A young male with idiopathic intracranial hypertension with essential thrombocythaemia

Palangasinghe DR*, Kularathna OAR2, Herath HMM3 and Pathirana KD4

1Senior Registrar in Medicine, University Medical Unit, Teaching Hospital Karapitiya, Galle, Sri Lanka
2Registrar in Medicine, University Medical Unit, Teaching Hospital Karapitiya, Galle, Sri Lanka
3Senior Lecturer in Medicine, Consultant Physician, Faculty of Medicine, University of Ruhuna, Sri Lanka
4Professor in Medicine, Consultant Neurologist, Faculty of Medicine, University of Ruhuna, Sri Lanka

Abstract

Background: Idiopathic intracranial hypertension (IIH) is a disorder characterized by increased intracranial pressure, normal cerebrospinal fluid (CSF) constituents, without a demonstrable cause for high intracranial pressure. Although commonly associated with obesity, medications like vitamin A, tetracycline and nalidixic acid, it is rarely associated with myeloproliferative disorders. We report a young male from Sri Lanka who developed idiopathic intracranial hypertension associated with essential thrombocythaemia (ET) both which improved with treatment of the latter.

Case presentation: A 24-year-old male presented with persistent frontal headache for two months with transient visual disturbances. On examination he was averagely built, had bilateral optic disc oedema with no ophthalmoplegia, reduced visual acuity or visual field defects. Neuroimaging did not reveal any structural abnormality or venous sinus thrombosis. CSF analysis revealed normal constituents with elevated CSF pressure. Full blood count revealed markedly elevated platelet count (997 x 10^3/mm^3) with normal other cell lines. Janus kinase (JAK)-2 mutation was positive and bone marrow biopsy was in keeping with ET. He was commenced on cytoreductive therapy for ET and aspirin which resulted in resolution of headache and improvement of platelet count to normal values.

Conclusion: IIH could rarely be associated with myeloproliferative disorders like ET. It is important to treat underlying ET which will result in improvement of symptoms secondary to IIH.

Background

Idiopathic intracranial hypertension (IIH) is a disorder which consists of symptoms and signs of increased intracranial pressure, normal cerebrospinal fluid constituents, without a demonstrable cause for high intracranial pressure. Usually they present with headache and visual symptoms. Headache is the most frequent and the starting symptom in more than 80% of patients [1]. While bilateral papilloedema is the most characteristic finding some may develop visual field defects and lateral rectus palsy as well. IIH is associated with many different conditions including medications (Vitamin A, Tetracycline, Nalidixic acid, Nitrofurantoin, NSAIDs), systemic arterial hypertension, polycystic ovarian disease, obesity, pregnancy and menstrual irregularities [1]. However, association of IIH with myeloproliferative disorders is very rare. In the literature, only few cases of idiopathic intracranial hypertension were found to be associated with chronic myeloid leukaemia and essential thrombocythaemia [2,3]. We report a case of idiopathic intracranial hypertension associated with essential thrombocythaemia (ET) in a Sri Lankan young male.

Case Presentation

A 24-year old previously healthy Sri Lankan male presented with a history of moderate to severe persistent headache, predominantly over frontal region for two months. It was not precipitated with coughing, sneezing or straining. It was associated with transient visual obscurations. He denied any history of fever, autonomic symptoms with headache, any altered behaviour or history of head injury. He had no past history of recurrent headaches. He was a non-smoker and denied any substance abuse including alcohol. He was not on any long-term medications.

On clinical examination, he was found to have bilateral disc oedema with absent retinal venous pulsation (Figure 1). But there was no ophthalmoplegia, abnormal pupillary light reflex or visual field defects. Visual acuity was 6/6 in both sides. No other focal neurological signs were detected. Blood pressure was 130/80mmHg. Rest of the clinical examination was unremarkable.

Full blood count showed Haemoglobin–13.4g/dL WBC 8740/mm^3, with normal differential count, Platelets - 997 x 103/mm^3. Magnetic resonance imaging brain (MRI) was normal. Magnetic resonance venogram (MRV) revealed no venous sinus thrombosis. Cerebrospinal fluid manometry revealed increased cerebrospinal fluid pressure (229cm H2O), but normal cerebrospinal fluid constituents (Neutrophils – 0/mm^3, Lymphocytes – 1/mm^3, Red blood cells – 4/mm^3, Protein – 45 g/dL, Glucose – 86 mg/dL, with random blood glucose of – 128 mg/dL). He did not have any evidence of an infection.
Our patient presented with headache and papilloedema with elevated CSF pressure without demonstrable structural abnormality in brain and normal CSF constituents. He also had marked thrombocytosis without any inflammatory disorder, chronic infection or chronic bleeding with positive JAK-2 mutation and evidence of megakaryocyte proliferation in bone marrow. He fulfilled the diagnosis of both IIH and ET. Even though the association of IIH with a myeloproliferative disorder is rare; ET is well known to cause cerebral venous sinus thrombosis [2]. MRI combined with MRV has high sensitivity (97%) and specificity (99%) in detection of dural venous sinus thrombosis [5]. In the absence of venous sinus thrombosis in venogram, there are postulated mechanisms with prothrombotic conditions to develop high intracranial pressure. One hypothesis is elevated platelet count giving rise to intermittent dural sinus blockage [3]. Other possibility is that unrecognized non-occlusive venous sinus thrombosis from MRV that might occlude the CSF drainage [6]. In the literature mixed prospectively and retrospectively investigated cohort studies showed that many prothrombotic conditions other than thrombocytosis are associated with IIH such as the presence of anticardiolipin antibodies, familial deficiency of anti-thrombin iii, polycythemia vera and increased fibrinogen level [6]. Therefore, looking for an associated such condition is of importance in evaluating a patient with IIH. Our patient’s clinical improvement was evident with the resolution of headache with the treatment for two weeks followed by resolution of thrombocytopenia after one month of treatment for ET. This case adds to the literature of association between the IIH and ET. At the same time, it shows the importance of searching for an underlying myeloproliferative disorder in a patient with IIH.

**Consent**

Written informed consent was obtained from the patient for the publication of this case and accompanying images.

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