Chest Wall Osteochondroma in Children: A Case Series of Surgical Management

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Background: Chest wall osteochondroma is a rare tumor in children. Even though the potential for malignant transformation or serious intrathoracic complications is low, it has led some centers to advocate surgical management of these bony tumors. We present our experience of the surgical management of costal osteochondromata.

Methods: Between January 1, 2006 and November 1, 2012 we saw 854 patients with solitary or multiple exostoses in our clinics. By reviewing our billing lists we found 7 children who had surgical management of chest wall osteochondromata. The indications for surgery were pain (3 patients), excision for confirmation of diagnosis (2 patients), recurrent pneumothorax (1 patient), and malignancy (1 patient).

Results: All patients made a good postoperative recovery with a median hospital stay of 1.8 days (range, 0 to 4 d). There was no recurrence of exostosis on follow-up (range, 8 mo to 2.6 y). One patient required surgery for excision of another chest wall osteochondroma at an adjacent location. No patient reported scar-related pain symptoms. No malignant transformation or intrathoracic complications occurred. We found ribs as the first site of presentation of multiple hereditary exostoses in 2 young patients.

Conclusions: Surgical management of thoracic osteochondroma, with excision for painful, symptomatic, malignant lesions or lesions adjudged to be at risk of intrathoracic complications, yields good outcomes in terms of symptom control, establishing histologic diagnosis, and prevention of thoracic complications.

Level of Evidence: Level IV—case series.

Key Words: osteochondroma, exostosis, chest wall

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Osteochondroma, also known as exostosis, is the most common benign bone tumor.[1] Osteochrondoma maybe solitary or can present as part of the condition of multiple hereditary exostoses (MHE).[2] MHE is an autosomal dominant condition associated with *EXT1* and *EXT2* genes mutations.[3–6] MHE in most cases involves the metaphyseal region of long bones, such as the femur or tibia, and usually presents after 2 years of age as multiple bony growths on the appendicular skeleton.[7]

Solitary chest wall osteochondromata are extremely rare.[2] They are usually a part of MHE[8] and constitute 1% to 1.5% of total cases of osteochondromata.[9,10] Although asymptomatic and diagnosed incidentally in most cases,[11] serious and even life-threatening complications can occur.[3,12–14]

Chest wall osteochondromata can present with local complications of cosmetic or physical discomfort,[15] intrathoracic complications of pneumothorax,[3,16] pericardial effusion,[17] hemothorax,[13,14,18–20] and consequent empyema[1] or with sarcomatous transformation of the osteochondromata.[4] These aforementioned serious complications have led some surgeons to advocate excision biopsy of chest wall lesions[21] and to perform preemptive resection for chest wall osteochondromata with bony spur.[1] In this study we report outcomes in 7 patients who had surgical management for the chest osteochondromata.

To our knowledge this is one of the largest studies for surgical management of chest wall exostoses published to date.

METHODS

After obtaining Institutional Review Board approval, we used our billing lists to identify all children who underwent surgery for chest wall osteochondromata at our institution between January 1, 2006 and April 1, 2012. Patients over 18 years of age and/or with postoperative follow-up of <6 months were excluded. Once identified, patients' electronic medical records were queried to obtain data: index and follow-up radiologic data; operative data including indications, complications, and recovery information; histologic diagnosis and information about pain; any recurrence and any evidence of malignant transformation on follow-up.

RESULTS

We saw 854 patients with solitary or multiple osteochondroma during the study period in our clinics. By running a query of our billing data we found 7 children, with >6 months of follow-up, who had surgical management of

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The authors declare no conflicts of interest.

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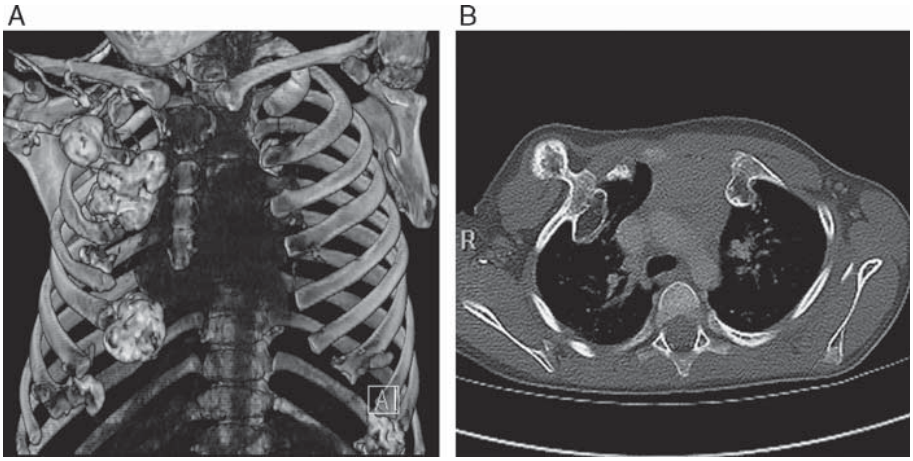

FIGURE 1. A, Three-dimensional computed tomographic (CT) scan showing multiple intrathoracic and extrathoracic osteochondromata. This patient had a known multiple hereditary exostoses diagnosis. The patient underwent multiple surgeries for symptomatic osteochondromata. During evaluation of painful osteochondromata of the chest wall we found these prominent intrathoracic and extrathoracic osteochondromata. B, Axial view CT scans showing intrathoracic and extrathoracic osteochondromata. As we expected based on this CT scan, intraoperatively we found several large osteochondromata along the anterior chest wall that abutted the heart and pericardium.

chest wall osteochondromata. The mean age of patients at the time of surgery was 9.7 years (range, 11 mo to 16 y), 4 were male (57%). The mean postoperative hospital stay was 1.8 days (range, 0 to 4 d). Five patients had MHE at presentation and 2 patients were found to have MHE at subsequent follow-up visits when they developed osteochondromata at other sites.

The various indications for excision biopsy were to remove painful bony masses in 3 patients (Fig. 1), to confirm the diagnosis of painless rib lesions in 2 patients, and to prevent another episode of recurrent pneumothorax in 1 patient (Fig. 2). Radical resection was indicated for chondrosarcoma arising from an osteochondroma in 1 patient. This patient who was nearing skeletal maturity and had a known MHE diagnosis presented with a painful growing mass over the anterior upper chest. The patient underwent incisional biopsy at another institution. He was diagnosed to have grade I chondrosarcoma and subsequently was referred to us for further management. Preoperatively a limited chest computed tomographic (CT) scan was obtained in addition to plain x-ray in all patients but the 2 younger ones.

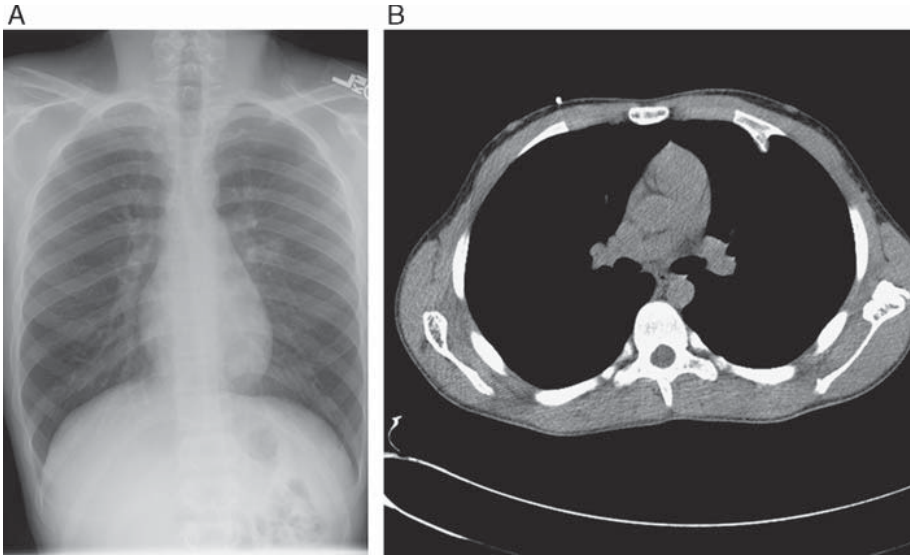

FIGURE 2. A, This chest x-ray shows no abnormalities in the patient who was treated for recurrent episodes of pneumothorax. B, This computed tomographic scan shows a pedunculated osteochondroma arising from the inner aspect of the left third rib as the cause of recurrent pneumothorax.

TABLE 1. Analysis of the Operative Patients

	Rib/ Laterality	Age at Presentation (y)	Indication for Surgery	Operation	Postoperative Stay (d)	Recurrent Osteochondroma During Follow-up	Duration of Follow-up (y)	Thoracic Complication During Follow-up
1	8/left	13.3	Painful rib exostosis	Excision	1	None	1.9	None
2	10/right	1.3	Excision biopsy due to parental concern	Excision	0	None	1.5	None
3	1/left	15.8	Chondrosarcoma of the rib	Wide resection of first and second rib	4	No, but developed a new fourth rib osteochondroma	2.1	None
4	8+9/left	16	Painful rib exostosis	Excision biopsy	2	None	1.3	None
5	3/left	15.3	Recurrent pneumothorax secondary to rib exostosis	Excision	3	None	2.6	None
6	8/right	0.9	Excision biopsy due to parental concern	Excision	0	None	2.6	None
7	Bilateral multiple ribs	5.5	Painful exostosis close to thoracic viscera	Excision	3	None	0.7	None

Because of the high dose of radiation, in the case of the 2 very young patients the procedures were planned based on physical examination and plain x-ray.

In 5 cases the osteochondromata were externally prominent. In the case with recurrent pnemothorax the osteochondroma was growing inwards toward the pleura. In the seventh case there were multiple extrathoracic and intrathoracic osteochondromata impinging on the pleura and the pericardium. All procedures were conducted through traditional open incisions. Procedures were performed by the orthopaedic surgeon in 3 cases and by the pediatric cardiothoracic surgeon in 2 cases. In 2 cases the procedures were performed by the orthopaedic surgeon in conjunction with the cardiothoracic surgeon.

For example, in the seventh case the cardiothoracic surgeon operated first with a median sternotomy. All the palpable large osteochondromata were removed from the inner surface of the chest walls. Then the procedure was turned over to the orthopaedic oncology team. At that point 2 separate short oblique incisions were made over the right chest wall to remove 2 symptomatic external osteochondromata. The asymptomatic osteochromata were not excised. A chest tube was inserted for 6 of the 7 patients at the end of procedure. There were no intraoperative or postoperative complications. We followed our patients for a mean duration of 1.8 years (range, 8 mo to 2.6 y) postoperatively. One patient had to undergo a second surgery for a new bony mass at the fourth rib. All

TABLE 2. Comparison of Various Case Reports for Pediatric Costal Osteochondromata

References	No. Patients	Mean Age (y)	Indication for Surgery	Operation	Complications	Duration of Follow-up (y)
Assefa[2]	1	14	Hemothorax	Excision by VATS	No	—
	1	15	Pneumothorax	Nonsurgical	No	—
	1	6	Hemothorax	Excision	No	—
Khosla[3]	1	17	Pneumothorax	Excision by VATS	No	0.3
Pham-Duc[9]	1	15	Hemothorax	Excision	No	—
Jin[12]	1	11	Hemothorax	Excision	No	—
Nakano[13]	1	15	Hemothorax	Excision	No	—
Huang[14]	1	9	Hemothorax	Nonsurgical	No	2
Cowles[17]	1	6	Hemothorax, pericardial effusion	Excision by VATS	No	—
Kim[15]	6	13.6	Pain and/or protrusion	Excision	No	—
Kuo[19]	1	15	Hemothorax	Excision by VATS	No	—
Asmat[20]	1	16	Hemothorax	Excision by VATS	No	—
Abdullah[21]	1	14	Diaphragmatic rupture and bowel obstruction	Exostosis excision and diaphragmatic	No	—
This study	7	9.7	Pain, confirmation of diagnosis, recurrent pneumothorax, malignant transformation	Excision	No	1.8

VATS indicates video-assisted thoracoscopic surgery.

patients remained symptom-free and none of the patients had a recurrence. None of the 7 patients developed new episodes of pneumothorax/hemothorax or malignant transformation. As all patients remained symptom-free during the follow-up period we did not obtain any post-operative images except in the following 2 cases. In the chondrosarcoma case, a CT scan was performed 6 months after surgery to investigate for any signs of a silent recurrence. We also obtained repeat magnetic resonance imaging (MRI) for the patient with multiple intrathoracic and extrathoracic osteochondromata after 6 months to see for recurrence. The individual patients' information is summarized in Table 1.

DISCUSSION

This case series of 7 patients who had surgical management of chest wall exostoses demonstrated 4 major patterns of presentation: the asymptomatic incidental finding, those with local pain symptoms without concern for thoracic complications, those with thoracic complications, and those with malignant transformation. This can be compared with other case reports as depicted in Table 2.

Chest wall tumors are rare in the pediatric population.[22] Approximately 50% of rib lesions in the pediatric age group are malignant.[15] The most common malignant lesion is Ewing sarcoma,[23] followed by osteosarcoma and chondrosarcoma.[15] Differential diagnoses of benign costal tumors, including but not limited to osteochondromata, aneurismal bone cyst, fibrous dysplasia, chondroma, and eosinophilic granuloma.[24]

Malignant transformation of a benign osteochondroma into a chondrosarcoma is a known complication of MHE.[4] The incidence of malignant transformation has been reported as high as 10% in MHE.[24] Chondrosarcoma is a slow growing malignancy and even recurrent cases can be managed by radical resection.[25] One of our patients had a grade 1 (low grade) chondrosarcoma and responded to surgery very well without any recurrence after 2.1 years of follow-up.

Pneumothorax is a frequently reported complication of costal osteochondroma in children, though it only made up one of our cohort, maybe due to mechanical interference with surrounding anatomic structures.[2,3,16] Involvement of the exostosis with the neurovascular bundle can also cause pain and hemothorax, the latter leading to empyema if suboptimally drained.[1]

The combined risk of thoracic complications and malignant transformation have led some centers to advocate careful follow-up[3] and the frequent use of excisional biopsy to exclude malignancy and in cases where thoracic complications are a concern.[21] Even after resection, recurrences might occur or a new osteochondroma could develop at a separate site, hence vigilance should be maintained. Our follow-up modalities included clinical examination, plain x-ray (Fig. 2), CT scan (Fig. 2), and MRI. Some authors would advocate for MRI as the best modality for more deeply located lesions like thoracic osteochondromata.[4]

Two patients in our study presented early in childhood (average age 1.1 y) with bony swelling of the rib with parental concern and anxiety. They did not have any bony swelling elsewhere at the time of surgery. However, these patients developed multiple osteochondromata over the next few years on follow-up. MHE usually presents after the age of 2 years as multiple bony growths on the appendicular skeleton.[7] Interestingly, ribs were the first site of presentation of MHE for these 2 young patients.

We recognize some limitations in our study. First, it is a retrospective study with its inherent limitations. Second, it would be more desirable if we could follow-up with our patients for a longer period of time.

In conclusion chest wall osteochondromata are typically asymptomatic. Asymptomatic cases do not need further evaluations unless they develop symptoms on regular follow-up. However, we conclude that they should always be given due consideration in patients with MHE presenting with pain or concerning physical examination findings. In these circumstances they should be evaluated with an x-ray and possible advanced imaging like CT scan or MRI. They should be considered in the differential diagnoses list for patients with spontaneous hemothorax or pneumothorax. The surgical excision of chest wall osteochondroma is recommended only in symptomatic patients. This procedure gives good symptomatic relief and can prevent life-threatening complications.

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