Comment on "Hypophosphatasia Associated with Pseudotumor Cerebri and Respiratory Insufficiency"

Sir,

I read the article by Dr. Teber *et al*.[1] regarding hypophosphatasia and pseudotumor cerebri. Although an article of great interest, certain details remained suspect. While it is true that many metabolic derangements have been associated with pseudotumor cerebri, the authors state that "hypophosphatasia is an extremely rare cause of pseudotumor cerebri." A causal relationship between hypophosphatasia and pseudotumor cerebri is yet to be conclusively demonstrated in the literature.

It is also implied in the article that the diagnosis of pseudotumor cerebri is made in this infant based on the elevated opening CSF pressure of 430 mm H2O. Although elevated opening CSF pressures are associated with pseudotumor cerebri, this is not diagnostic. The diagnosis of pseudotumor cerebri still hinges on the modified Dandy Criteria set forth in 1985, which also require that the patient have normal to small slit ventricles on imaging with no intracranial mass.[2]

This patient's CT scan revealed "mild dilatation in third and lateral ventricules [sic]," suggesting that this patient in fact did not have pseudotumor cerebri. A patient presenting with bulging fontanelles, increased intracranial pressure, and with dilated ventricles on neuroimaging suggests the diagnosis of pediatric hydrocephalus. Hydrocephalus is in fact a much more common disorder in this age group, as infantile pseudotumor cerebri would itself be a rare presentation, much less secondary to a rare etiology. This behooves us to rule out more common etiologies before searching for an obscure one.

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Author's Reply

Sir,

Dr Kapoor pointed out that a causal relationship between hypophosphatasia and pseudotumor cerebri (PTC) is yet to be conclusively demonstrated in the literaure. It is well known that hypophosphatasia and PTC are both rare conditions and their existence together is extremely rare and the causal relationship is not clear[1]. This is why we have published this case. There is no case report about the relationship of PTC and hypophosphatasia in Pubmed but there are few reports about PTC and nutritional ricets. They emphasized that the pathogenesis of pseudotumour cerebri is uncertain but derangement of calcium or phosphorus metabolism is responsible through alterations in cellular energy utilization, membrane structure and function, or intercellular ion concentrations.[2,3,4]

The second comment was about the ventricular dilatation that exists in our patient. Our patient's CT scan is not consistent with hydrocephalus, there was only mild dilatation in third and lateral ventricules due to cerebral atrophy. Probably this dilatation already existed just before the diagnosis of PTC. We don't think that this mild dilatation can cause the symptoms of increased intracranial pressure or an opening CSF pressure of 430 mmH2O. As he did'nt have hydrocephalus, the symptoms of pseudotumour cerebri responded well to the corticosteroid therapy.

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