CASE REPORT

Disappearing large calcified thoracic disc herniation in a patient with thalassaemia

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SUMMARY

Regression of herniated disc fragments with subsequent improvement in clinical symptoms has been reported in the lumbar and cervical spine. Such regressions in the thoracic spine are extremely rare. We report a case of a 38-year-old patient with thalassaemia who had regression of a large calcified herniated thoracic disc causing cord compression, with subsequent herniation of a second calcified disc at a different level and discuss the possible aetiopathogenesis. This is the first such case reported in the thalassaemia population.

BACKGROUND

It is known that disc degeneration and calcification is more common in patients with thalassaemia than in the general population.¹ Regression of herniated disc fragments with subsequent improvement of clinical symptoms, usually in the lumbar and cervical spine, has also been well reported. Such regressions in the thoracic spine are extremely rare. We report an interesting case of regression of a large calcified herniated thoracic disc with cord compression, with subsequent herniation of a second calcified disc at a different level, and discuss the possible aetiopathogenesis. This is the first such case reported in the thalassaemia population.

CASE PRESENTATION

A 38-year-old woman with β -thalassaemia major presented to our office in November 2010, with significant upper back pain. On clinical examination, the patient showed thoracic tenderness; however, motor strength, sensory examinations as well as deep tendon reflexes were normal and myelopathy was absent. The patient indicated to suffer from fatigue and arthralgia.

At 2 years of age, the patient had been diagnosed with β thalassaemia major and underwent multiple blood transfusions thereafter; in addition, iron chelation therapy was given due to iron overload. A liver biopsy indicated some degree of fibrosis and a FerriScan showed secondary haemochromatosis. The patient had a total splenectomy and her surgical history was positive for appendectomy and cholecystectomy. Her family history was positive for a brother suffering from thalassaemia.

INVESTIGATIONS

A CT scan was performed, which indicated a large calcified disc herniation at T6–7 level and a non-herniated calcified disc at T7–8 (figure 1A, B).

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes:

- ▶ Hard disc herniation
- ▶ Calcified disc
- ▶ Infectious aetiology
- ▶ Malignancy

TREATMENT

Owing to significant cord compression in the thoracic spine, surgical decompression was recommended. However, while the patient was waiting to decide on the surgery, her symptoms improved with conservative treatment management. Therefore, no surgical intervention was required.

OUTCOME AND FOLLOW-UP

The patient appeared for follow-up at our clinic in September of 2012, after her back pain was aggravated secondary to a slip and fall. She continued to remain neurologically intact. Interestingly, her MRI at this time revealed a new herniation of the previously identified non-herniated calcified disc at the T7–8 level while the previously seen calcified disc herniation at T6–7 was no longer visible (figure 2A, B). Our initial reaction on seeing the new imaging was that it was a spinal level localisation error, but repeated counting of the levels utilising the first and last ribs as reference confirmed that the level of pathology had indeed changed from T6–7 to T7–8.

Seven weeks later, a follow-up CT scan demonstrated progression of the calcified disc herniation at T7–8, which had now extruded and confirmed the disappearance of the calcification of the T6–7 disc (figure 3). The subsequent MRI correlate scan 1 week later confirmed these findings. The new central disc herniation at T7–8 demonstrated less cord compression than was seen initially with the T6–T7 disc herniation (figure 4); localisation of levels was confirmed by counting of the first and last ribs.

Since this new herniation was smaller than before and the patient remained neurologically intact, we decided in favour of close observation.

DISCUSSION

Spontaneous regression of disc herniation is well known to neurosurgeons. This, however, happens more commonly with soft disc herniations where the protruding nucleus pulposus either degenerates or is resorbed over time, leading to remission of symptoms. Large calcified disc herniations are very unlikely to resorb, especially in the thoracic spine. In a series of 858 patients, Martinez-Quinones *et al*² found that 37 soft disc herniations regressed with time and only one of them was in the thoracic spine.

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Figure 1 (A) Initial presentation of upper back pain. Right paramidline and midline sagittal non-contrast CT images of the thoracic spine demonstrate a calcified extruded disc at T6/T7 and a calcified non-herniated disc at T7/T8. (B) Initial presentation of upper back pain. Non-contrast 3 mm axial images of the thoracic spine confirm the large right paracentral calcified disc extrusion at T6/7 and the calcified non-herniated disc at T7/8.

There is only one, well-documented (by CT and MRI) case in the English literature of spontaneous regression of a symptomatic calcified thoracic disc herniation.³ This patient was a 36-year-old woman who was initially thought to have a calcified thoracic meningioma but was later presumed to have a herniated disc due to spontaneous regression of most of the lesion and concurrent improvement in symptoms. We came across three other reported cases of regression of thoracic disc herniation in the world literature (reported in Spanish and French journals).

Martinez-Quinones *et al*² reported on a 47-year-old man who had a regression in his T6–7 disc herniation. It is not clear from the report if this patient had partial or complete regression, or if the disc was calcified. In France, Coevoet *et al*⁴ reported a MRI documented regression of a large thoracic disc herniation after 1 year. Morandi *et al*⁵ reported on a 55-year-old man who had a CT and MRI documented regression of his thoracic disc after 6 months of conservative treatment. It is unclear if any of these three reported cases were calcified.

Figure 2 (A) Almost 2 years later, thoracic back pain was aggravated by a slip and fall. Sagittal T1 and T2 non-contrast MRI of the thoracic spine demonstrate resolution of the previous herniation and calcification at the T6/T7 disc with residual degenerative signal (low T2). There is, however, new herniation of the previously calcified disc at T7/T8, which demonstrates low T1 and T2 signal. (B) Almost 2 years later, thoracic back pain was aggravated by a slip and fall. Axial non-contrast T2-weighted MR images of the thoracic spine at T7/T8 demonstrate a small right paracentral disc herniation without cord compression.

Figure 3 Follow-up non-contrast sagittal and 2.5 mm axial CT images were obtained of the thoracic spine 7 weeks later. The sagittal image confirms disappearance of the calcification at the T6/T7 disc and demonstrates progression of the calcified herniated disc at T7/T8, which is now extruded. Axial images at, above and below the T7/T8 disc demonstrate a moderate calcified central disc protrusion.

In general, thalassaemia syndromes are related to decreased or lack of production of one of the globin chains within haemoglobin leading to a hypochromatic microcytic anaemia. Anaemia itself is theorised to precipitate degenerative changes in the intervertebral discs. Additionally, the anaemia necessitates multiple blood transfusions leading to excessive iron deposition—causing the production of damaging free radicals—which accelerates degenerative disc disease. Iron overload is also known to affect joint cartilage and synovium, especially of large joints. Spinal abnormalities are known to occur in patients with thalassaemia, haemochromatosis and other iron excess states, as well as a result of iron chelation therapy.^{1 6–8} Haemochromatosis often leads to chondrocalcinosis, although this is uncommon in thalassaemia. Iron chelation therapy, such as deferoxamine,

which is used to combat the deleterious effects of iron overload, can itself sometimes adversely affect the spine due to interference with mineral metabolism of enchondral ossification and/or possibly direct toxicity.⁷

Aessopos *et al*⁶ reported intervertebral disc calcification (IDC) in 7 of 30 (23.33%) β -thalassaemia intermedia patients. The patients with IDC had more back pain but otherwise had similar laboratory parameters (of iron excess, extramedullary haematopoiesis, etc) compared to those without IDC. They suggested the possibility of early iron chelation therapy in order to prevent complications from IDC. Incidence of IDC in the general adult population is approximately 5–6% and approaches 45% in those over the age of 85 years.⁶ Presence of IDC in this patient was likely secondary to her underlying condition and

Figure 4 Non-contrast MRI 1 week later confirmed the calcified extruded disc at T7/T8, not significantly changed from the CT. Again, there is degenerative, non-calcified signal at T6/T7, unchanged since the MRI 8 weeks prior. Axial images demonstrate the calcified central protrusion, which indents the ventral cord without significant cord compression or abnormal signal.

not to iron chelation therapy, as treatment was initiated after imaging documentation of two levels of IDC.

Several authors have investigated the causes of back pain in thalassaemia. Symptoms have been ascribed to expansion of bone marrow causing pressure on cortical bone, scoliosis, disc degeneration, osteoporotic compression fractures and extramedullary haematopoiesis, which may result in spinal cord compression, IDC and deferoxamine-induced bone dysplasia.^{1 9–11} Hartkamp *et al*⁷ reported spinal deformities in 22 patients with β -thalassaemia treated with deferoxamine. Two of these patients had asymptomatic IDC while six patients had narrowing of the disc space. Interestingly, one of these patients presented at age 7 years 11 months, with IDC at levels T1–2 and T3–4, which had resorbed, as indicated by the radiograph taken 4 years 9 months later and new calcification had developed at levels T4–5 and T5–6. This is somewhat similar to our case where calcification disappeared at one level, only to reappear at another. However, unlike in our case, these discs were not herniated and the patients in this study were asymptomatic.

Various theories have been put forward for spontaneous regression of disc herniations. These include re-accommodation of the herniated nucleus pulposus in the disc space, dehydration and desiccation of the disc, inflammatory reabsorption of the herniated material, pressure from cerebrospinal fluid pulsations and role of epidural venous plexus in its res.^{2 12} In comparison to a rare occurrence in adults, idiopathic IDC in children tends to regress in 61% of cases. This is attributed to richer vascular supply of the paediatric intervertebral disc and a more active metabolism. It is possible that the patient in this case, at age 38 years, may still benefit from some of these qualities. Also, in this case, it could be postulated that herniation of the IDC facilitated its resolution in keeping with the above theory of reabsorption. In a period of 2 years, the herniated IDC at T6/7 disappeared, while the originally non-herniated IDC at T7/8 persisted and subsequently herniated. This theory could be tested with delayed follow-up imaging in approximately 18 months.

We believe that, in our case, deposition of calcification in the intervertebral disc spaces at T6/7 and T7/8 on initial presentation was related to the patient's underlying thalassaemia. It is unclear what caused the initial disc herniation at T6/7 but the subsequent IDC herniation at T7/8 was likely related to the reported history of the patient's 'slip and fall'. The aetiology of the disappearance of the calcification and herniation at T6/T7 is unclear. Possibilities include that the patient is privy to an active metabolism and good vascular supply to the disc—as is documented in children—or that her findings support the hypothesis of resorption similar to that seen in non-calcified herniations. Less likely is the possibility of resolution secondary to the patient's iron chelation therapy, as the literature usually sites adverse affects on the spine. The findings in our case suggest that, in patients with thalassaemia without a myelopathy, conservative therapy should be considered along with continued MR surveillance since the calcified discs may resolve on their own without surgical intervention.

We report an extremely unusual case of complete regression of a calcified thoracic herniated disc with cord compression at

one level, with subsequent herniation of a calcified thoracic intervertebral disc at a different level. Various theories have been put forward to explain the increased incidence of IDC in patients with thalassaemia, but it is unclear if there is a predisposition to herniation and what would cause the regression of calcified disc herniation. Our patient's pathophysiology may be related to that postulated for non-calcified herniated disc and/or an incidental rich vascular intervertebral disc supply and metabolic system. Clinical follow-up and delayed follow-up imaging will be of value. It is important to note that no surgical treatment was needed because of the spontaneous resolution of the disc herniation.

Learning points

- Calcification of an intervertebral disc can be a complication of haemochromatosis.
- Transfusion-dependent β -thalassaemia, with associated iron overload, can result in such calcification of intervertebral discs.
- Observation of calcified discs might be sufficient if patients do not show any neurological impairments.

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