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Fibromyalgia as A Sole First Presentation of Recurring Idiopathic Intracranial Hypertension in a child. A Case Report and Literature Review

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ABSTRACT

Idiopathic intracranial hypertension (IIH) is a condition characterized by raised intracranial pressure (ICP) with no evidence of brain pathology, also known as pseudo tumor cerebri. Headache and visual obscuration are the most common presenting symptoms for IIH with a frequency of (68-84%) respectively. Other symptoms of IIH include neck pain, back pain, and radicular pain in the arms and legs resulting from increased spinal pressure and forced filling of the spinal nerves with CSF.

Case summary:

We present an 11-year-old boy known to have chronic kidney disease stage II, due to obstructive uropathy. He had a history of Idiopathic intracranial hypertension that was treated completely and condition resolved, then presented with generalized pains and aches without headaches or visual symptoms and found to have IIH for the second time, the symptoms resolved with appropriate treatment.

Conclusion:

Widespread pain and several other characteristics of IIH share similarities with characteristics of fibromyalgia (FM) and chronic fatigue syndrome (CFS), two overlapping chronic pain conditions.

Keywords: Idiopathic intracranial hypertension; Fibromyalgia; Papilledema

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Introduction

Idiopathic intracranial hypertension (IIH) is a well-recognized disease which was first described in 1893 by Heinrich Quinckae as pseudo tumor cerebri syndrome, then was relabeled in 1955 by Foley to be called benign intracranial hypertension [1].

It is defined as increased intracranial pressure without evidence of brain pathology; infectious, obstructive or vascular.

In the 1980s, physicians started to become more aware of vision complication, after which the name of “benign” was changed to the current name idiopathic intracranial hypertension [1].

Multiple theories explained the pathophysiology of IIH and its relation with cerebrospinal fluid (CSF) Considering flow dynamics, as a central theme, both in increased secretion and decreased absorption [2,3]. Other proposed theories in which obesity, corticosteroid use, vitamin A toxicity, hormonal changes, aquaporin and natriuretic peptide are proposed as possible contributors to the development of IIH [2–8].

Friedman and Jacobson [9] set diagnostic criteria for IIH. According to those criteria, IIH can be classified as:

-**Definite:**

When high opening pressure is >28 cm H2O or >25cm H2O if child is not sedated and not obese.

In the presence of either papilledema or abducens nerve palsy.

- **Probable:**

Normal opening pressure in presence of papilloedema

-**Suggestive:**

Raised opening pressure with at least three neuroimaging finding of increased Intracranial Pressure (ICP), when both papilledema and abducens nerve palsy are absent.

Those criteria were revised in 2014 by IIHTT study (Idiopathic Intracranial Hypertension Treatment Trial) making the opening pressure cut off more than 20 cm H2O [10]. Headache and visual obscuration used to be the most common presenting symptoms for IIH (around 68-84%respectively) [10].

Fibromyalgia is a rare condition in pediatrics. It was reported in few cases in adult patients in association with IIH. However, the authors are not aware of this association being described in pediatric age group with fibromyalgia as the presenting manifestation of IIH.

Case Report

An 11 years old male patient, who is known to have chronic kidney disease (stage II) due to obstructive uropathy, presented with history of headache suggestive of increased intracranial pressure. He was diagnosed with idiopathic intracranial hypertension with prior presentation of body aches and headaches, examination revealed papilledema. LP showed high opening pressure reaching up to 59cm H2O. MRI and MRV were unremarkable.

He was treated with Acetazolamide 25mg/kg/day after which his symptoms resolved and his eye examination normalized in few months in repeated examination. Medication was weaned gradually over 6 months until discontinued. He remained in good condition for around four months when he presented with severe muscle pain in his flanks, arms, shoulders and back. This was associated with abdominal pain that interrupted his sleep. Pain was responding partially to over-the-counter analgesics. His condition was associated with low mood and school absences.

Parents sought medical advice multiple times in the emergency room. No visual symptoms nor headache were reported. Physical examination showed an overweight boy with BMI (Body Mass Index) of 26.7 kg/m2. Multiple muscular point of tenderness in his upper and lower body parts. Repeated fundus examination was done by ophthalmologist on weekly bases with pupil dilation. None of them showed papilledema or other abnormalities. Inflammatory markers and rheumatological work up were unremarkable. Lumber, thoracic spine X-rays were within normal. Ultrasound abdomen did not show abnormalities.

As his symptoms were concurrent with COVID-19 restrictions and the diagnosis was challenging, he was referred to psychiatrist for evaluation which revealed no significant disorders. After

one month of recurrent pains, his eyes examination started to show signs of early optic disc swelling. Lumber puncture showed an opening pressure of 48 cm H2O. His symptoms resolved immediately after lumber puncture. Acetazolamide was restarted again. Follow up after 6 weeks of therapy showed complete resolution of his symptoms and normal ophthalmological examination.

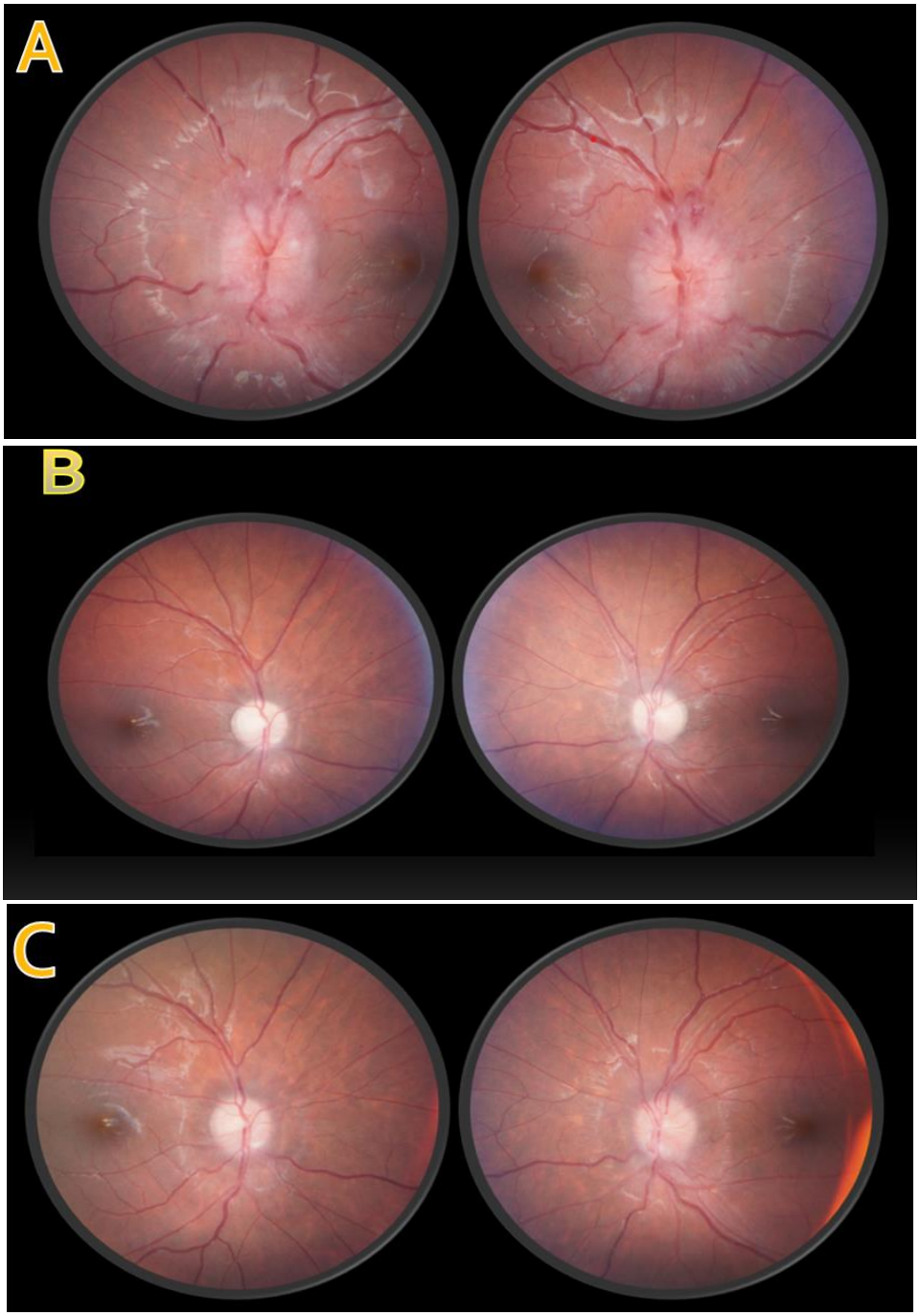

Image A: Fundus examination revealed bilateral papilledema at the presentation
Image B: Fundus examination revealed resolved papilledema in both eyes after the management
Image C: Fundus examination revealed relapsed bilateral papilledema within the second presentation

Discussion

Idiopathic intracranial hypertension (IIH) is commonly present with headache and papilledema. Papilledema is the result of increase of the cerebrospinal fluids (CSF) pressure which in turn causes direct nerve injury. In a similar way, few reports proposed a hypothesis that increase in CSF pressure may affect spinal nerves leading to syndromes such as chronic fatigue syndrome and fibromyalgia [13–16].

A report on the electrodiagnostic studies in 17 adult female patients with fibromyalgia described neurogenic findings in all patients, with a marked difference in myotome abnormalities based on the level of nerve involvement. Needle Electromyography at the sacral levels showed more abnormalities in comparison with lumbar ones. This was attributed to the difference of hydrostatic pressure of CSF in the spinal canal [11].

Hulens et. al. reported 30 adult patients with fibromyalgia or unexplained widespread pains that responded to spinal tapping [15]. The definition of high pressure was CSF opening pressure over 20 CM H2O [16]. Fourteen out of the 30 patients had an opening pressure of more than 20 cm H_2O, with all patients including the normal pressure group showing benefit from the spinal tap. The authors concluded that high intracranial pressure (ICP) is a possible cause of chronic pains and tapping is a possible treatment. In a different report, Hulens et. al. described the overlap between symptoms of patients with fibromyalgia and IIH, especially those without papilledema [12].

We acknowledge that Fibromyalgia diagnostic criteria [17] require the presence of symptoms for 3 months. Despite that our patient presented with less than that period in his second illness, it is probable that his initial presentation is also related to fibromyalgia. Knowing that he received treatment for the first presentation that might mask or alleviate the typical symptoms to fulfill the criteria at that time.

To our knowledge, our patient is the first in pediatric age group who shows increase in ICP together with fibromyalgia-like symptoms without the classical symptoms of IIH. This is also the first report of such presentation recurring after a period of initial treatment. It is not clear whether there is an association with his kidney problem, however the association of IIH with obesity has been well documented.

Further research is needed to evaluate the hypothesis in pediatric patients with non-specific pains and aches. This unusual presentation may lead to delay of diagnosis and treatment of IIH with atypical presentation.

Conclusion

Idiopathic intracranial hypertension (IIH) has a wide variety of symptoms. The link between IIH and Fibromyalgia has been described in adults with fibromyalgia-like presentation of IIH. We describe a patient in the pediatric age group. Idiopathic intracranial hypertension should be considered in patients with fibromyalgia like presentation.

References

[1] Burkett JG, Ailani J. An Up to Date Review of Pseudotumor Cerebri Syndrome. Vol. 18, Current Neurology and Neuroscience Reports. Current Medicine Group LLC 1; 2018.

[2] Cleves-Bayon C. Idiopathic Intracranial Hypertension in Children and Adolescents: An Update. Headache. 2018 Mar 1;58(3):485–93.

[3] Baykan B, Ekizoğlu E, Altiokka Uzun G. An update on the pathophysiology of Idiopathic intracranial hypertension alias pseudotumor cerebri. Agri. 2015 Apr 1;27(2):63–72.

[4] Libien J, Kupersmith MJ, Blaner W, McDermott MP, Gao S, Liu Y, et al. Role of vitamin A metabolism in IIH: Results from the idiopathic intracranial hypertension treatment trial. J Neurol Sci. 2017 Jan 15;372:78–84.

[5] Subramaniam S, Fletcher WA. Obesity and Weight Loss in Idiopathic Intracranial Hypertension: A Narrative Review. Vol. 37, Journal of Neuro-Ophthalmology. Lippincott Williams and Wilkins; 2017. p. 197–205.

[6] Portelli M, Papageorgiou PN. An update on idiopathic intracranial hypertension. Acta Neurochir (Wien). 2017 Mar 1;159(3):491–9.

[7] Sinclair AJ, Ball AK, Burdon MA, Clarke CE, Stewart PM, Curnow SJ, et al. Exploring the pathogenesis of IIH: An inflammatory perspective. J Neuroimmunol. 2008 Sep 15;201–202(C):212–20.

[8] Markey KA, Uldall M, Botfield H, Cato LD, Miah MAL, Hassan-Smith G, et al. Idiopathic intracranial hypertension, hormones, and 11β-hydroxysteroid dehydrogenases. J Pain Res. 2016 Apr 19;9:223–32.

[9] Friedman DI, Jacobson DM. Diagnostic criteria for idiopathic intracranial hypertension. Neurology. 2002 Nov 26;59(10):1492–5.

[10] Wall M, Kupersmith MJ, Kieburtz KD, Corbett JJ, Feldon SE, Friedman DI, et al. The idiopathic intracranial hypertension treatment trial clinical profile at baseline. JAMA Neurol. 2014;71(6): 693–701.

[11] Hulens M, Bruyninckx F, Rasschaert R, Vansant G, De Mulder P, Stalmans I, et al. Electrodi-

agnostic Abnormalities Associated with Fibromyalgia. J Pain Res. 2020;13:737–44.
[12] Hulens M, Rasschaert R, Vansant G, Stalmans I, Bruyninckx F, Dankaerts W. The link between idiopathic intracranial hypertension, fibromyalgia, and chronic fatigue syndrome: exploration of a shared pathophysiology. J Pain Res. 2018;11: 3129–40.
[13] Higgins JNP, Pickard JD, Lever AML. Chronic fatigue syndrome and idiopathic intracranial hypertension: Different manifestations of the same disorder of intracranial pressure? Med Hypotheses. 2017;105:6–9.
[14] Hulens M, Dankaerts W, Stalmans I, Somers A, Vansant G, Rasschaert R, et al. Fibromyalgia and unexplained widespread pain: The idiopathic cerebrospinal pressure dysregulation hypothesis. Med Hypotheses. 2018 Jan;110 (December 2017):150–4.
[15] Hulens M, Rasschaert R, Dankaerts W, Stalmans I, Vansant G, Bruyninckx F. Spinal fluid evacuation may provide temporary relief for patients with unexplained widespread pain and fibromyalgia. Med Hypotheses. 2018 Sep;118 (April):55–8.
[16] Distelmaier F, Sengler U, Messing-Juenger M, Assmann B, Mayatepek E, Rosenbaum T. Pseudotumor cerebri as an important differential diagnosis of papilledema in children. Brain Dev. 2006 Apr;28(3):190–5.
[17] Wolfe F, Clauw DJ, Fitzcharles M-A, Goldenberg DL, Häuser W, Katz RL, et al. 2016 Revisions to the 2010/2011 fibromyalgia diagnostic criteria. Semin Arthritis Rheum. 2016;46(3):319–29.