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Case Report

Pseudotumor cerebri in 11 months old child – A case report with review of literature

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Abstract

Pseudotumor cerebri is defined as a clinical entity where there are signs and symptoms of raised intracranial pressure but the higher mental and neurological functions are not altered. Many times the etiology is unknown. A thorough history and clinical examination is necessary along with exclusion of possible structural causes of intracranial hypertension. Headache, papilledema and visual disturbances are the characteristic symptoms in a child but in a younger infant it becomes all the more difficult to arrive at diagnose. The prognosis is excellent if the offending drug or cause is identified and abated. This case study reported a case of Pseudotumor cerebri (PTC) in 11 months old child.

Key words

Pseudotumor cerebri, Benign intracranial hypertension, Papilledema, Nalidixic acid.

Introduction

Benign intracranial hypertension (ICH) or Pseudotumor cerebri (PTC) is a syndrome characterised by high cerebrospinal fluid pressure without any underlying structural or systemic cause [1]. The presenting symptoms of idiopathic ICH are known to vary with age and there are assertive reports of this entity in the pediatric population. The older children usually have associated headache, neck pain, diplopia, or infrequent visual disturbances and often experience intracranial noises. The diagnosis in younger infant is challenging as they may only have irritability or apathy [2]. Idiopathic intracranial hypertension without papilledema is well-described in adults [3] and is rarely reported in the pediatric population [4]. PTC in 11 months old child without papilledema is highlighted in this case report.

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11 months old male baby was seen in the casualty late night with symptoms of lethargy, recurrent episodes of non projectile vomiting, abdominal pain, fowl smelling semi-loose stools with blood streaks and mucous since the previous day. History revealed that the family had travelled for a pilgrimage and had food from hotels. He was treated for symptoms of the diarrhoeal episode and was on medications elsewhere. He became lethargic since the previous night with recurrent vomiting and had low grade fever for the last 2 days. There was no associated rash, ear discharge, seizures or any trauma. Birth history revealed that he was born to 29 years old mother at 37 weeks gestation via spontaneous vaginal delivery weighing 3.6 kg with normal apgar score. His parents were of non-consanguineously married. He was immunized to date and had normal milestones of development. On general physical examination, baby was tired, crying, at times was very irritable. He had low grade fever (99.2^0F), heart rate 102/minute, and respiratory rate 36/minute, weight 7.36 kg, anterior fontanelle was bulging and prominent with pulsations in the supine as well as in the sitting position. **(Photo - 1A, Photo - 1B)** His oxygen saturation was 96%. All peripheral pulses were equally felt. Blood pressure was 94/56 mm Hg in right upper limb. He was having moderate dehydration. His respiratory effort remained good and was very irritable at times inconsolable even after breastfeeding. Abdominal examination did not reveal any mass in the abdomen and there was no hepatosplenomegaly. Cardiovascular system examination was normal. The child was admitted and his general condition improved with symptomatic treatment given for fever with Paracetamol drops and for vomiting with Domperidone suspension. His dehydration was corrected with intravenous Ringer lactate as he was vomiting. Breast feeding, oral feeds and fluids started subsequently. He did not have any neurological deficits and his fundus examination was normal and there was no papilledema. He had received Nalidixic acid (NA) suspension for the dysentery prior to the admission in our hospital. A diagnosis of PTC, secondary to Nalidixic acid, was considered, but decided to rule out other causes of bulging anterior fontanel including meningitis. The laboratory investigations revealed Hb (12 g/dl), PCV (33.7%), total count (9000/µl), neutrophils (48%), lymphocytes (46%), eosinophils (3.7%), monocytes (2.1%), basophils (0.2%), ESR (31mm/hr), and platelets count (210000/µl) were all found to be normal. Serum Na^+ 133 mEq/l, K^+ 4.4 mEq/l, Cl^- 91 mEq/l, bicarbonate 26 mEq/l, glucose 144 mg/dl, total Ca^{++} 9.0 mg/dl and C Reactive protein was normal (<0.8 mg/dl). NA was not given further. He was started on oral Cefixime suspension 8 mg/kg/day in 2 divided doses along with Zinc sulphate solution 10 mg/day and oral rehydration solution. The baby's clinical condition was followed-up. The bulging anterior fontanel was present for 2 more days and became normal on the 4^{th} day after admission. **(Photo - 1C)** The child was discharged with advice for continuing breast feeding and to follow-up.

Discussion

PTC is a condition of ICH without localizing signs or neurological deficits except for papilledema and normal cerebrospinal fluid constituents with normal intracranial structure [5]. The PTC may be primary PTC (idiopathic ICH) or arise from a detectable secondary cause. PTC in the primary group are generally associated with a normal brain parenchyma and without ventriculo megaly, mass lesion, or underlying infection or malignancy can also cause this type of PTC and also observed in adolescent obese children and

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women [5]. The secondary PTC group causes are multiple and includes causes such as cerebral venous abnormalities, cerebral venous sinus thrombosis. Also a decreased CSF absorption from past intracranial infection or subarachnoid hemorrhage and endocrine disorders such as Addison disease, hypoparathyroidism, Cushing's disease and chronic kidney failure, anemia can cause PTC [2, 3]. There are many medications which can cause PTC and they are birth control pills, cyclosporine, minocycline, NA, nitrofurantoin, phenytoin, sulfa drugs, tamoxifen, tetracycline, vitamin A. Hormones such as human growth hormone, thyroxine (in children), leuprorelin acetate, levonorgestrel, anabolic steroids medical and starting or withdrawal from chronic corticosteroids can cause PTC [6]. The pathogenesis of PTC is a cascade involving cerebral edema, increased cerebral blood volume, and also decreased CSF absorption [1, 7]. For ICH to occur there should be a prolonged elevation of the intracranial pressure, above 200 mm H_2O. In most patients, it manifests as severe headache, papilledema, transient visual disturbances, diplopia, tinnitus, nausea, vomiting, and central nervous system and mental abnormalities, as well as dysfunctions of the cardio-respiratory systems [7]. The presenting symptoms of benign ICH are known to vary with age where by the older children usually have associated headache, neck pain, diplopia, or infrequent visual disturbances and often experience intracranial noises. The diagnosis in younger infant is challenging as they may only have irritability or apathy [2, 7].

A good history taking regarding onset of the signs and symptoms along with a thorough clinical examination is necessary. It is pertinent to look for signs of suspected raised ICH like bulging anterior fontanel in an infant, neurological deficits, positive signs of meningeal irritation and fundoscopy for not missing papilledema help clinch the diagnosis in an irritable child in the setting of PTC [2, 3]. The diagnosis of PTC is based on high degree of clinical suspicion but the tests that may be done include computed tomography (CT) scan of the brain, ophthalmology examination, including visual field testing, magnetic resonance imaging (MRI) of the brain with MR venography and Lumbar puncture for CSF analysis can be done [1]. There are standard revised diagnostic criteria such as modified Dandy criteria for the diagnosis of PTC as is mentioned in the literature review [5, 6]. According to these criteria, LP must be included in the route to diagnose of PTC. In this case, when the NA was withdrawn, the condition improved and thus ignored the invasive procedure such as LP and other imaging (e.g. CT, MRI). However, weight gain or exposures to a substance associated with the PTC are associated with relapse of PTC and there will be recurrent papilledema [3].

In most patients with idiopathic PTC, without papilledema, laboratory and neurologic investigations are normal. The child in this case had bulging anterior fontanel and signs of raised ICH such as irritability and vomiting, but not associated with papilledema as mentioned in other studies [3]. NA, a quinolone is still used in the treatment of acute dysentery, and has been known to cause PTC in infants and young children. The recommended dose is 55 mg/kg/day in 3 divided doses [8]. In this case, the child reported received the drug and developed PTC. Fortunately, no delay in medical intervention excluded the possible serious side effects such as metabolic acidosis, associated with the NA toxicity.

The treatment of PTC involves initially the removal and discontinuation of possible offending medication or hormonal preparation. The management for ICH is by medical treatment, drug therapy, or surgical intervention. Fluid and salt restriction is

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routinely advised. A combined therapy with acetazolamide and furosemide is recommended as an effective first-line method of treating raised intracranial pressure in children with PTC [9]. It is advised not to use corticosteroids as the first choice for treatment and the LP and diuretic therapy should precede its administration. An LP can also help relieve CSF pressure in the brain and prevent vision problems.

CSF shunting procedures namely (lumboperitoneal or ventriculoperitoneal shunt) may be employed, if medical treatment fails. Papilledema should not be missed. When chronic papilledema threatens visual function optic nerve sheath decompression may also be done to prevent vision loss [3]. Weight loss is advised in adults and adolescents with obesity in the primary PTC. PTC disappears on its own within 6 months in some subset of patients. It is found that even with immediate intervention, visual loss can occur [10]. Transient visual abnormalities were very common in a subset of patients with absent papilledema in PTC [11] and so vision has to be monitored on follow-up. The child does not have any deficits.

This case study concluded that awareness of causes of PTC among the pediatricians and treating physicians is very important. Even though there is a risk of vision in a minority of cases, the prognosis in PTC is excellent. PTC has good prognosis, if diagnosed early and correctly with the simple non invasive tools such as proper history taking and clinical examination as the diagnosis is mainly of exclusion of causes.

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References

1. Moffat FL. Pseudotumor cerebri. Can J Neurol Sci, 1978; 5: 431-6.
2. Weig SG. Asymptomatic idiopathic intracranial hypertension in young children. J Child Neurol, 2002; 17: 239-41.
3. Marcelis J, Silberstein SD. Idiopathic intracranial hypertension without papilledema. Arch Neurol, 1991; 48: 392-9.
4. Beri S, Gosalakkal JA, Hussain N, Balky AP, Parepalli S. Idiopathic intracranial hypertension without papilledema. Pediatr Neurol, 2010; 42: 56-8.
5. Friedman DI, Liu GT, Digre KB. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. Neurology, 2013; 81(13): 1159-65.
6. Liguori C, Romigi A, Albanese M, Marciani MG, Placidi F, Friedman D, Digre K, Liu G. Revised diagnostic criteria for the pseudotumor cerebri syndrome in adults and children. Neurology, 2014; 82(19): 1752-3.
7. Kabat AG. Intracranial hypertension. Optom Clin, 1996; 5: 153-79.
8. Riyaz A, Aboobacker CM, Sreelatha PR. Nalidixic acid induced pseudotumour cerebri in children. J Indian Med Assoc, 1998; 96: 308-314.
9. Schoeman JF. Childhood pseudotumor cerebri: Clinical and intracranial pressure response to acetazolamide and furosemide treatment in a case series. J Child Neurol, 1994; 9: 130-4.
10. Kesler A, Gadoth N. Pseudotumor cerebri (PTC--an update). Harefuah, 2002; 141: 297-300, 312.
11. Amacher AL, Spence JD. Spectrum of benign intracranial hypertension in children and adolescents. Childs Nerv Syst, 1985; 1: 81-6.

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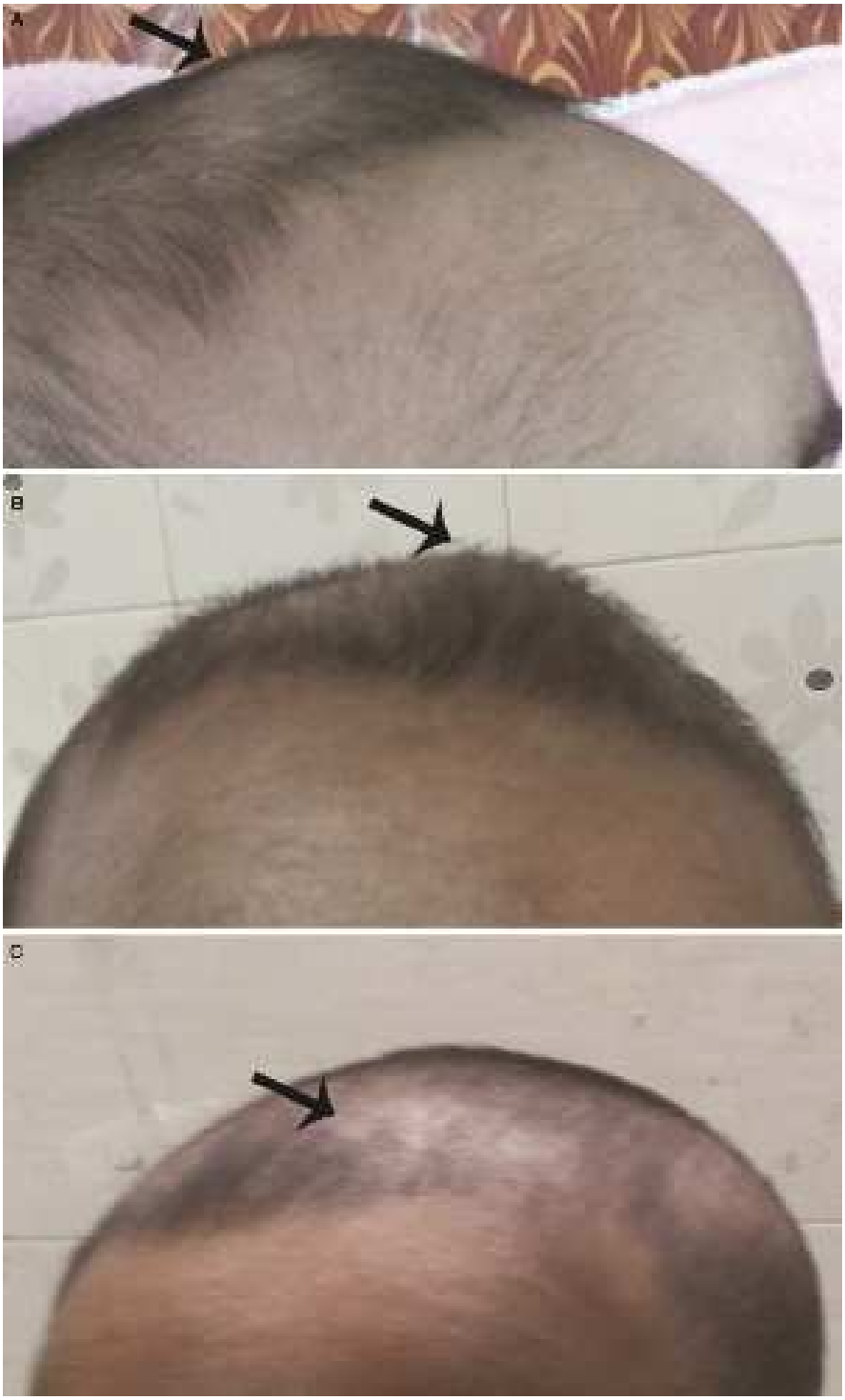
Photo – 1: Child with the bulging anterior fontanel in the **A)** supine; **B)** sitting position and **C)** Anterior fontanel returned to be normal on 3rd hospital day.



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