Intervertebral Disc Calcification and Klippel-Feil Syndrome

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ABSTRACT

Intervertebral disc calcification is rare in the pediatric population and is associated with sudden neurological manifestations. Although commonly symptomatic, conservative management yields excellent prognosis in the vast majority of cases. The following case illustrates the finding of intervertebral disc calcification in a patient with vertebral body segmentation anomaly consistent with Klippel-Feil Syndrome. As both entities are associated with potential neurological sequelae, this case of coexistent pathologies highlights the importance of recognizing the potential presence of intervertebral disc calcifications in pediatric Klippel-Feil Syndrome patients.

CASE REPORT

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An 8-year-old male with recently diagnosed Klippel-Feil Syndrome (KFS) presented with acute onset of non-traumatic neck pain radiating to the left upper extremity and accompanied by paresthesias. Neurological exam was unremarkable for a focal neurologic deficit. Non-contrast CT of the cervical spine (5.2 mGy) demonstrated non-segmentation anomaly at the C6-C7 level consistent with KFS. In addition, there were multilevel intervertebral disc calcifications and a large ossified ventral epidural component within the spinal canal at C4-C5, representing ossification of the posterior longitudinal ligament (OPLL). This contributed to severe spinal canal stenosis and encroached on the left C4-C5 neural foramen, consistent with the patient's left sided C5 radiculopathy.

DISCUSSION

Etiology & Demographics:

Intervertebral disc calcification (IVDC) is a rare condition of unknown etiology in the pediatric population, thought to be an underlying inflammatory process secondary to a recent infectious or traumatic trigger [1]. Overall incidence is low, with a recent literature review from 1997 to 2017 finding 50 cases of IVDC in pediatric patients ranging in age from 45 days to 16 years old [2]. Furthermore, no greater than 400 cases have been described since the first case report in 1924 [3]. The combination of IVDC with OPLL, as seen in the patient above, is much less frequently encountered. Only 8 previous cases have been reported in the literature [4]. Overall, a greater majority of pediatric IVDC patients are male and average age of presentation is 8 years old [2].

Imaging Findings of Intervertebral Disc Calcification:

On CT images, oval shaped calcifications are typically seen in the intervertebral disc nucleus pulposus, rather than the annulus. Furthermore, calcifications are usually seen at one level in the cervical spine. Signal intensity of pediatric IVDC on MR imaging can be hypointense at both T1 and T2. Additionally, changes in signal intensity can be seen in the marrow of adjacent vertebral bodies. While some patients may have resolution of calcification on follow up imaging, residual changes found include persistent disc calcification, flattening of the vertebral body, and osteophytes [5].

Differential Diagnosis:

The differential diagnosis for this entity includes calcium pyrophosphate dihydrate disease/pseudogout, ochronosis/alkaptonuria, amyloidosis, osteochondroma, and osteoma.

In contrast to pediatric IVDC, the calcifications seen in ochronosis/alkaptonuria and amyloidosis are usually found in multiple levels throughout the spine [6].

While calcium pyrophosphate dihydrate disease/pseudogout can be associated with single level intervertebral disc calcifications, such calcifications are usually extensive and can result in destruction of the intervertebral disc [7].

Osteochondromas are benign bone lesions arising from the epiphyseal growth plate. Though commonly found in the long bones, the spine can be affected. On CT, the lesion classically involves continuity between the cortex and medullary region of the bone [8]. Osteochondromas seen on MR imaging typically have mixed signal characteristics depending on the overall size of the lesion, and amount of calcification and marrow within the lesion [9].

Osteoid osteoma, another benign spinal lesion, is typically seen at the posterior of the vertebral column [10]. On CT, osteomas are seen as a lytic lesion with centralized calcification and a surrounding sclerotic rim. Osteomas are heterogenous on MR imaging, however, there may be inflammation seen in adjacent structures [11].

Treatment & Prognosis:

Pediatric IVDC has a favorable prognosis, with most cases having excellent neurological outcomes with conservative management only. Such management includes bed rest, analgesics, physical therapy, and cervical orthosis [12]. Furthermore, follow up imaging on many of these pediatric patients demonstrates complete or near-complete resolution of the disc calcifications [13-15]. Adult KFS patients are known to have higher rates of cervical spine degeneration compared to the general population [16]. With regard to pediatric KFS patients, a prior case report described IVDC that only occurred between fused spinal segments. The authors proposed that calcification was related to vertebral fusion, possibly due to lack of normal mechanical stress between the fused vertebrae [17]. Additionally, the presence of OPLL, as seen in the patient above, has been noted in patients with congenital cervical fusions. The formation of OPLL may be an adaptation to increased adjacent level mechanical stress [14]. Recognition of the potential for IVDC in pediatric KFS patients is important, as it can impact diagnosis and treatment of symptoms. As emphasized previously, IVDC has an excellent prognosis with conservative management. Only rare cases consisting of severe, progressive neurological compromise require surgical intervention [17]. A recent review of the literature found only 20 cases of pediatric IVDC requiring surgical intervention [18]. The patient above was managed conservatively with pain control and physical therapy. He was discharged with outpatient neurosurgery follow up.

TEACHING POINT

Pediatric Klippel-Feil Syndrome (KFS) patients may potentially have coexistent Intervertebral Disc Calcification (IVDC). Recognition of the potential for IVDC in KFS patients is important, as it can impact diagnosis and treatment of symptoms.

REFERENCES

1. Fortes M, Kadom N. 11-year-old boy with history of prior trauma now presents with neck and shoulder pain and torticollis. American Journal of Neuroradiology. 2011. Available at: http://www.ajnr.org/content/cow/11282011. Accessed March 29, 2020.

2. Dushnicky MJ, Okura H, Shroff M, Laxer RM, Kulkarni AV. Pediatric idiopathic intervertebral disc calcification: Single-center series and review of the literature. The Journal of Pediatrics. 2019;206:212-216. PMID: 30466792

3. Yang HS, Chen DY, Yuan W, Yang LL, Tsai N, Lin QS. Paresis associated with aconuresis caused by intervertebral disc calcification at c7-t1: a case report and review of the literature. Spine (Phila Pa 1976). 2010;35(10):E434-E439. PMID: 20393391

4. Zhu J, Sun K, Xu X, et al. A Preliminary Attempt of Nonintervention in the Treatment of Patients with Intervertebral Disc Calcification Combined with Ossification of the Posterior Longitudinal Ligament. World Neurosurg. 2019;129:181-185. PMID: 31146042

5. Diehn FE, Maus TP, Morris JM, et al. Uncommon manifestations of intervertebral disk pathologic conditions. Radiographics : a review publication of the Radiological Society of North America, Inc. 2016;36(3):801-823. PMID: 27082664

6. Jacobson JA, Girish G, Jiang Y, Sabb BJ. Radiographic evaluation of arthritis: Degenerative joint disease and variations. Radiology. 2008;248(3):737-747. PMID: 18710973

7. D Resnick, G Niwayama, T G Goergen, et al. Clinical, radiographic and pathologic abnormalities in calcium pyrophosphate dihydrate deposition disease (CPPD): Pseudogout. Radiology. 1977;122(1):1-15. PMID: 186841

8. Murphey MD, Choi JJ, Kransdorf MJ, Flemming DJ, Gannon FH. Imaging of osteochondroma: variants and complications with radiologic-pathologic correlation. Radiographics. 2000;20(5):1407-1434. PMID: 10992031

9. Sinelnikov A, Kale H. Osteochondromas of the spine. Clin Radiol. 2014;69(12):e584-e590. PMID: 25282617

10. Marcove RC, Heelan RT, Huvos AG, Healey J, Lindeque BG. Osteoid osteoma. Diagnosis, localization, and treatment. Clin Orthop Relat Res. 1991;(267):197-201. PMID: 2044277

11. Gadiya A, Shah K, Patel P, Nene A. Osteoid Osteoma of Cervical Spine: A Case Report and Review of Literature. J Orthop Case Rep. 2019;9(1):78-81. PMID: 31245326

12. Tsutsumi S, Yasumoto Y, Ito M. Idiopathic intervertebral disk calcification in childhood: A case report and review of literature. Childs Nerv Syst. 2011;27(7):1045-1051. PMID: 21494885

13. Garg M, Kumar S, Satija B, Gupta R. Pediatric intervertebral disc calcification: A no touch lesion. Journal of craniovertebral junction & spine. 2012;3(1):23-25. PMID: 23741126

14. Wang G, Kang Y, Chen F, Wang B. Cervical intervertebral disc calcification combined with ossification of posterior longitudinal ligament in an-11-year old girl: Case report and review of literature. Childs Nerv Syst. 2016;32(2):381-386. PMID: 26210494

15. Mac-Thiong J, Leduc S, Marton D, Duhaime M, Morin B. Herniation of a calcified cervical disc into the foramen transversarium in an 8-year-old child. Spine. 2004;29(16):E352. PMID: 15303044

16. Nouri A, Martin AR, Lange SF, Kotter MRN, Mikulis DJ, Fehlings MG. Congenital cervical fusion as a risk factor for development of degenerative cervical myelopathy. World Neurosurgery. 2017;100:531-539. PMID:28130167

17. Dussault RG, Kaye JJ. Intervertebral disk calcification associated with spine fusion. Radiology. 1977;125(1):57-61. PMID: 897188

18. Chu J, Wang T, Pei S, Yin Z. Surgical treatment for idiopathic intervertebral disc calcification in a child: case report and review of the literature. Childs Nerv Syst. 2015;31(1):123-127. PMID: 25192983

FIGURES

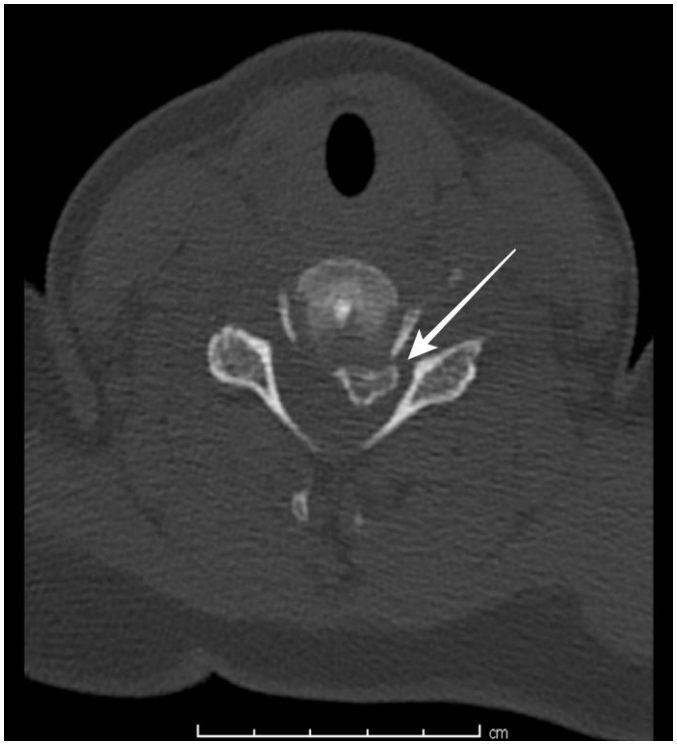

Figure 1: 8-year-old male with intervertebral disc calcification and Klippel-Feil Syndrome
Findings: Axial view demonstrates ossified posterior longitudinal ligament at C4-C5 contributing to severe left-sided spinal canal stenosis and encroachment on the left C4-C5 neuroforamen as indicated by the arrow
Technique: Phillips Ingenuity CT, 332 mAs, 100 KVp, helical 0.9mm axial slices, non-contrast

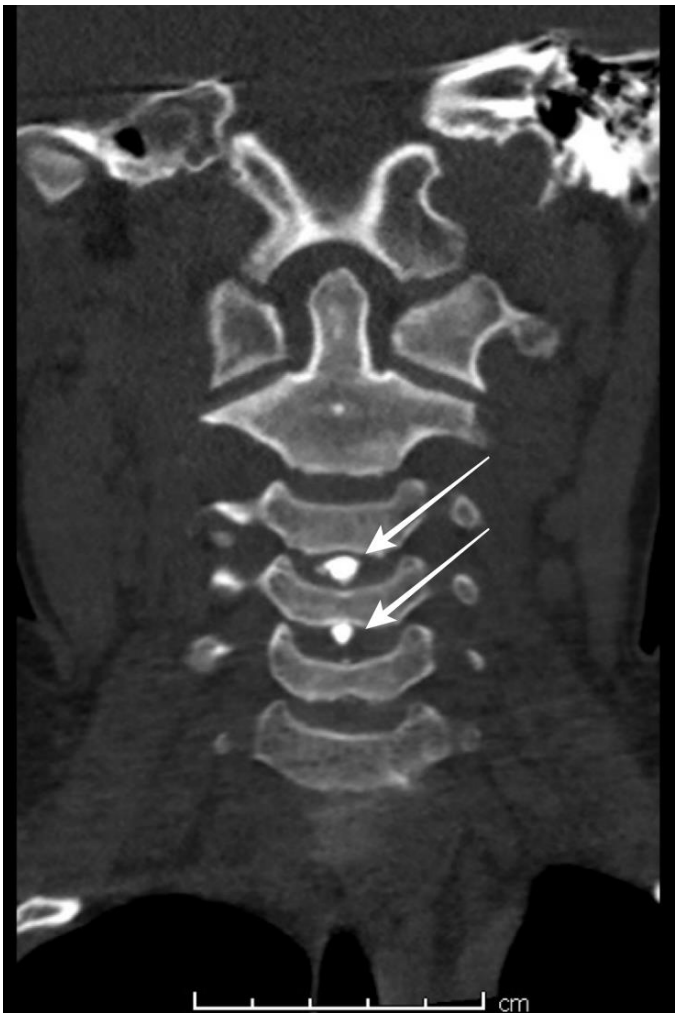

Figure 2: 8-year-old male with intervertebral disc calcification and Klippel-Feil Syndrome
Findings: Coronal view demonstrates arrows indicating intervertebral disc nucleus pulposus calcification at C3-C4 and C4-C5
Technique: Phillips Ingenuity CT, 332 mAs, 100 KVp, helical 0.9mm axial slices, non-contrast

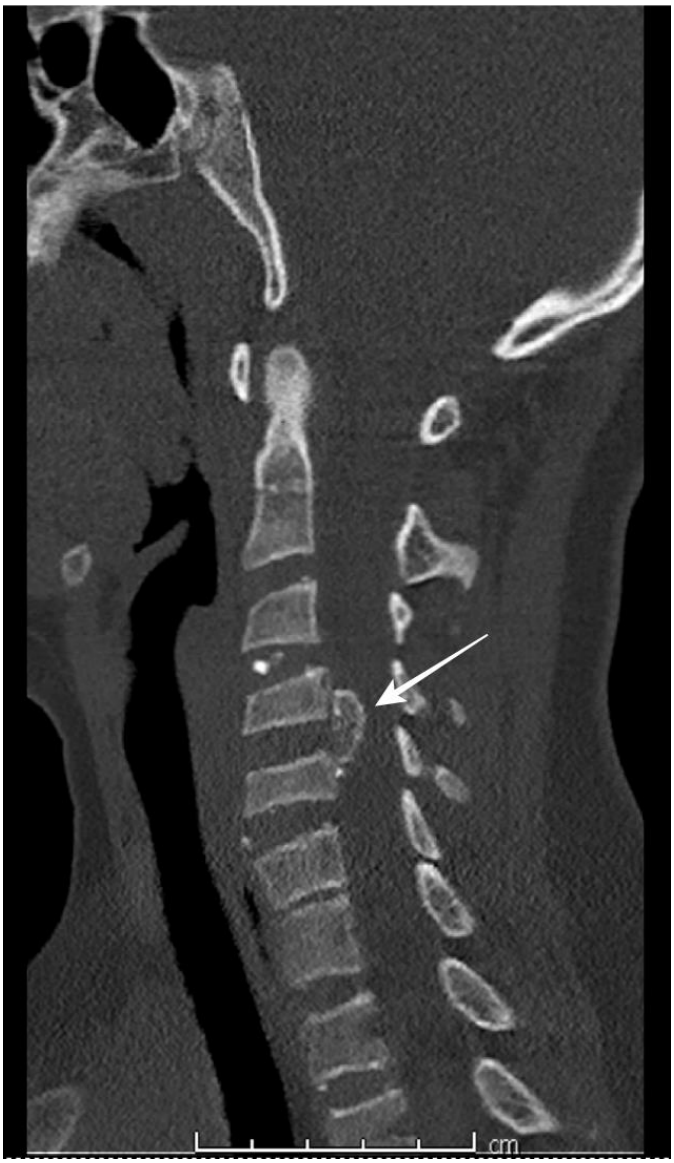

Figure 3: 8-year-old male with intervertebral disc calcification and Klippel-Feil Syndrome
Findings: Sagittal view demonstrates C3-C4 and C4-C5 intervertebral disc nucleus pulposus calcification and an arrow indicating ossified posterior longitudinal ligament dorsal C4-C5
Technique: Phillips Ingenuity CT, 332 mAs, 100 KVp, helical 0.9mm axial slices, non-contrast

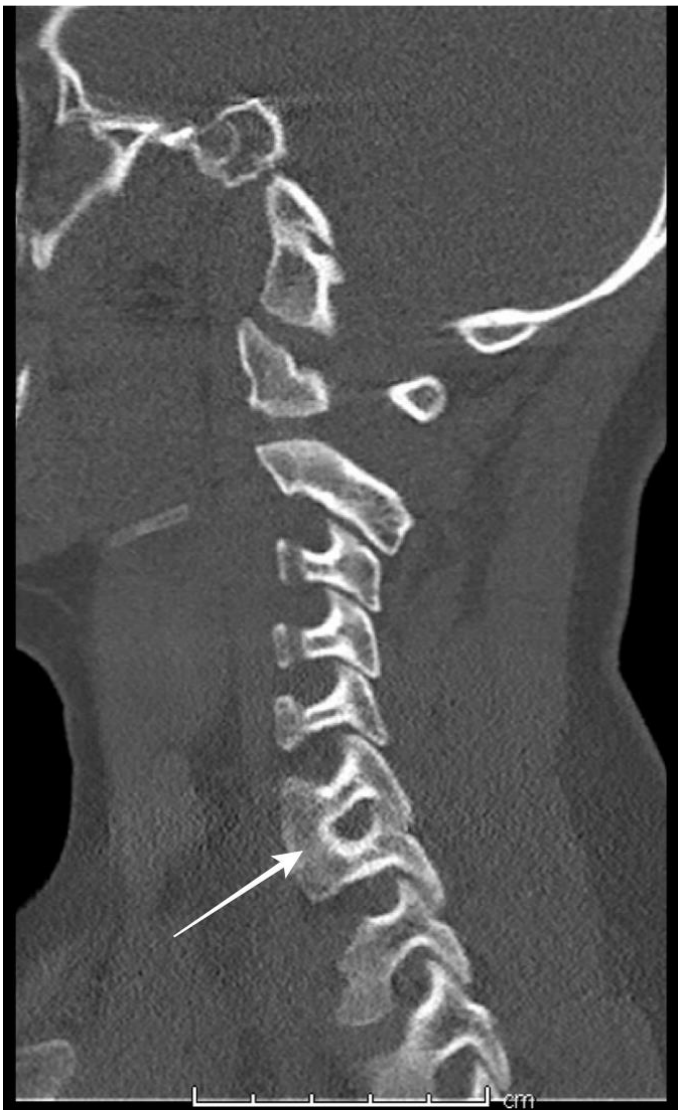

Figure 4 (left): 8-year-old male with intervertebral disc calcification and Klippel-Feil Syndrome
Findings: Additional sagittal view demonstrates left-sided mild, incomplete vertebral body and posterior elements segmentation of C6-C7 as indicated by the arrow consistent with KFS
Technique: Phillips Ingenuity CT, 332 mAs, 100 KVp, helical 0.9mm axial slices, non-contrast

Etiology	Unknown etiology, thought to be an underlying inflammatory process secondary to a recent infectious or traumatic trigger.
Incidence	50 cases reported in the literature from 1997 to 2017. No more than 400 cases total since first case report in 1924.
Gender Ratio	Greater prevalence in male patients.
Age Predilection	Average age is 8 years old.
Risk Factors	Trauma or infection can be a predisposition; however, many cases are found incidentally.
Treatment	Conservative management for the majority of cases, with surgery only needed for patients with evidence of myelopathy.
Prognosis	Favorable, many patients have complete resolution of symptoms.
Findings on Imaging	Lesions are typically seen in the cervical spine, with one level affected. Oval shaped intervertebral disc nucleus pulposus, rather than annulus, disc calcifications are found on radiographs and CT imaging. Signal intensity of calcifications on MR imaging can be hypointense at both T1 and T2. Additionally, changes in signal intensity can be seen in the marrow of adjacent vertebral bodies. While some patients may have resolution of calcification on follow up imaging, residual changes found include persistent disc calcification, flattening of the vertebral body, and osteophytes.

Table 1: Summary table for intervertebral disc calcification in the pediatric population.

Calcium pyrophosphate dihydrate disease/Pseudogout	Radiograph and CT: extensive, often destructive, intervertebral disc calcification
Ochronosis / Alkaptonuria	Radiograph and CT: multilevel intervertebral disc calcification
Amyloidosis	Radiograph: multilevel intervertebral disc calcification
Osteochondroma	Radiograph and CT: continuity between the cortex and medullary region of the bone MR: mixed signal characteristics depending on the overall size of the lesion, and amount of calcification and marrow within the lesion
Osteoma	CT: lytic lesion with centralized calcification and a surrounding sclerotic rim MR: heterogeneous lesion with possible inflammation in adjacent structure

Table 2: Differential diagnosis table for intervertebral disc calcification in the pediatric population.

ABBREVIATIONS

IVDC = Intervertebral disc calcification
KFS = Klippel-Feil Syndrome
OPLL = Ossification of the posterior longitudinal ligament

KEYWORDS

Intervertebral disc calcification; Klippel-Feil Syndrome; Cervical spinal stenosis; Ossified posterior longitudinal ligament; Computed Tomography