Pediatric idiopathic intervertebral disc calcification of the cervical spine

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To the Editor: Pediatric idiopathic intervertebral disc calcification (PIIVDC) is a rare childhood disease characterized by calcification of the intervertebral disc and can progress to inflammation or extrusion. The cause of intervertebral disk calcification in children remains unclear. Increasing inflammation and extrusion can eventually lead to neck or spinal pain in some patients. Most reported cases are asymptomatic or mildly symptomatic, and the diagnosis is largely incidental. PIIVDC is thought to be self-limited with favorable outcomes even when treated conservatively by immobilizing the spine and using analgesic therapy. However, there are no gold guidelines for the treatment of this disease at present. In this study, we reported 12 children with cervical disc calcification who presented to our hospital, nine of whom received non-interventional treatment with satisfactory outcomes, in an attempt to outline the presentation, clinical features, and clinical management of this rare disease.

Database records were reviewed retrospectively to identify patients aged 0 to 18 years with PIIVDC who were treated in outpatient and inpatient departments of our center between January 2005 and April 2019 and were followed up for at least a year after diagnosis. Patient data including age, gender, symptoms, medical history, physical, neurological and imaging findings, management, follow-up periods, and outcomes were collected and analyzed. The research protocol was approved by the Institutional Review Board of Changzheng Hospital (Shanghai, China).

Twelve patients (nine boys and three girls), aged between 5 years and 18 years, diagnosed with cervical disc calcification, and treated in our hospital between 2005 and 2019, were enrolled in this study. All patients were asymptomatic at the time of presentation. Of these, 10 children (10/12) had neck pain; 3 (3/12) had symptoms of torticollis; 4 children (4/12) had neurologic symptoms involving the upper limbs, including 1 (1/12) with muscle weakness in the hand; and four (4/12) had numbness or radiating pain in the hands. No bladder or anal sphincter dysfunction was observed. Three children (3/12) had a history of neck injury, including two (2/12) who had fever prior to the onset of other symptoms.

All 12 patients underwent anteroposterior and lateral radiographs of the cervical spine. Seven patients (7/12) underwent computed tomography (CT) to confirm the calcified density of the nucleus pulposus. Five patients (5/12) underwent magnetic resonance imaging (MRI) of the cervical spine to confirm the extent of spinal cord compression [Figure 1]. Focal dense areas of enhancement were detected in the respective disc spaces. Most children had one-level involvement of the cervical spine. Five of the 12 disc calcification lesions (41.7%) were located at the C4/5 level. Only one child (1/12) had radiographs showing involvement of two calcified intervertebral discs at C2–C3 and C3–C4 with flattening of the C3 vertebra.

Considering that the patient’s symptoms were not very severe and that the disease was benign and self-limiting, 9 of the 12 patients (75%) were treated with non-interventional strategies including rest, intensive follow-up observation, and avoidance of strenuous activities, whereas the other three patients (25%) were treated with analgesics or a neck brace for persistent neck pain or stiffness. All the patients were followed up at 3 months, 6 months, and 12 months, and annually thereafter until both the symptoms and calcification had resolved. All patients achieved good therapeutic outcomes with no worsening of symptoms after treatment. Five patients (5/12) achieved complete symptom relief in <3 months. The nine patients who received non-interventional treatment did not experience any exacerbation or recurrence of symptoms. Follow-up imaging showed that 8 of the 12 patients (66.7%) achieved complete resolution of disc space calcification within 2 years and 4 (4/12) patients had a reduction in the volume of the calcifications during the follow-up period but did not achieve complete resolution.

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A 5-year-old boy was admitted to our hospital with a sudden onset of neck pain, torticollis, and focal neurological deficiency. He did not have any specific medical history. The symptoms started 2 months earlier with neck pain and torticollis due to a trauma-induced neck strain. The patient was referred to a local hospital where he underwent an X-ray examination which did not reveal any abnormalities. The symptoms improved on day 5 after the onset. A week before the boy was presented to our department, he started to experience weakness in his left upper limbs. He was then brought to our department, where physical examination revealed limited activity of the neck and a muscular deficit of the left musculus deltoideus. Cervical spine radiographs showed C4–C5 intervertebral disc calcification. Admission CT scan demonstrated calcification of the nucleus pulposus with lateral herniated calcification at the C4–C5 level. MRI revealed decreased signal intensity of the involved disc on T1- and T2-weighted images, causing serious spinal cord compression. Other intervertebral disc levels and all vertebrae seemed normal. The patient was treated conservatively under clinical supervision only and was advised to avoid strenuous exercise. After 2 weeks of treatment, there was a marked improvement in the left upper limb weakness. A neurological examination in 3 months showed complete resolution of the symptoms. A CT scan of the cervical spine performed 3 months after the initial presen-
tation, showed continued disappearance of the calcified disc and extrusion.

Since the first case of PIIVDC was described, there have been very few reports of PIIVDC. About 70% of the PIIVDC cases are located in the cervical spine. The etiology of disc calcification in children remains unclear. It is generally believed that genetic and/or metabolic defects play a role in the pathogenesis of PIIVDC. Other suggested etiologies include inflammation triggered by infection, reduced nutrient supply, or trauma. There is compelling evidence that increased nutrient shortage promotes degeneration, knowing that the intervertebral discs of children are in a high metabolic state. Subsequent inflammation causes the nutrients to diffuse and probably reverse disc degeneration, causing the spontaneous resolution of the calcified lesions that often are observed.

PIIVDC involves calcified lesions in the intervertebral disc, most commonly located in the nucleus pulposus. Even though calcifications can be discovered in any part of the spine, the cervical spine is most frequently affected in childhood. Patients with cervical PIIVDC often present with neck pain and occasionally associate with neurological dysfunction. Some cases are usually found asymptomatic and found incidentally on imaging examination. Associated lesions can lead to posterior disc herniation and, although rare, can cause nerve root or cord compression.

There is no gold treatment guideline available for PIIVDC so far. In our series, none of the cases developed malignant symptoms and all resolved spontaneously with only conservative treatment, including analgesics, anti-inflammatory drugs, cervical collars, or even without any intervention. This highlights the benign nature of this condition. Dushnicky et al. and Zhu et al. also reported their non-interventional treatment and all the patients recovered fully. We believe that, even with any clinical intervention the calcified lesion may aggravate and the recurrence of symptoms may occur as the natural condition progresses until inflammation triggers the point of regression of the calcified lesions. Therefore, in most cases, intervention and immobilization may not be required.

Based on the experience obtained in our center, we have summarized some principles for the treatment of PIIVDC as follows: (1) For patients who are asymptomatic or have mild neurological deficits, especially those <10 years of age, non-interventional measures should be taken, including rest and intensive follow-up observation. Patients should be followed up intensively and be advised to avoid strenuous activities. (2) Conservative treatment including analgesics, cervical collars, and traction should be recommended for pediatric patients with persistent and intolerable symptoms, such as neck pain, stiffness, torticollis, and back pain. Symptoms usually resolve spontaneously within 6 months by conservative treatment. The size and location of calcification are not related to the time of resolution of either the symptoms or the calcification. Therefore, regular radiological follow-up examinations are required in these patients. (3) Surgical treatment should be recommended for patients with progressive neurologic deterioration, whether with or without an adequate course of conservative treatment. Critical attention should be paid to the aggravation of symptoms, which usually manifest as weakness of the extremities. So far as surgical treatment is concerned, either anterior cervical corpectomy/discectomy or posterior laminoplasty/laminectomy can be chosen depending on individual circumstances.

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