

Clinical features and treatment outcomes of Langerhans cell histiocytosis of the spine

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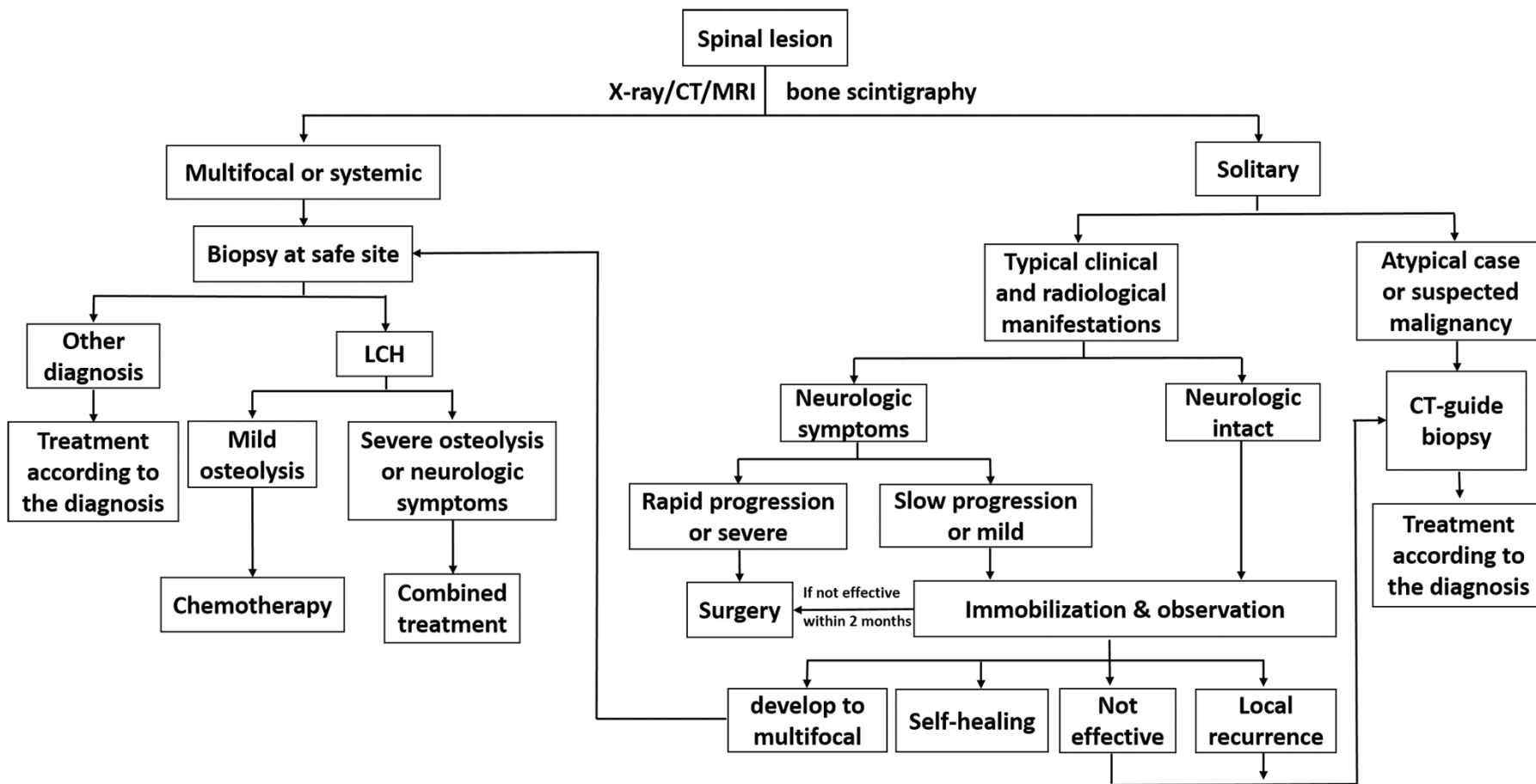
Background

- Langerhans cell histiocytosis (LCH) of the spine is a relatively rare condition with unknown etiology.
- The diagnosis and treatment protocols for spine LCH remain controversial.
- We evaluated the efficacy and safety of our proposed diagnosis and treatment protocol introduced in 2009.

Methods

- We retrospectively reviewed 110 patients with spine LCH who had been diagnosed and treated in our hospital from October 1997 to November 2015.
- The indications for Computed tomography (CT)-guided biopsy and surgery for spine LCH have become more stringent since 2009.
- In cases of a solitary spinal lesion, immobilization and/or observation were usually first suggested.
- Chemotherapy was suggested for cases with multifocal LCH lesions, and low-dosage radiotherapy was restricted to recurrent solitary lesion.
- Our indication for surgery: only for patients with muscle strength grade less than 3/5, rapid deterioration of neurological function within 2 weeks, persistent neurological symptoms over 2 months, severe deformity, and/or segmental instability.

Our Treatment Protocol



Results

- This series included 69 male and 41 female patients (age range, 1–52 years).
- Pain was the most common symptom (93.6%, 103/110).
- Pathologic diagnosis was achieved in 72 cases (65.5%).

Results

- CT-guided biopsies were performed in 91.3% (42/46) and 73.2% (41/56) of cases before and after 2009, respectively ($P = 0.02$).
- Immobilization and/or observation were performed in 25.9% (14/54) and 75.0% (42/56) cases before and after 2009, respectively ($P < 0.001$).
- Approximately 35.2% (19/54) and 10.7% (6/56) cases had surgery ($P = 0.002$) before and after 2009, respectively.
- Ninety-eight cases (89.1%) were followed up for a mean 66.3 (range, 24–159) months.
- During the follow-up, no significant difference was found in the outcomes between the two groups treated before and after 2009 ($P = 0.64$).

2 year old boy

multiple LCH lesions in left C1 lateral mass, sternum and bilateral ilium.

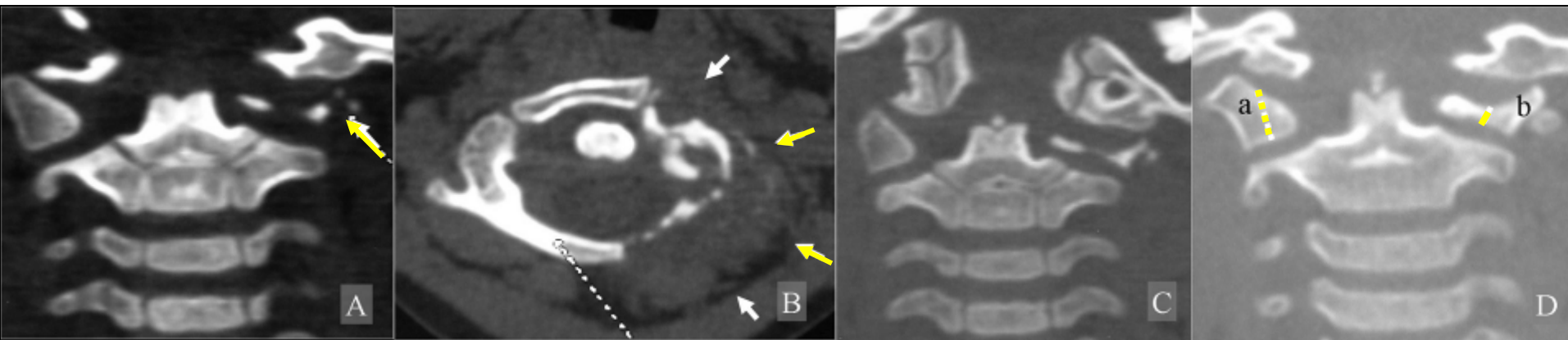
neck pain for 6 months

immobilization for 2 months and chemotherapy for 2 years.

at diagnosis

6 month

44 month



**lateral mass to transverse process,
posterior arch and paravertebral muscle**

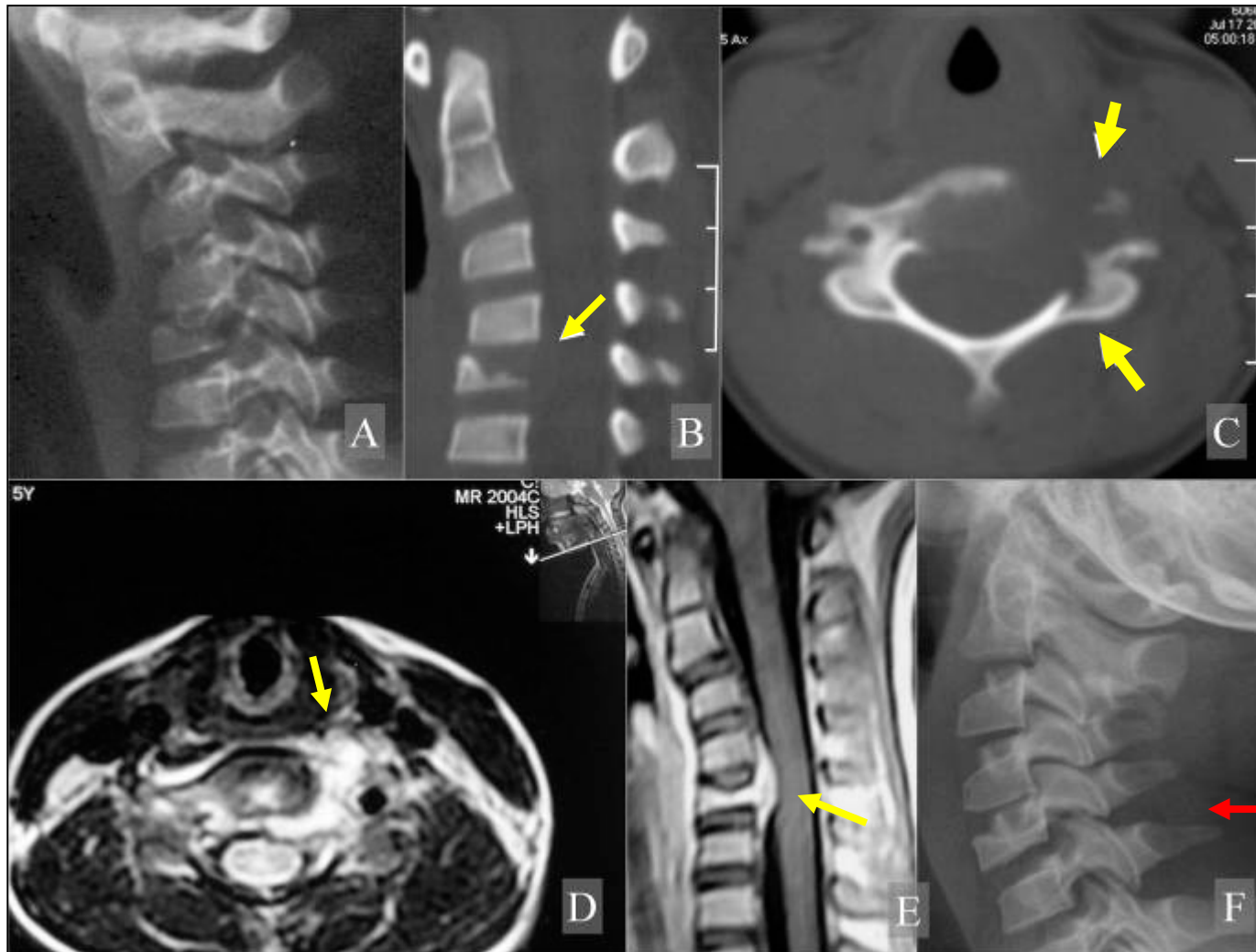
slight remodeling

**height ratio
improved 34.4%
($b/a \times 100\%$).**

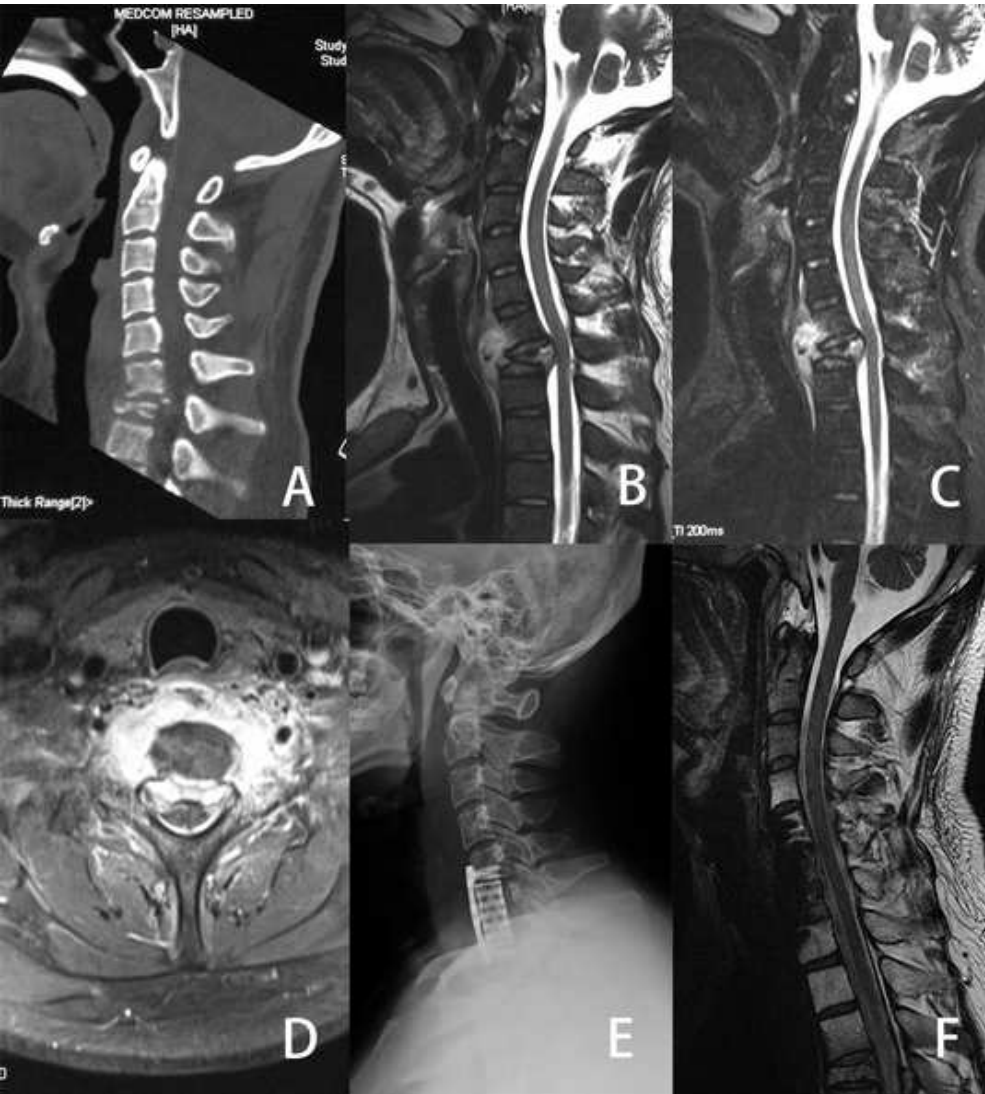


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5 year old boy with solitary C5 lesion, symptom spontaneous disappearance with immobilization for 4 weeks



Case3



A 29-year-old man presented in 2012 with neck pain for 2 months and upper extremity numbness. A skeletal survey revealed solitary C6/7 bony destruction with paravertebral extension, which was diagnosed as Langerhans cell histiocytosis by computed tomography-guided biopsy. With immobilization, his neck pain was relieved, while the sensory loss of his upper extremities persisted. After 5 months, his Langerhans cell histiocytosis developed to multifocal, involving T4 and L5 vertebrae, right temporal bone, and 3rd and 7th right ribs. Curettage of C6/7 was carried out with fusion and instrumentation followed by chemotherapy. (A) Sagittal computed tomography showed bony destruction of C6/7 vertebrae with cervical kyphosis. (B, C) Sagittal T2-weighted magnetic resonance imaging and magnetic resonance imaging with contrast demonstrated spinal cord compression. (D) Axial magnetic resonance imaging revealed paravertebral and epidural soft-tissue extension. (E) Lateral radiography after surgery. (F) At 48-month follow-up, the patient was symptom-free, sagittal T2-weighted magnetic resonance imaging showed no local recurrence.



Conclusions

- Biopsy is not mandatory for typical spine lesions of LCH.
- Given the self-healing tendency of spine LCH, immobilization and/or observation remain the first-choice treatments for LCH lesions.
- Conservative biopsy and treatment protocols might be more appropriate for spinal LCH.
- Surgery indication should be more stringent.



Our Spine Tumor MDT



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Disclosure declaration

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