and not in contact with the optic chiasm. There is mild thickening of the pituitary stalk with homogenous hyper-intensity in post-contrast images [Figure 1a-c].

She was diagnosed as a case of microprolactinoma and managed with 0.5 mg of cabergoline twice weekly. She showed clinical improvement in the form of regular menses and cessation of galactorrhoea. Her headache did not improve as we had expected and we continued her with cabergoline for another 6 months. Her repeat prolactin was normal and she continued to complain of headache and occasional blurring of vision. In view of persisting symptoms, a repeat neuro-imaging was done which revealed pituitary microadenoma (0.6 × 0.5 cm^2) as described previously. Visual field examination revealed marked constriction of peripheral vision [Figure 2a] and bilateral early papilledema. Evaluation of the cortical venous sinuses by magnetic resonance venography revealed all patent sinuses. She was considered to have intra-cranial hypertension of obscure etiology and a guarded lumbar puncture was done after giving mannitol and acetazolamide. Her opening CSF pressure was 25-mm Hg and biochemical evaluation of the CSF was normal. She was diagnosed as a case of IIH and treated with acetazolamide along with cabergoline. Her symptoms and headache improved markedly and repeat perimetry showed completely normal visual fields [Figure 2b and c]. During the last follow-up, she is asymptomatic with no headache, visual defect, diplopia, and normal fundus examination.

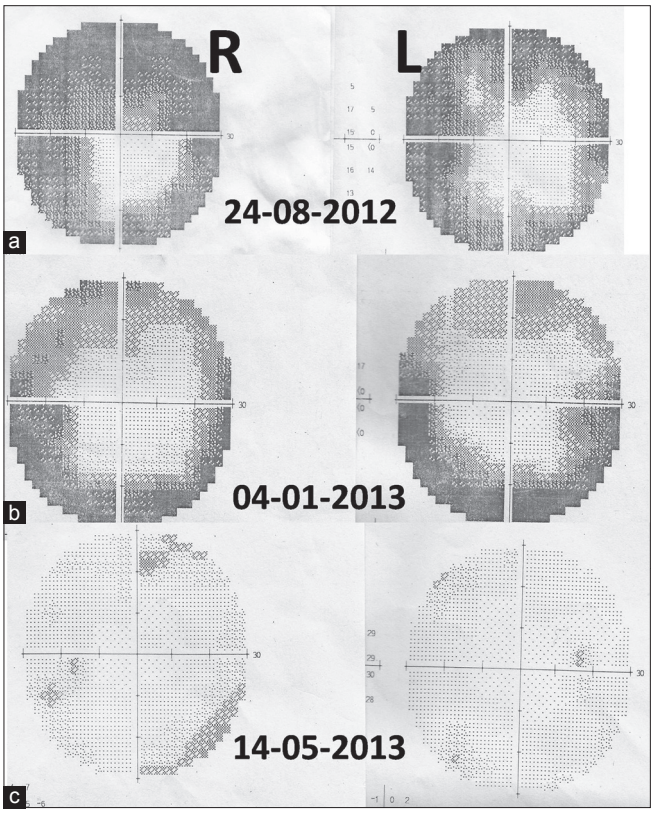

Figure 2: Perimetry showing visual field defects (a) and gradual improvement (b, c)

Discussion

To the best of our knowledge, this is the second case reported in English literature of association between microprolactinoma and idiopathic intra-cranial hypertension. The previous report pertains to a 17-year-old girl with similar association and she required a lumboperitoneal shunt to relieve her symptoms.[3] Our patient had typical features of raised ICP and microprolactinoma. Her initial management with cabergoline resulted in biochemical and radiological improvement with persisting headache. Her visual fields also worsened gradually along with the development of papilledema, prompting us to look for an alternate etiology. She was diagnosed to have IIH as per modified Dandy criteria and acetazolamide resulted in complete normalization of her symptoms and visual fields.[4]

Idiopathic intra-cranial hypertension is an entity without clear pathogenetic mechanisms. Decreased CSF absorption or elevated venous pressure leads to elevation in the ICP and the characteristic features of the condition. The term pseudotumor cerebri is not in vogue to remove the stigma associated with a tumor and benign intra-cranial hypertension is also not popular, due to the understanding that IIH is not benign, if untreated. The visual complications of IIH include field defects and optic atrophy in long standing cases.[5] IIH was reported in patients with pituitary tumors compressing the cavernous sinus leading to decreased CSF absorption.[6] Our patient had normal venography and microadenoma suggesting an alternate mechanism of this association. The endocrine disorders associated with IIH include obesity, Addison disease, hypoparathyroidism, and hypothyroidism.[3]

To conclude, we report the rare association of idiopathic intra-cranial hypertension with microprolactinoma. Our report suggests that IIH should be considered in all patients with unexplained visual field defects along with intra-sellar lesions.

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