Langerhans cell histiocytosis with multiple spinal involvement

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Langerhans’ cell histiocytosis (LCH)

- a clonal proliferation and accumulation of a specific histiocyte: the Langerhans’ cell
- LCH is manifested either localized (unifocal or multifocal) or systemic.
- Clinical variants of LCH includes:
  - eosinophilic granuloma (EG)
  - Hand–Schüller–Christian disease
  - Letterer–Siwe disease
The incidence of vertebral involvement varies between 6.5 and 25% of all skeletal LCH cases.

Multiple spinal LCH, including multifocal spinal lesions (lesions in two or more separated vertebrae) and/or solitary lesions (one lesion involving several consecutive vertebrae), is even rarely reported.

A search of the MEDLINE databank located 13 studies and totally 50 cases of LCH with multiple spinal lesions.

We reported five additional cases and a meta-analysis of all cases was also performed.
Results

- From 1997 to 2006, 42 consecutive patients with spinal LCH were diagnosed in our department.
- Only 5 (11.9%) had spine lesions involving multiple vertebrae
- 3 cases with consecutive lesions, 1 with separated lesions and 1 with combined (both consecutive and separated) lesions
- 4 male and 1 female, and the mean age at diagnosis was 18 (range: 12 to 23) years old
14 yrs boy, intermittent back pain and stiffness for 6 months. ESR 15 cm/h
CT and MRI revealed T11 vertebral plana and irregular T10 with slight regional kyphosis (Fig2 a-d) and lamina extension (Fig2 e & f)
LCH confirmed by percutaneous needle biopsy
The patient had immobilization and radiotherapy. One month after radiotherapy, his symptoms completely resolved.
The shape of collapsed vertebrae did not change at 57 month follow-up (Fig2 h-k)
A 23-yrs lady, low back pain for 2 weeks. ESR was 61 cm/h

Bone scan (Fig1 e) revealed intensely osteoblastic lesion at atlantoaxial spine, L3, left iliosacral joint, left humerus and left scapula. CT and MRI showed osteolytic lesion at S1 and L3 with epidural and paravertebral extension (Fig1 c,d,g)

Percutaneous needle biopsy of the S1 lesion under CT guidance stained positive for CD-1a and S-100.

The patient had chemotherapy and local radiotherapy. One month later, her pain completely resolved. She remained free of symptoms without disease recurrence at 36 month follow-up.
• 22 yrs man, neck pain for 2 years. The pain exaggerated in the past 20 days with left arm weakness (muscle power Grade 4/5). ESR was 41 mm/hr.
• CT and MRI manifested C4-6 vertebral body bony destruction with paravertebral and epidural extension (Fig3 a-e), which led to the suspected diagnosis of tuberculosis (TB).
• The patient underwent curettage and reconstruction. The postoperative pathological diagnosis was LCH.
• From retrospective view, this case should not been simply diagnosed as TB for the intact adjacent endplates and discs. He was symptom free at 61 ms follow-up.
Age and Sex

- Langerhans cell histiocytosis was usually regarded as a pediatric disease. LCH was reported to have a peak incidence between 5 and 10 years of age with male predilection.

- Recent studies have reported as many as 39% of diagnosed cases in adults.

- The gender distribution was nearly equal in children, whereas male was predominant in the adult cases.
Distribution

- Thoracic spine lesion was still the most common region (48.9%), and then came the cervical (27.5%) and lumbar spine (23.6%).
- The average number of involved vertebrae was 3, but it reaches 10 in one case.
- Extraskeletal disease was reported in 8-10% cases with spine LCH involvement, but it was 31.1% in this meta-analysis.
Prognosis

- generally fair. Depends largely on the visceral extension of lesions, although any effects of loading on the pathologic bone tissue must be taken into account.

- LCH have shown that resolution occurred at a rate unaffected by the mode of treatment.

- There is no clear evidence that treatment affects the natural history of this entity.

- Robert pointed out that the spinal complications in LCH are due more to the aggressiveness of the therapy than to the natural progression of the condition itself.
Proposed protocol

based on the results of this study and a review of the literature
Thanks for your attention