Paediatric spinal Langerhans cell histiocytosis requiring corpectomy and fusion at C7 and at Th8–Th9 levels

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SUMMARY

An 11-year-old girl was treated by corpectomy and anterior fusion because of the destruction of the C7 vertebral body. Pathological studies were not conclusive. The outcome was excellent, but 18 months later, she required thoracic corpectomy with anterior fusion owing to the impending kyphotic fracture of the Th8 vertebral body. Langerhans cell histiocytosis was now recognised and chemotherapy was given. 3 years later, the disease appears well controlled with normal shape of both the operated vertebral levels and maintenance of the movements of the adjacent vertebrae.

BACKGROUND

Spinal Langerhans cell histiocytosis (LCH) is rare in childhood and rarely requires surgical treatment. The literature includes very few patients managed by cervical corpectomy and there are just two cases treated by thoracic corpectomy. No patient needing corpectomy at both cervical and thoracic levels has been previously reported. As a consequence of its rarity, very few papers focus on the management of paediatric spinal localisations.

CASE PRESENTATION

An 11 year-old girl from Morocco was admitted to our emergency room with severe neck pain and without history of trauma. Neurological examination was unremarkable. Anamnesis was positive for lung tuberculosis. One year before, at another hospital, the asymptomatic collapse of the Th9 vertebral body had been occasionally diagnosed and interpreted as a stable consequence of healed Pott's disease.

INVESTIGATIONS

Neuroradiological assessments (figure 1) showed the osteolytic destruction of the C7 vertebral body with posterior listhesis of C6. The collapsed body was surrounded by enhancing pathological soft tissue as far as the epidural space. No sign of myelopathy was evident. The known collapsed Th9 body appeared stable, without any sign of cord damage, spinal kyphosis and/or spinal instability.

DIFFERENTIAL DIAGNOSIS

The neuroradiologist listed the possible diagnoses in the order of probability: Pott's disease, lymphoproliferative disease, LCH and metastasis of unknown origin. Chest x-ray and laboratory investigations were completely normal. The tuberculin skin test was intensely positive (TST). Abdominal

ultrasounds and bone scintigram did not show any other lesion.

TREATMENT

Through an anterior cervical approach, C7 corpectomy was performed with excision of scarred pathological tissue, body reconstruction and interbody fusion by autologous iliac strut graft fixed by mini-plate and screws. Histological examination did not show any neoplastic or granulomatous tissue, but revealed a subacute inflammatory process with necrotic areas and marked osteolysis caused by scattered osteoclast reabsorption. Microbial staining and cultural examinations resulted negative. A general histological diagnosis of inflammatory/infective spondylitis was given.

OUTCOME

Postoperatively, the neck pain completely resolved. The patient was discharged in a few days. One year later, she was quite asymptomatic and radiograms showed the good evolution of the arthrodesis (figure 2). However, 18 months after surgery, she came back because of acute dorsal pain.

Figure 1 Initial radiological assessments: (A) cervical radiogram (lateral view) showing a *vertebra plana* at C7 level; (B) CT scan (sagittal reconstruction) showing the destroyed C7 body surrounded by pathological soft tissue; (C) T2 weighted MRI (sagittal view) showing the listhesis at C7 level (white arrow) and the known collapse of the Th9 body (white arrowhead) which appears stable and without kyphotic deformity.

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Figure 2 Follow-up cervical radiogram obtained 1 year after C7 corpectomy showing the optimal degrees of fusion and stabilisation without any residual deformity.

Neurological examination was still unremarkable, but a clear kyphotic posture was evident. Neuroradiological studies showed that the known injured Th9 vertebra was grossly unchanged, but the adjacent Th8 vertebra was now collapsed with kyphotic deformity (figure 3). The intervertebral discs appeared uninvolved and no sign of myelopathy was evident. There was no

Figure 3 Readmission radiological assessments (18 months after cervical surgery): (A and B) CT scan (sagittal and coronal reconstructions) showing the impending fracture and collapse of the Th8 vertebral body adjacent to the known injured Th9 body; (C) T2-weighted MRI (sagittal view) showing the correct evolution of the previously operated cervical level (white arrow), the kyphotic collapse of the Th8 vertebral body (black arrow), and the known lesion of the Th9 vertebral body (arrowhead).

problem at the operated cervical site. Again, the differential diagnosis concerned Pott's disease and spinal LCH. Blood examinations, chest x-ray and abdominal ultrasounds were again negative while the TST was still intensely positive.

REOPERATION

Through a left-side thoracotomy, both the Th8 and Th9 vertebral bodies were removed and reconstructed using an expandable titanium cage filled by bone substitute. The kyphosis was corrected and the spine was fixed by anterior plate and screws. The pathological studies were quite similar to those of the first operation. This time, the study included immunochemistry which allowed the diagnosis of LCH since the CD-1A antigen and the S-100 protein were found. The first surgical specimen from C7 was re-evaluated and the diagnosis of spinal LCH was now obtained at this level too.

FURTHER OUTCOME AND FOLLOW-UP

The patient fully recovered and pain completely resolved. Further skeleton or visceral LCH localisations were excluded. One month after surgery, the patient underwent chemotherapy consisting of a combination of prednisone and weekly vinblastine for some months.

Follow-up radiograms showed adequate vertebral fusion with correct spinal alignment, and without any measurable limitation in spinal mobility (figure 4). Three years later, the patient was in perfect good health and further MRI showed the adequate evolution of both the operated levels (figure 5). The LCH appeared well controlled without any new manifestation.

DISCUSSION

Clinical aspects

Spinal LCH is generally rare.^{1–3} The differential diagnosis should include aneurysmal bone cyst, osteogenesis imperfecta, Gaucher's disease and neoplastic lesions.^{2 4–6} Some radiological patterns could assist the diagnosis of spinal LCH (symmetrical vertebral body collapse, maintenance of the disc spaces, absence of soft tissue masses^{1 6 7}) but isolate spinal LCH may represent a real diagnostic challenge.^{4 8}

Our patient did not present classical radiological features of LCH and, unfortunately, the immunochemistry was not used in the first surgical sample which undoubtedly represented a mistake. However, cases of spinal LCH with not-effective pathological diagnosis, and difficult differentiation between tuberculosis and spinal LCH have been reported also by others.^{7 9} Anyway, even if we had obtained the diagnosis of LCH since the first operation, we think the indication for chemotherapy would have been at least debatable in the presence of a single proved localisation.⁶

Indications for treatment

Most spinal LCHs are likely to require no treatment at all.^{4 8–10} The endochondral ossification remains classically undisturbed³ that means the vertebral body maintains the potential for the recovery of its height.^{1 4 6 11} Presently, radiation therapy is rarely advocated⁶ and chemotherapy is reserved for disseminated or aggressive diseases.^{4 6} Surgical treatment should be considered just in the uncommon cases of spinal cord compression^{2 4 5 8 9} asymmetric vertebral body breakdown with kyphosis or scoliosis^{3 6 8 12} and/or vertebral instability.⁷

Surgical techniques

Partial removal or simple curettage are rarely appropriate for spinal LCH.⁶ Since the LCH usually affects only the vertebral

Figure 4 Postoperative CT scan: (A and B) sagittal and coronal reconstructions showing the correct placement of the expandable cage and screws with good spinal alignment.

body,^{1 6} the anterior vertebral approaches result particularly suitable allowing relatively easy vertebral corpectomy, fusion and fixation by supplemental plate and screws.^{2-7 9 10 13 14}

Figure 5 Follow-up MRI (T2-weighted image) obtained, respectively, 5 and 3 years after cervical and thoracic surgeries showing adequate fixation and vertebral reconstruction at both levels.

Undoubtedly, the anterior routes to paediatric spine pose special problems especially for what concerns the size and the lack of dedicated stramentarium. However, in our experience,^{13 14} the modern versatile expandable cages and the new specifically designed plates usually permits optimal vertebral realignment and kyphosis correction. In general, in paediatric cases, autografts should be preferred^{3 8 10} but, if the patient is not too young, fusion/fixation at one/two levels may be easily tolerated, and metal instruments can be harmlessly and successfully used.^{2 3}

Review of similar published cases

The medical literature reports just a total of about 50 paediatric patients with spinal LCH requiring major surgery.^{1-10 12 15-18} Multiple localisations at different spinal levels are quite exceptional¹⁰ and paediatric patients with vertebral LCH requiring corpectomy and anterior fusion are quite uncommon.²⁻¹⁰ Almost all the reported anterior approaches to spinal LCH concern the cervical level and thoracic corpectomies have been previously reported only in two patients.^{4 9} Anterior approaches at two different levels (both thoracic) have been reported only in one patient but to perform curettage rather than corpectomy.⁶

LCH causing vertebral instability first at cervical level and later at thoracic level has been never described first. Accordingly, the patient we are reporting is really unique: she is the first to be successfully treated for spinal LCH using corpectomy and fusion at two different vertebral levels and at two different times.

Learning points

- ▶ Langerhans cell histiocytosis (LCH) should be always ruled out in case of spontaneous vertebral collapse in childhood.
- ▶ Paediatric spinal LCHs rarely require surgical treatment, but when this occurs, aggressive surgery and chemotherapy may be decisive.
- ▶ Since the LCH localisation, vertebral corpectomy with anterior fusion/fixation represents the best surgical option at any vertebral level even in children.

Competing interests None.

Patient consent Obtained.

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