

## Conservative Treatment in a 6 Year Old With Extensive Eosinophilic Granuloma Involving the Craniocervical Junction.

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### Introduction

Langerhans Cell Histiocytosis(LCH) is a rare disease that results from the aberrant proliferation of dendritic cells which can infiltrate the spine resulting in instability. In this report, we present a patient with lesions involving the C1, C2 and occipital condyle. Despite previous reports of treatment by surgical curettage and fusion, we report a favorable outcome using chemotherapy and halo-thoracic brace fixation.

### History and Examination:

A 6 years old girl presented with sudden onset left sided neck pain and torticollis. She has cervical lymphadenopathy. There was no neurological deficits.

### Investigations:

CT and MRI of the craniocervical junction showed multiple lytic and expansile lesions involving the right occipital condyle, clivus, C1 lateral mass and C2 odontoid process resulting in atlanto-axial and occipital-cervical subluxation. Biopsy of cervical lymph node showed LCH, which stained positive for CD1a. Bone marrow trephine did not show any tumour involvement.

### Halo-thoracic brace fixation

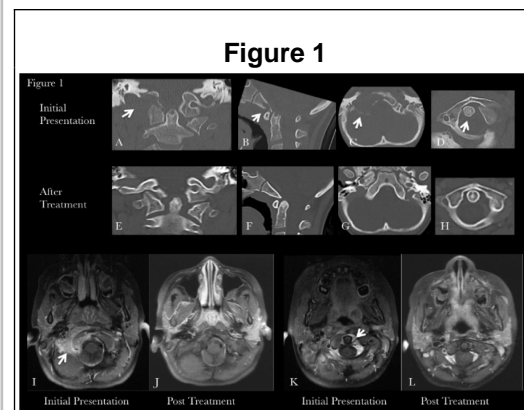
Her atlanto-axial subluxation was reduced awake under fluoroscopic guidance, followed by halo-thoracic brace fixation. Subsequent CT showed satisfactory reduction of the subluxation.

### Chemotherapy

She was commenced on Vinblastine, which was stopped after 2 cycles due to lack of response on interval imaging. It was then switched to Cladribine with good response, and this was continued for a total of 7 cycles. Post treatment MRI showed complete resolution of the tumours.

### Outcome

Her brace was removed after 10 months and she regained nearly full range of neck rotation with no neurological deficits. Post treatment CT scan showed near complete reconstitution of the bones which were initially destroyed by LCH.



**Figure 1:** A-H: CT images showing the coronal, sagittal and axial views of C1, C2 and occipital condyle pre and post-treatment. The lytic lesions in the occipital condyle (A,C) and clivus (B) resolved with near complete reconstitution of the bone (E,F,G). The atlantoaxial subluxation (D) resolved by the end of treatment (H). I-L : MRI with contrast showing the resolution of contrast enhancing lesions in the occipital condyle (I,J) and around the transverse ligament (K,L)

### Discussion:

Our patient had LCH lesions causing craniocervical junction instability. The traditional approach of surgical excision, fixation and fusion would result in limited range of neck rotation, which is not desirable especially in children. Our patient did not have spinal cord compression, therefore surgical decompression was not indicated. Histology was established from biopsy of affected cervical lymph nodes. Her occipital-cervical subluxation and C1/C2 subluxation were amenable to closed reduction, and she was fixed in a halo brace. Her LCH was treated primarily with chemotherapy and she responded well.

There were two important observations. At 10 months after treatment, it was noted that her ligamentous laxity has improved and the lytic bone lesions were replaced by normal bone assuming the normal anatomical conformation. Previous report by Ha 1 showed that vertebral bodies involved by LCH with collapse of height, when managed conservatively with orthosis resulted in bone reformation at osteolytic lesions. This was a well described phenomena. Some reports have even argued for long term immobilization in patients with spinal instability until sufficient bone reconstitution has occurred to restore stability 2. This was likely due to the immobilization which facilitates the reparative phase of bone healing as well as the preservation of the periosteum (otherwise removed during surgical curettage) which provided vascular supply and progenitor cells necessary for bone healing 4 .

Ligamentous laxity in our patient is caused by creep : the deformation and elongation of spinal ligaments as a result of the constant load applied by the expansive soft tissue component of the LCH tumor. We hypothesize that immobilization and chemotherapy removes the load applied on the ligaments allowing adequate time for the ligaments to undergo the tissue remodeling cascade a couple of months after injury 5. Furthermore, the early diagnosis means that the strain on the ligaments were relieved during the microfailure stage which provides it with the best chance for tissue remodeling.

### Conclusion:

Here, we demonstrated that conservative treatment with cervical immobilization and chemotherapy resulted in satisfactory outcomes in patients with LCH causing cranio-cervical junction instability. In this case LCH lesions regressed and bone lesions were reconstituted to restore normal anatomical morphology.

### References

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