Rib and sternal lesions in pediatric Langerhans cell histiocytosis: treatment and outcomes

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BACKGROUND

Langerhans cell histiocytosis (LCH) is a rare disorder characterized by proliferation and tissue infiltration of dendritic cells, most commonly occurring in children.[1] Though it can occur in any organ, it often manifests as skeletal lesions with rib and sternal involvement being common. Due to its ubiquitous nature, LCH lesions can be located in various sites or anatomical systems as either unifocal, located in one system and site, or multifocal, found in in one system but multiple sites.[2] Although evident in pediatric populations, few studies have characterized and reviewed the eventual outcomes after treatment of isolated rib and sternal LCH lesions, especially in regards to treatment and long-term sequelae.

PURPOSE

We aimed to characterize a series of unifocal and multifocal skeletal LCH lesions in the rib and sternum clinically and radiographically in order to determine the success and recurrence rates with different treatment modalities in a pediatric population at a tertiary children's hospital.

METHODS

A retrospective analysis found 11 patients younger than 18 years old with a diagnosis of unifocal or multifocal LCH lesions of the rib or sternum. Clinical presentations, lesion sites, additional skeletal lesions, biopsy site, radiographic findings, treatments, recurrence rates, and length of follow-up were reviewed and recorded.

Table 1. Classification, Treatment, and Outcomes

Case	Class	Treatment	Recurrences	Total follow-up
1	multifocal	Vinblastine, Prednisolone	0	1.4 years
2	multifocal	Vinblastine, Prednisone	0	4.8 years
3	multifocal	Cytarabine, methylprednisolone, methotrexate	1	7.7 years
4	multifocal	Vinblastine, Prednisone	2	3.4 years
5	multifocal	Cytarabine	0	1.4 years
6	unifocal	Wide resection with intralesional steroid injection	0	0.4 years
7	unifocal	Curettage with intralesional steroid injection	0	4.4 years
8	unifocal	Methylprednisolone injection	0	2.2 years
9	unifocal	Resection	0	2.2 years
10	multifocal	Vinblastine, methylprednisolone, methotrexate	0	13.2 years
11	multifocal	Cytarabine and clofarabine for new lesion	0	5.0 years

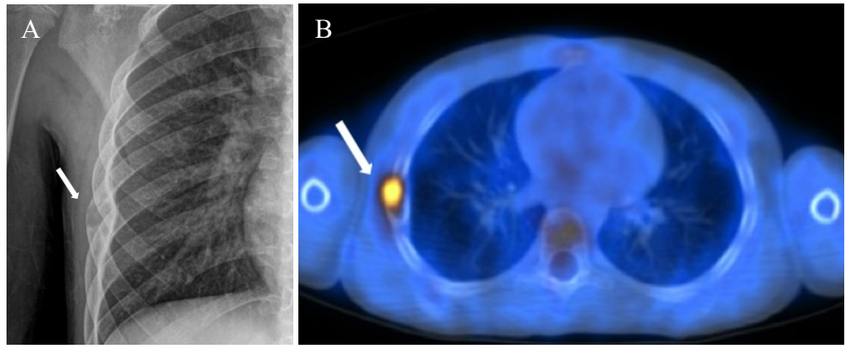

Figure 1. An eight year old female presenting with right sided rib pain (Case 7). (A) Anteroposterior radiographic scan demonstrating osteolytic lesion at the 6th rib. (B) Axial view of positron emission tomography (PET) scan demonstrating hyperintensity at the 6th rib.

RESULTS

Out of 686 patients queried for LCH diagnosis, 4 were isolated rib lesions and 7 were multifocal rib/sternum lesions. The most common radiographic finding was a lytic bone lesion. The most common clinical presentation was pain, with bone pain or deformity occurring in all patients with unifocal lesions. All unifocal lesions were treated locally via surgical resection plus intraoperative steroid injection or steroid injection alone. All other cases were multifocal and treated with chemotherapy. There were two cases of recurrence, with both occurring in the multifocal group. Median length of follow-up was 3.4 years (Table 1). There was no mortality recorded.

CONCLUSION

Surgical resection with intra-lesional corticosteroid injection is an appropriate option for unifocal rib and sternal LCH lesions. Rib lesions as part of a multifocal presentation may be managed adequately with chemotherapy. Resection and steroid therapy alone may be reasonable for pediatric single-system multifocal skeletal lesions that are anatomically accessible and small in number and size, in contrast with a full chemotherapy regimen and associated ill effects.

REFERENCES

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