Rare Cause of Pseudotumor Cerebri in Children

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Abstract
Pseudotumour cerebri may be idiopathic or secondary. Clinicians must take care to exclude secondary causes of raised intracranial pressure in all patients. We report a case that illustrates the importance of asking about dietary intake and supplements when evaluating a patient with pseudotumour cerebri.

Keywords: Pseudotumour cerebri, intoxication, vitamin A, raised intracranial pressure.

Aims

Pseudotumor cerebri (PTC) is a syndrome of elevated intracranial pressure that occurs typically in obese young women and characterized by headache, papilledema, increased intracranial pressure, and occasionally, visual field defects with sixth nerve palsies [1]. Many explanations have been proposed to have been responsible of this syndrome [2].

Pseudotumour cerebri may be idiopathic or secondary. Clinicians must take care to exclude secondary causes of raised intracranial pressure in all patients, particularly in children and teenager with normal body mass index [3].

We report a case for whom ingestion of vitamin A megadose was associated with neurological symptoms of benign intracranial hypertension.

Presentation of Case

An eight-Year-old girl presented with complaints of severe generalised headache that was worse on waking, with vomiting, blurring vision and diplopia. Those symptoms lasted 9 days. The clinical examination was normal apart a bilateral convergent strabismus. Body mass index was 12.5 kg/m² for a normal between 13.2 to 22.9 kg/m² (-2 z-scor). Her visual acuity was 10/10 right and left eyes. Fudus examination showed optic disc swelling bilaterally (stage 1).

Sensory examination showed homonymous horizontal diplopia in primary position and distance vision, maximal in near vision on the left version (left lateral rectus field of action). Ophthalmic examination revealed no cause for optic disc swelling and neurological examination was otherwise normal. Magnetic resonance imaging (MRI) and venography showed no space-occupying lesion, venous thrombosis or stenosis. Full blood count and renal function were normal. Cerebrospinal fluid (CSF) opening pressure was 27 cm H2O, in lateral decubitus position, with normal constituents. The history taking identified an ingestion of a large amount of tuna liver. The dosage of vitamin A (Retinol) in her plasma, nine days later, was elevated to 2.72 μmmol/l (Normal: 0.90-1.20).

Based on all these arguments we diagnosed her with pseudotumor cerebri following a vitamin A overdose. There was no need for an urgent lumboperitoneal shunt. She received a treatment based on acetazolamide 20mg/kg/day.

After 8 weeks of follow-up, the papilloedema had resolved in right eye and regressed in left eye. The diplopia had resolved as well. At this review, the only cause that could be incriminated was vitamin A overdose.

Discussion

This case report demonstrates the rule of vitamin A overdose in raised intracranial pressure. The first description of this association was made in 1856 by Elisha Kane, who noted vertigo and headache after ingesting polar bear liver [4].
The primary food sources of vitamin A are liver, dairy products, egg yolk, fish (retinol or retinyl esters) and yellow and green vegetables (carotene).

Our case involved well-meaning parents who gave to their daughter tuna's liver rich of vitamin A, thinking that such supplementation will strengthen their child’s immune system. Recommended nutritional intake of vitamin A for our patient is 1333UI per day (400µg/day).

The content of hepatic vitamin A storage is approximately 100 to 300mcg/g as retinyl esters. When a person consumes an excessive amount of vitamin A, there is an hepatotoxicity resulting from excessive retinyl ester and elevated circulating vitamin A, as retinol and retinyl esters, inducing a systemic toxicity [5].

Hypervitaminosis A has been reported to cause PCT in children and adults [6,7]. The pathophysiology is not clear. One of the hypotheses is that the elevated serum retinol is transported with the retinol binding protein to the CSF, where retinol acts like a toxin [8].

In addition to the risk of pseudotumor cerebri, systemic effects of vitamin A megadosage include dermatologic abnormalities (dry skin, pruritic, peeling skin, hair loss, cheilitis, stomatitis, gingivitis), skeletal system abnormalities (bone pain, tenderness, growth disturbance, osteoporosis, cortical hyperostosis, periosteal calcifications), teratogenic effects well described with vitamin A derivative acne medication [4].

All patients presenting an increased intracranial pressure should undergo magnetic resonance imaging with venogram to exclude other causes. After that, a lumbar puncture should be performed. The composition of the CSF is normal. Opening pressure greater than 28cm H2O in children is considered elevated. However, greater than 25 cm H2O is considered elevated in those not sedated during the lumbar puncture and non-obese children [8].

Assessing vitamin A status in persons with subtoxicity or toxicity is complicated because serum retinol concentrations are nonsensitive indicators in this range of liver vitamin A reserves [9].

The treatment of PCT goal’s is to prevent vision loss. There are no randomized clinical trials for evidence-based recommendations in the treatment of pediatric pseudotumor cerebri. Acetazolamide is frequently recommended in the treatment of pediatric patient. The recommended starting dose is 15 – 25mg/kg/day in 2 to 3 divided doses per day. This can be gradually increased up to 100mg/kg, without exceeding 2g/day in children and 4g/day in adolescents. Other treatment can be used as alternative to acetazolamide, such as furosemide, topiramate, corticosteroids. [4] The treatment should be taking until the resolution of papilledema. The follow up is based on the visual assessments, optic nerve appearance, and functional symptoms of elevated intracranial pressure. In the case of secondary pseudotumor cerebri, it is a priority to remove the offending agent. If the medical therapy is not enough, surgical interventions such as an optic nerve sheath fenestration or cerebrospinal fluid shunting can be proposed [8].

Conclusion

Pseudotumor cerebri can cause, if undetected, permanent blindness. The emergency pediatrician must be aware of this condition. Our case highlights the importance of asking about dietary intake and supplements when evaluating a patient with pseudotumour cerebri.

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References