Acute multifocal dystonic reaction: An unusual presentation of vitamin D-deficient rickets

Sir,

A 7-year-old boy born of non-consanguineous marriage presented with inability to open his mouth. He had episodic painful spasm of the jaw muscles since last 7 h with each attack lasting between 2 and 5 min. During each attack he was unable to open his mouth due to spasm of bilateral masticatory muscles. This was followed by complete recovery in between the attacks. His sensorium was normal during the attacks. The painful spasm of the jaw muscles subsided spontaneously within 5 min after admission. There was a history of intermittent spasm of the right hand since 1 day. There was no history of spasm in other muscles of the body. There was no history of fever, convulsion, dyspnea, paresthesia, altered sensorium, involuntary movements, drug intake, or trauma. He was completely immunized as per his age. His developmental and birth history was normal. He was breastfed till 19 months and weaning started at 10 months of age. He was a pure vegetarian and a detailed dietary history revealed deficient intakes of vitamins D. There was no history of any treatment received for rickets in the first 2 years of life. On admission, he was hemodynamically stable. Hyperventilation test and Trousseau sign were negative. Chvostek sign was positive bilaterally. His weight was 17 kg and height was 111 cm (both below 5th percentile for age and gender). Systemic examination was normal. Mild pallor was present. There was no pain

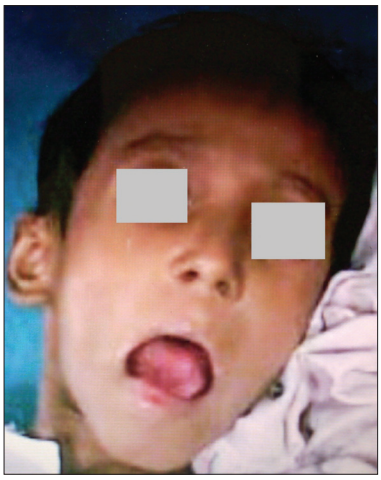

Figure 1: Clinical photo showing spasm involving bilateral jaw muscles

or restriction of movements at the temporomandibular joint bilaterally. A repeat attack of painful spasm of jaw muscles occurred while examining the oral cavity. During this episode he was not able to close his mouth [Figure 1]. He also developed carpopedal spasm of the right hand. Intravenous calcium gluconate (100 mg/kg) was given, which was followed by complete resolution of the spasm within 2 min. His investigations revealed the following: Serum calcium (Ca) 7.1 mg/dL, serum magnesium 2.2 mg/ dL, serum phosphorous (P) 2.9 mg/dL, alkaline phosphatase 981 IU/L , serum parathormone 272 pg/mL (N, 9–65 pg/mL), 25-OHD 12 ng/mL (N, 30.0–74.0 ng/mL). His hemogram was suggestive of iron deficiency anemia. The Ca × P product of our patient was 20.59. Radiography of the wrist showed evidence of rickets in the form of metaphyseal splaying/fraying and decreased bone density. His liver and renal function test, arterial blood gas analysis, and serum electrolytes were normal. A diagnosis of acute multifocal (multiple noncontiguous body parts affected) dystonic reaction secondary to vitamin D–deficient rickets (VDDR) was made. He was started on vitamin D, calcium, and iron supplements. On follow-up after 3 months he is asymptomatic and well.

VDDR is an important and common problem in the developing countries. Presentations of VDDR vary widely, from asymptomatic to life-threatening situations.[1] Various atypical presentations, such as myelofibrosis, stridor, and dilated cardiomyopathy have also been described.[2-4] Thompson *et al* described a case of a 32-year-old woman with episodic and unilateral spasm of masticatory and facial muscles due to tetany. To the best of our knowledge our case is the first to be described with bilateral jaw muscle spasm due to tetany. The various causes of tonic or clonic jaw spasms include tetanus, bruxism, trauma, tetany, hysteria, pontine lesions, cold exposure, and convulsion.[5] These should be differentiated from involuntary movements of the jaw, which occur in conditions, such as generalized and cranial dystonia, acute dystonic reactions to neuroleptic drugs, and tardive dyskinesia. Various masticatory movements may also be seen in epileptic automatisms and in response to oral/facial stimulation in patients with severe damage of the cerebral hemispheres/brainstem.[5] The prompt resolution of spasms after intravenous calcium distinguishes our case from the other causes. In VDDR, bone mineralization has a definite relation to the Ca × P product in the serum. Rickets would be present if the Ca × P product is below 30 as seen in our patient; and it would not develop if this product is above 30.[6] Vitamin D deficiency leads to impaired intestinal absorption of calcium, resulting in decreased serum calcium. This results in secondary hyperparathyroidism that initially maintains serum calcium levels at the expense of the skeleton. However, with prolonged vitamin D deficiency resulting in osteomalacia, calcium stores in the skeleton become relatively inaccessible, since osteoclasts cannot resorb unmineralized osteoid, and frank hypocalcemia ensues.[7] This could probably explain the hypocalcemia seen in our patient. Various hypotheses have been put forward to explain the mechanism of dystonia in vitamin D deficiency. The increase in serum calcium due to secondary hyperparathyroidism can stimulate dopamine release, resulting in dystonia.[8] As per another hypothesis, calcitriol is known to decrease the production of inflammatory cytokines in the brain. Thus in the absence of calcitriol, excessive production of inflammatory cytokines can occur, which could be responsible for the dystonia.[9]

In conclusion, physicians should be aware of the different ways in which hypocalcemia can present to ensure its early diagnosis and treatment. Also, vitamin D deficiency rickets should be considered in the differential diagnosis of children presenting with bilateral jaw muscle spasm.

Syed Ahmed Zaki, Vijay Lad
Department of Pediatrics, Lokmanya Tilak Municipal General Hospital and Medical College, Sion, Mumbai, India

Correspondence:
Dr. Syed Ahmed Zaki, Department of Pediatrics, Lokmanya Tilak Municipal General Hospital and Medical College, Sion, Mumbai- 400 022, Maharashtra, India.
E-mail: drzakisyed@gmail.com

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