Pediatric Oncall Journal
Volume : 21, Issue 1:28-29
DOI: https://doi.org/10.7199/ped.oncall.2024.3

PEDIATRIC ONCALL
CHILD HEALTH CARE

CASE REPORTS

RARE CAUSE OF PSEUDOTUMOR CEREBRI IN CHILDREN

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ABSTRACT

The benign intracranial hypertension called Pseudotumour cerebri may be primary or secondary. Secondary causes should be excluded first. We report a case that illustrates the great significance of asking about dietary intake when exploring a patient with pseudotumour cerebri.

Keywords

Pseudotumour cerebri, intoxication, vitamin A, raised intracranial pressure

ARTICLE HISTORY

Received 24 February 2022

Accepted 1 June 2022

ABBREVIATIONS :

CSF : Cerebrospinal fluid

MRI : Magnetic resonance imaging

PTC : Pseudotumor cerebri

Case Report

Aims :

Pseudotumor cerebri (PTC) is a benign elevated intracranial pressure that occurs usually in obese young adults, specially women. This syndrome is characterized by headache, papilledema and possible sixth nerve palsy with visuel field defects.[1] It may be idiopathic or secondary.[2] Secondary and treatable causes of raised intracranial pressure should be excluded in all patients, particularly in children and teneeger with normal quetelet index.[3] We report a case for whom ingestion of vitamine A overdose was associated with neurological symptoms of benign intracranial hypertension.

Presentation of Case :

A eight Year-old girl complained of severe headache that get worse when waking, with vomiting, blurring vision and diplopia. Those symptoms lasted nine days. The clinical examination was normal apart a bilateral convergent strabismus. Quetelet index was 12,5 kg/m^2 for a normal between 13.2 to 22.9 kg/m^2 (-2 z-scor). Her visuel acuity was 10/10 in right and left eyes. Fuduscopic 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). Ophtalmic examination revealed no cause for optic disc swelling. Neurological examination was normal. The exploration using Magnetic resonance imaging (MRI) and venography showed no space-occupying lesion, venous thrombosis or stenosis. Blood tests were normal, including blood count, hepatic and renal function. Cerebrospinal fluid (CSF) opening pressure was 27 cm H2O, in lateral decubitus position, with normal analysis. The history taking identifed an ingestion of a large amount of tuna liver. The dosage of vitamine A (Retinol) in her plasma, nine days after ingestion, was elevated to 2.72 µmmol/l (Normal: 0.90-1.20).

Based on all these arguments, the diagnosis of pseudotumor cerebri secondary to a vitamin A overdose was retained.

There was no need for an urgent lumboperitoneal shunt. She received a treatment based on acetazolamide 20 mg/kg/day. After 8 weeks of follow-up, the papilloeodema 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 condition was firstly descripted in 1856 by Elisha Kane, who reported vertigo and headache after having polar bear liver.[4] The vitamin A exists in many food sources like liver, dairy products, egg yolk, fish, yellow and green vegetables.[4] 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

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vitamin A for our patient is 1333 UI per day (400 µg/day).

The content of hepatic vitamin A storage is approximatively of 100 to 300 mcg/g as retinyl esters. If 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 hypothesis is that the elevated serum retinol is transported with the retinol binding protein to the CSF, where retinol acts like a toxin.[8]

Besides of the risk of pseudotumor cerebri, systemic effects of vitamine A megadose include dermatologic abnormalities as dry skin, pruritic, peeling skin, hair loss, cheilitis, stomatitis, gingivitis; skeletal system abnormalities as bone pain, tenderness, growth disturbance, osteoporosis, cortical hyperostoss, periosteal calcifications; teratogenic effects well noticed with 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 28 cm H2O in children is considered eleveted. However, greater than 25 cm H2O is considered abnormal in those not sedated during the lumbar puncture and non-obese children.[8]

The assessment of vitamin A status in persons with subtoxicity or toxicity may be difficult 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. Acetazolamid is frequently used in the treatement of pediatric patient. The recommended starting dose is 15-25 mg/kg/day in 2 to 3 divided doses per day. This can be gradually increased up to 100mg/kg, without exceeding 2 g/day in children and 4 g/day in adolescents. Other alternatives can be used such as furosemide, topiramate, corticosteroids.[4] The treatment should be taking until the resolution of papilledema. The follow-up is based on the visuel assessments, optic nerve appearance and functional symptoms of elevated intracranial pression. 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 performed.[8]

Conclusion

Pseudotumor cerebri can cause, if undetected, permanent blidness. 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.

Authors Contribution

All authors had access to the data and participated in writing the manuscript.

Compliance with Ethical Standards

Funding None

Conflict of Interest: None of the authors have any conflicts of interest to disclose.

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