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The Spine Journal 14 (2014) 768–776

THE SPINE JOURNAL

Clinical Study

Surgical treatment based on pedicle screw instrumentation for thoracic or lumbar spinal Langerhans cell histiocytosis complicated with neurologic deficit in children

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Received 7 September 2012; revised 19 March 2013; accepted 29 June 2013

Abstract

BACKGROUND CONTEXT: Surgical indications and procedures for spinal Langerhans cell histiocytosis (LCH) in children are still controversial. Reports containing large samples of surgically treated patients are few in the currently available literature, and the reported operative procedures were also somewhat obsolete. So, further investigation based on large-sample cases and using improved surgical techniques is beneficial and helpful to refine the treatment strategy.

PURPOSE: To recommend a reasonable treatment strategy for thoracic or lumbar spine LCH in children complicated with neurologic deficit.

STUDY DESIGN/SETTING: Retrospective/academic medical center.

PATIENT SAMPLE: Twelve children aged from 2 to 16 years old with the diagnosis of thoracic or lumbar spinal LCH accompanied by neurologic deficit received surgical treatment from January 2005 to January 2010.

OUTCOME MEASURES: Frankel scale for neurologic function, fusion of the mass, and recurrence of the lesion.

METHODS: All 12 patients presented initially with local pain and progressive neurologic detriment. Neurologic evaluation revealed two patients with Frankel Grade B, eight with Grade C, and two with Grade D. Radiographic features were positive for typical vertebra plana, a space-occupying mass in the spinal canal compressing neural elements, and a spinal canal encroachment rate more than 50%. Posterior instrumentation with pedicle screw combined with anterior corpectomy, decompression, and support bone graft was performed in the first seven patients as a one-stage procedure. In the remaining five patients, posterior pedicle screw fixation, laminectomy for decompression (via excision of the tumor-like mass), and repair of laminae with allograft bone block were performed. The collapsed vertebral body was left untouched. No chemotherapy or radiotherapy was administrated postoperatively in any of the cases.

RESULTS: The mean follow-up duration was 43.3 months. The mean operation time was 330 minutes with combined procedure and 142 minutes with single posterior approach (p=.000). The average blood loss was 933 mL with combined procedure and 497 mL with single posterior approach (p=.039). Three of seven patients who received combined surgery encountered approach-related complications, that is, one with intercostal neuralgia and two with pleural effusion. No severe neurologic deteriorate, instrumentation failure, or disease recurrence was detected at follow-up. Neurologic function completely recovered in all 12 patients from 2 to 12 weeks after surgery. The anterior bone graft fused and shaped well in all seven patients, and allograft bone block for lamina repair also achieved complete fusion in the remaining five patients. The internal fixator was removed at 3 to 5 years (average 4.1 years) after initial operation in six patients. No deformity, including scoliosis and kyphosis, has been identified during follow-up period in both procedures.

FDA device/drug status: Not applicable.

Author disclosures: ***G-HL:*** Nothing to disclose. ***JL:*** Nothing to disclose. ***X-BW:*** Nothing to disclose. ***BW:*** Nothing to disclose. ***KP:*** Nothing to disclose.

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1529-9430/$ - see front matter
http://dx.doi.org/10.1016/j.spinee.2013.06.104

CONCLUSIONS: For spinal LCH patients, neurologic deficit is a main indication for operative treatment to prevent permanent and serious consequences. Surgery provides an opportunity for rapid recovery of neurologic function. Both combined and single-stage posterior approaches based on pedicle screw instrumentation techniques are similarly effective in relieving neurologic compression. However, single-stage posterior approach is more favorable with less complications, and preserving involved vertebral body is not a latent hazard of recurrence.

Keywords: Langerhans cell histiocytosis; Children spine; Neurologic deficit; Surgical treatment; Pedicle screw instrumentation

Introduction

Langerhans cell histiocytosis (LCH) is a lesion that results from the abnormal reticuloendothelial system. In most cases, LCH is not an aggressive neoplasm. The natural history is often self-limited and resolves spontaneously, especially in children and young adults [1–3]. The most vulnerable population are children under 10 years old, and the spine is one of the commonly involved location of the lesion. Although the growing spine is involved, major kyphotic or scoliotic deformity is seldom seen because the lesion usually involves the whole vertebral body incurring nearly symmetrical destruction and vertebral collapse; moreover, asymmetrical collapse also has nothing to do with deformity [4–6]. Treatment principles for vertebral LCH have been variable. Without neurologic deficit, authors trend to conservative treatment method including simple observation, bracing immobilization, steroid, or radiation [7,8]. Some have advocated that immobilization and radiation are also appropriate in children with mild neurologic deficit [7,9]. However, there are a minority of patients who suffer this kind of disease with more severe neurologic deficits because of spinal cord or cauda equina compression. We believe that the neural elements should be decompressed and then fused; as until now, there is no clear evidence to prove that neurologic status will stop deteriorate or improve with conservative treatment [6,10]. So, for the sake of neurologic safety, operative treatment becomes inevitable.

As far as we know, few studies focus on the surgical treatment of LCH. There is no standardized guideline established regarding the aspects of the operation, including surgical approaches (bone grafting, corpectomy, and others) and instrumentation methods. The existence of controversies and uncertainties in operative treatment gives us impetus to present our 12 cases to share with others. In this study, we reveal the outcome of vertebral LCH complicated with neurologic deficit in children at a single spine center and give the recommendation for surgical treatment.

Materials and methods

Twelve children aged 2 to 16 years (average 9.58 years) were admitted to our spine surgery center from January 2005 to January 2010 with the diagnosis of thoracic or lumbar spinal LCH complicated by neurologic deficit, including four females and eight males. With the approval of the institution's institutional review board, demographic data, medical record, and radiographic findings of these patients were reviewed thoroughly (Table). The most common initial symptom was local dorsalgia that was exacerbated by movement. The lateral decubitus position was found to be the most comfortable position for the patients. Diabetes insipidus has not been encountered in any patients. Neurologic symptoms, on average, occurred about 2 to 4 weeks after the initial dorsalgia. The time from onset to admission was 1 to 2 months. Neurologic evaluation revealed Frankel Grade B function in two patients, Grade C in eight, and Grade D in two. Preoperative diagnosis was mainly based on symptoms and laboratory investigations such as complete blood count, erythrocyte sedimentation rates, and radiographic features (vertebra plana, disc space maintained). Owing to the relatively emergent neurologic situation of these patients, preoperative biopsy has not taken so as not to delay first-rate surgical opportunity.

Laboratory tests

The results of a complete blood count, complete metabolic profile, and urinalysis were normal. Erythrocyte sedimentation rate elevated in eight cases ranging from 25 to 49 mm/h but with normal C-reactive protein.

Magnetic resonance imaging characteristics

The involved vertebrae were T3 (2), T4 (1), T8 (2), T11 (1), T12 (1), L1 (3), L2 (1), and L3 (1). Another focus was detected in the skull of two patients by roentgenograph. In 10 patients, the involved vertebrae were evenly collapsed and the two end plates came into contact with each other, resulting in the typical coin-shaped vertebra plana. In the rest of the two patients, the anterior half of the involved vertebra completely collapsed, whereas the posterior half maintained its height, resulting in an obvious wedge-shaped and medium kyphosis. All patients were positive for typical vertebra plana, a space-occupying mass in the spinal canal compressing neural elements, and a canal encroachment rate more than 50% via magnetic resonance imaging. Abnormal soft tissue surrounding the vertebral body in transverse section and prespinal mass in sagittal