Children Presenting with Calcified Disc Herniation: A Self-Limiting Process.

Chang Ho[1*], Stanley Chang[1], Daniel Fulkerson[2], Jodi Smith[2]

1. Department of Radiology, Riley Hospital for Children, Indianapolis, USA

2. Department of Neurosurgery, Riley Hospital for Children, Indianapolis, USA

* **Correspondence:** Chang Ho, Indiana University School of Medicine, Department of Radiology, 705 Riley Hospital Drive, Indianapolis, IN 46202, USA
(*cyho@iupui.edu*)

Radiology Case. 2012 Oct; 6(10):11-19 :: DOI: 10.3941/jrcr.v6i10.1162

ABSTRACT

We present two cases of disc herniation associated with juvenile intervertebral disc calcification, a rare, self-limiting process which typically resolves with conservative treatment. Recognition of this entity prevents unnecessary diagnostic workup and possible surgical intervention. A review of the literature for this rare entity is discussed.

CASE REPORT

CASE REPORT

Case 1:

A 7-year-old boy presented with an episode of severe neck pain for 2 weeks duration with initial onset during sleeping. There was no history of fever, recent infection, or trauma. On examination, the patient reported neck pain with flexion and extension; otherwise the physical exam was normal. There were no neurologic deficits, including no weakness, paresthesias, or gait abnormalities. The patient was afebrile. Laboratory values were not obtained. A CT scan was performed which demonstrated calcification of the nucleus pulposus of the C2-C3 intervertebral disc with a large calcified extrusion into the ventral epidural space with superior migration. The extruded, calcified fragment measured 5 x 8 x 12 mm in AP, transverse, and craniocaudal dimensions. There was resultant moderate canal stenosis and mass effect on the spinal cord (Figure 1).

Based on the results of the CT, he was referred to a pediatric neurosurgeon who, despite the moderate canal stenosis and mass effect on the spinal cord, elected to treat him conservatively. He was placed in a cervical collar and his pain was managed with ibuprofen. The patient returned to clinic for additional follow up visits at 1, 2, 3, and 6 months, with incremental improvement and eventual resolution of his pain. He subsequently returned to normal activities, except for contact sports. At the time of his six month follow up, a CT scan of the cervical spine demonstrated a marked decrease in disc calcification with near complete resolution of the extruded calcified disc material. There was persistent calcification of the nucleus pulposus with extension to the posterior annulus, but the canal stenosis and mass effect on the spinal cord had completely resolved (Figure 2).

Case 2:

A 4-year-old female presented with neck pain after falling off a bed. Besides the cervicalgia, she had no muscle weakness, sensory disturbance, or gait problems. Moreover, she had no fever or signs of systemic infection. Laboratory studies were not obtained. Plain films, CT scan, and MRI of the cervical spine revealed calcifications in the disc space at C4-C5 and C5-C6 with anterior herniation at C4-C5 (Figure 3).

The patient was treated conservatively with analgesics and had rapid improvement. She was asymptomatic until three years later when she fell in a hole while running outside. At that time she began complaining of neck pain again. Despite the pain, she had a normal neurologic exam with full strength and neck range of motion. Once again, there was no history of

fever or prior infection. No laboratory studies were performed. Plain films and a CT scan of the cervical spine demonstrated complete resolution of the disc calcification at the C4-C5 level; however, decreased but persistent calcification was seen at the C5-C6 level with superior herniation into the inferior endplate of C5 (Figure 4). Again, she was treated conservatively with complete resolution of her pain.

The imaging findings and clinical history for both cases suggested a diagnosis of juvenile intervertebral disc calcification; the resolution of symptoms and near-complete resolution of imaging findings with conservative management were confirmatory.

DISCUSSION

Juvenile intervertebral disc calcification is a rare entity whose etiology is poorly understood. First described by Baron in 1924, there have been at least 150 cases described in the literature since that time [1]. Inflammatory or posttraumatic etiologies have been suggested. A recent study hypothesized a viral-induced vasculitis resulting in ischemia due to interruption of blood supply from the endplate as a possible cause [2]. Disorders of calcium metabolism have not been found in these patients [1,3].

Patients typically present between the ages of 5 to 10 years, with males affected more often than females. The most common presentation is neck pain; however, the imaging findings have been seen in asymptomatic patients, as well. Associated symptoms which may be present include paraspinal muscle spasms, torticollis, and fever. Laboratory values which can be seen include elevation of the white blood cell count and erythrocyte sedimentation rate, suggesting a possible infectious or inflammatory etiology. Most patients do not present with a clear-cut preceding event. Single-level or multilevel disc calcification has been reported. Involvement of a single level is typically seen in the cervical spine, which is also the most common location of symptomatic patients [2,4,5].

Characteristic imaging findings include calcification of the nucleus pulposus, with or without associated disc herniation. On MRI, the intervertebral discs have been described to be hypointense on T1 and T2 weighted sequences, compatible with dense calcification [3]. Imaging findings tend to mirror the clinical course with complete or near-complete resolution over several months. However, residual calcification of the intervertebral discs can persist for years [4,5]. Intervertebral disc herniation and/or marrow edema have also been described [1,6].

Differential diagnoses include other entities which may cause disc calcification, including metabolic derangement such as alkaptonuria (ochronosis), acromegaly, hyperparathyroidism and homocystinuria. However, these processes are likely to involve more than a few levels of the spine and will likely include other joints as well [7,8]. Rarely, hemorrhage of the disc in hemochromatosis may cause eventual calcification. Other causes of disc calcifications are more likely to be seen in adults, including arthritic causes such as ankylosing spondylitis, gout and calcium pyrophosphate dihyrate disease (pseudogout), each with their own typical radiographic findings. Calcification of Sharpey's fibers is seen early, while "bamboo spine" may be seen in late cases of ankylosing spondylitis, while tophaceous deposits of urate cause bony endplate erosion, edema and bony proliferation, similar to findings in other commonly affected joints in the setting of gout [9]. Pseudogout is typically associated with calcifications of both the anterior and posterior longitudinal ligament and chondrocalcinosis of other joints. Amyloidosis deposits into the discs may also demonstrate calcification, although with preference for the annulus fibrosis rather than the nucleus pulposus [10]. Finally, disc calcification in adults are most commonly related to osteoarthritic degeneration and aging [8]. Other than the associated changes of the vertebral bodies and ligaments in inflammatory arthritides, the appearance of disc calcification may be similar. However, the age of presentation and the lack of any significant medical history should help to differentiate juvenile intervertebral disc calcification from other causes.

Patients are typically treated with conservative management including collar immobilization of the cervical spine and non-steroidal anti-inflammatory medications. Surgery is usually reserved for patients with intractable pain or progressive neurologic deficits [4,5,11]. Clinical symptoms typically resolve over a matter of weeks to months with complete or near-complete resolution of radiographic imaging findings over several months. At least 4 cases in the literature suggest the persistence of both radiologic findings of disc calcification and neck pain over many years, indicating that a minority of these rare cases may not be self-limiting [12]. There is one case in the literature with persistent myelopathy even after treatment with surgical decompression; however, most cases demonstrate complete resolution of symptoms [13].

Although previous cases have been documented in the scientific literature [6], very few of these have been reported in the recent radiology literature with significant extrusion. The true incidence of this entity may be under-reported as patients may be asymptomatic or not present for imaging [14]. Despite dramatic imaging presentation if associated with disc extrusion, recognition of this entity will prevent radiologists from recommending extensive diagnostic workup and from suggesting surgical intervention unless there are progressive neurologic deficits or intractable pain.

TEACHING POINT

Children without a significant medical history presenting with calcified, extruded intervertebral discs on spine imaging should be conservatively managed if not presenting with neurologic deficits or intractable pain. Followup imaging may be helpful to document spontaneous resolution.

REFERENCES

1. Grage LR, Egeblad M. Juvenile interverterbal disc calcification. Eur.Radiol. 1992; 2:377-381

2. Swischuk LE, Jubang M, Jadhav SP. Calcific Discitis in Children: Vertebral Body Involvement (Possible Insight Into Etiology). Emerg Radiol 2008 15:427-430.PMID: 18607651

3. Barrett MO, Anderson JT, Rao B, et al. Multilevel pediatric cervicothoracic intervertebral disc calcifications, Am J Orthop. 2008; 37(7) E126-E128. PMID: 18795190

4. Swick HM. Calcification of interverterbal discs in childhood. J Pediatr. 1975; 86(3):364-9. PMID: 1113224

5. Smith RA, Vohman MD, Dimon JH 3rd, et al. Calcified cervical intervertebral discs in children. Report of three cases. J Neurosurg. 1977 Feb;46(2):233-8. PMID: 833640

6. Mittal P, Saggar K, Sandhu P, Gupta K. Case report: Acute calcific discitis with intravertebral disc herniation in the dorsolumbar spine. Indian J Radiol Imaging 2010;20:205-7. PMID: 21042446

7. Baeva M, Buena A, Dhimes P. AIRP best cases in radiologic-pathologic correlation: ochronosis. Radiographics. 2011 Jul-Aug;31(4):1163-8.8. PMID: 21768245

8. Chanchairujira K, Chung CB, Kim JY, et al. Intervertebral disk calcification of the spine in an elderly population: radiographic prevalence, location, and distribution and correlation with spinal degeneration. Radiology. 2004 Feb;230(2):499-503. PMID: 14752191

9. Duprez TP, Malghem J, Vande Berg BC, et al. Gout in the cervical spine: MR pattern mimicking diskovertebral infection. AJNR Am J Neuroradiol. 1996 Jan;17(1):151-3. PMID: 8770267

10. Ladefoged C, Fedders O, Petersen OF. Amyloid in intervertebral discs: a histopathological investigation of surgical material from 100 consecutive operations on herniated discs. Ann Rheum Dis. 1986 Mar;45(3):239-43. PMID: 3954473

11. Dönmez H, Mavili E, Ikizceli T, et al. Pediatric intervertebral disc calcification. Diagn Interv Radiol. 2008; 14(4):225-7. PMID: 19061169

12. Morris IM, Sheppard L. The Persistence of Clinical and Radiological Features after Intervertebral Disc Calcification of Childhood. Br J Rheumatol 1986; 25:219-221. PMID: 3708237

13. Mahlfeld K, Kayser R, Henning G. Permanent Thoracic Myelopathy Resulting from Herniation of a Calcified Intervertebral Disc in a Child. Journal of Pediatric Orthopaedics B. 2002 Jan 11 (1):6-9. PMID: 11866083

14. Lernout C, Haas H, Rubio A, Griffet J. Pediatric intervertebral disk calcification in childhood: three case reports and review of literature. Childs Nerv Syst 2009;25:1019-23. PMID: 19424706

FIGURES

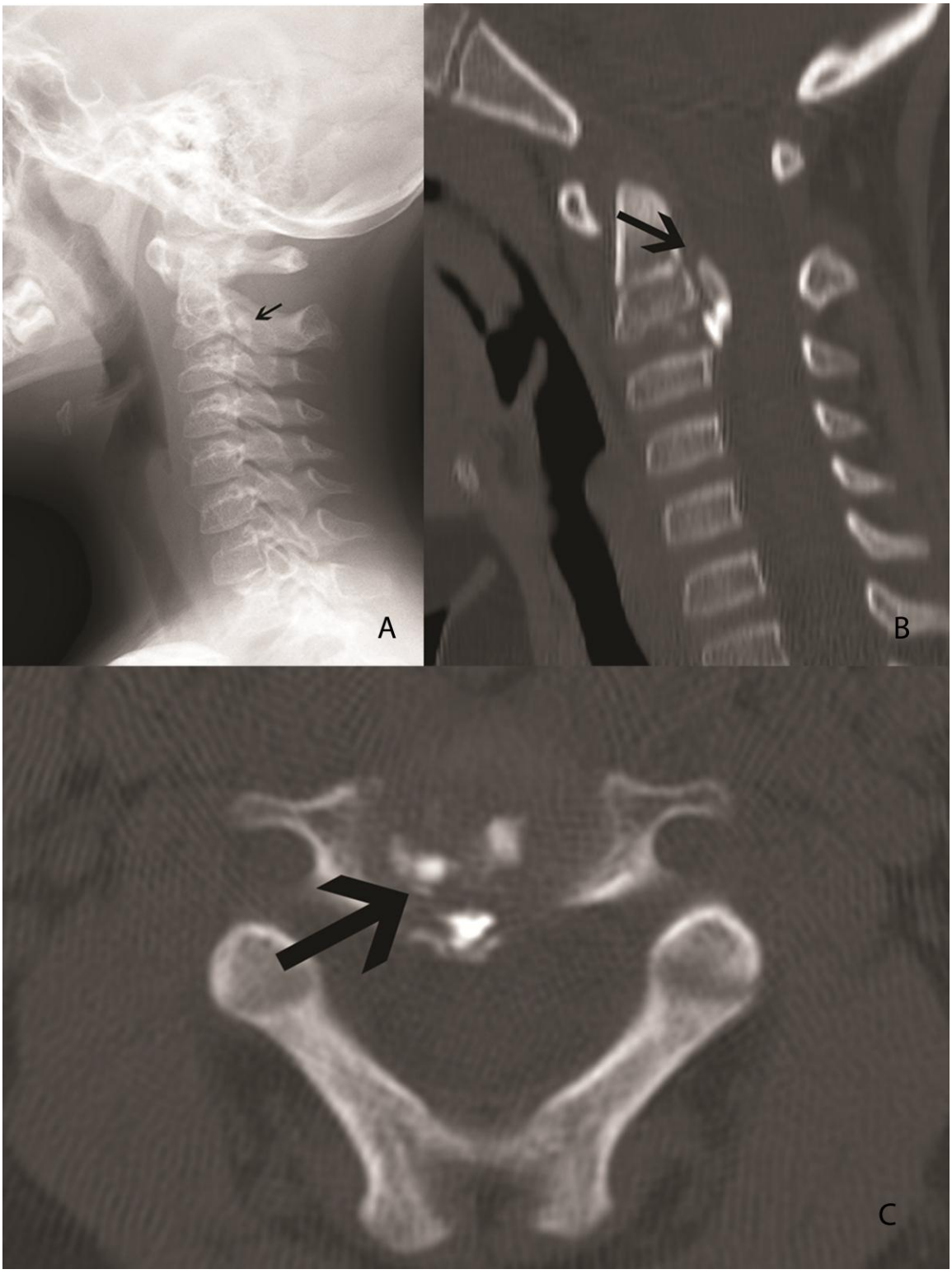

Figure 1: 7-year-old male with juvenile intervertebral disc calcification presenting with neck pain. (A) Sagittal plain film (protocol: 75kVp, 500mA), (B) CT (GE Lightspeed 16 protocol: 120 kVp, 280 mAs, 2 mm slice thickness reformats) sagittal reformat, and (C) axial CT of the cervical spine demonstrating calcification of the C2-C3 disc space with extrusion and cranial migration of 12mm causing indentation of the thecal sac and cervical spinal cord (arrows).

Figure 2: 7-year-old male with juvenile intervertebral disc calcification, follow-up imaging at six months. CT (Philips iCT 128, protocol: 120 kVp, 200 mAs, 2 mm slice thickness reformats) (A) sagittal reformat and (B) axial CT soft tissue window demonstrate incomplete resolution of disc calcification at C2-C3 with significant resolution of the extruded fragment and mass effect on the spinal cord (arrows). The patient was symptom-free at the time of imaging.

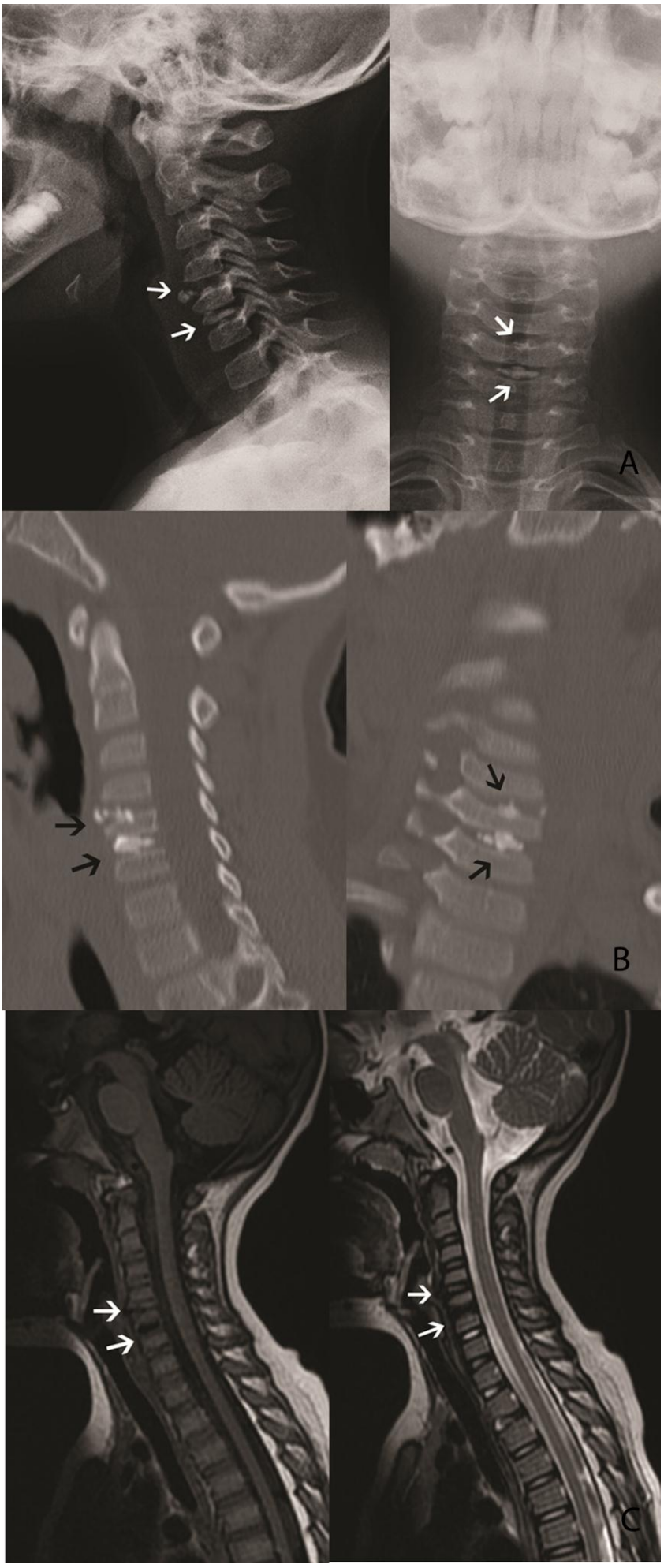

Figure 3: 4-year-old female with juvenile intervertebral disc calcification presenting with neck pain after falling out of bed. (A) Sagittal and AP plain films (protocol: 75 kVp, 500mA), (B) sagittal and coronal CT reformats (GE Lightspeed VCT 64, protocol: 120 kVp, 228 mAs, 2 mm slice thickness reformats), and (C) sagittal MRI FLAIR T1 (GE Signa Excite 1.5T, protocol: TR 1869, TE 24, TI 650, slice thickness 3 mm) and T2 (GE Signa Excite 1.5T, protocol: TR 3000, TE 107, slice thickness 3 mm) images of the cervical spine demonstrate disc calcification at the C4-C5 and C5-C6 levels (arrows). There is a small amount of disc protrusion anteriorly at C4-C5. On MRI (C), the disc calcification appears hypointense on both T1 and T2 weighted images.

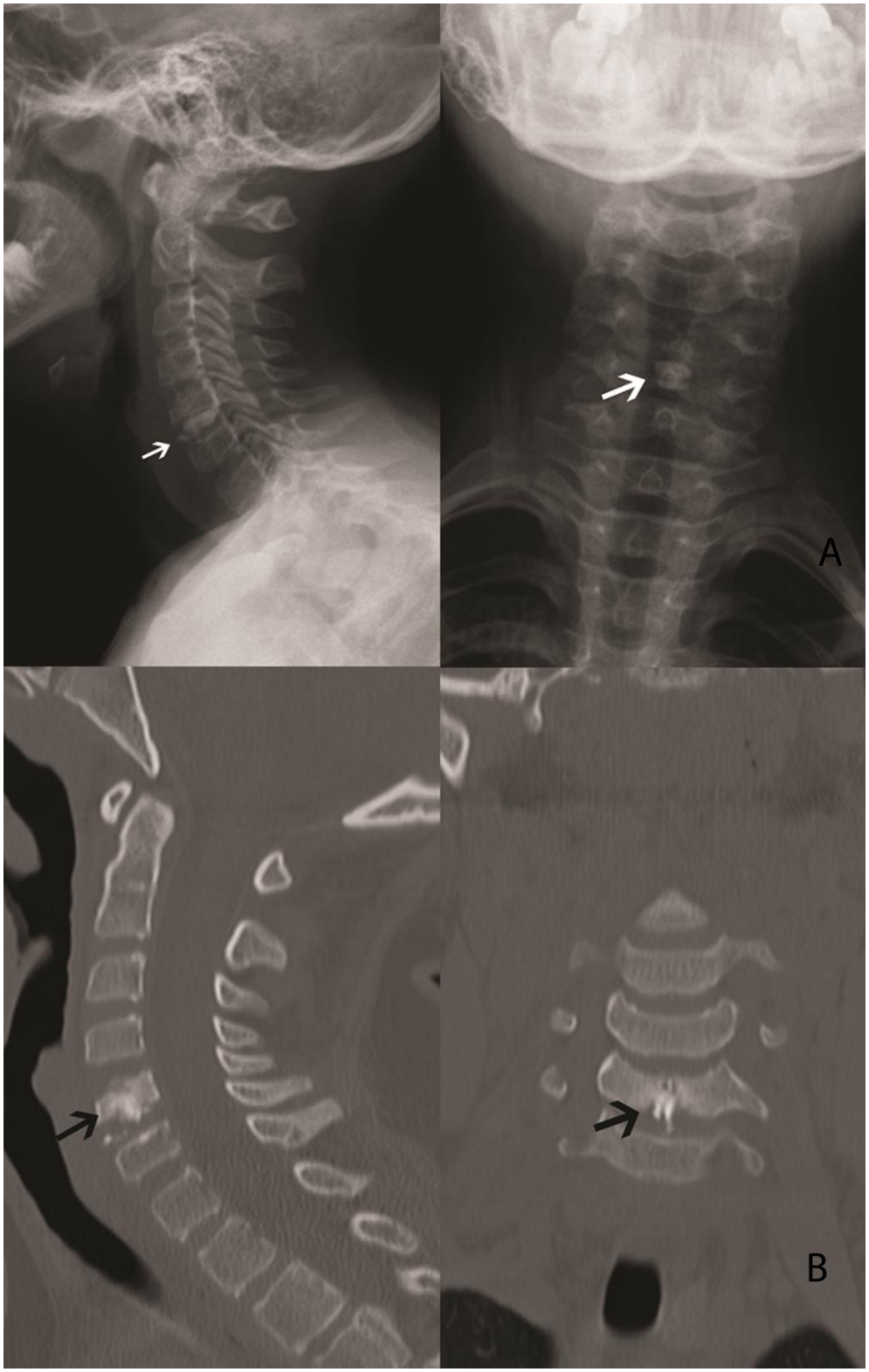

Figure 4: 4-year-old female with juvenile intervertebral disc calcification presenting with neck pain after falling out of bed. 3-year follow-up imaging after presenting with neck pain related to falling in a hole. (A) AP and lateral plain films (protocol: 75kVp, 500mA), and (B) sagittal and coronal CT reformats (GE Lightspeed VCT 64, protocol: 140 kVp, 274 mAs, 1.25 mm slice thickness reformats) of the cervical spine show persistent calcification of the disc at C5-C6 (arrows) with resolution of previous disc calcification at C4-C5. The residual disc calcification appears to cause end plate herniation of the inferior endplate of C5. The patient was asymptomatic shortly after imaging with conservative treatment.

	X-Ray	CT	MRI –T1	MRI – T2	Other Characteristics
Juvenile intervertebral disc calcification	Hyperdense disc	Hyperdense disc	Hypointense disc	Hypointense disc. Hyperintense bone marrow edema has been described	Cervical spine most common, a single or a few levels, childhood presentation
Metabolic processes • **Ochronosis** • **Hyperparathyroidism** • **Acromegaly** • **Homocystinuria**	Hyperdense disc	Hyperdense disc	Hypointense disc	Hypointense disc	Almost all levels of the spine involved. Other joints likely involved as well.
Gout	Hyperdense disc, with endplate erosion and bony overgrowth	Hyperdense disc, with endplate erosion and bony overgrowth	Hypointense disc	Hypointense disc, Hyperintense bone marrow from erosion and edema	Other joints likely involved with characteristic findings
CPPD (pseudogout)	Hyperdense disc, involving anterior and posterior longitudinal ligament	Hyperdense disc, involving anterior and posterior longitudinal ligament	Hypointense disc	Hypointense disc	In addition to ligamentous calcification, may see chondrocalcinosis of other joints.
Amyloidosis	Hyperdense disc	Hyperdense disc, more likely to involve the annulus (periphery).	Hypointense disc	Hypointense disc	Although both annulus and pulposus can be involved, there is a preference for the annulus.
Ankylosing Spondylitis	Hyperdense disc	Hyperdense disc	Hypointense disc	Hypointense disc	May see bamboo spine in late stage of disease with Sharpey's fibers affected in the early stage
Degeneration and Aging	Hyperdense disc	Hyperdense disc	Hypointense disc	Hypointense disc	Advanced age, when associated with disc degeneration presents with endplate changes and osteophytes

Table 1: Differential diagnosis table for calcified intervertebral discs

Etiology	Unknown, possibly post-infectious or inflammatory
Incidence	300 cases reported in the literature (from 1924 to 2011)
Gender Predilection	Male > Female
Age Predilection	5 – 10 years of age
Risk Factors	None
Treatment	Typically conservative, surgical management indicated with neurologic deficits or intractable pain.
Prognosis	Typically excellent
Findings on Imaging	X-ray and CT: Hyperdensity in the intervertebral discs, may be associated with disc herniation. MRI: Hypointensity on T1 and T2 from disc calcification

Table 2: Summary Table - Clinical and Imaging Findings of Juvenile Intervertebral Disc Calcification

ABBREVIATIONS

CT = computed tomography
MRI = magnetic resonance imaging

KEYWORDS

juvenile; idiopathic; intervertebral disc calcification; disc herniation; spine