Journal of Cardiothoracic Surgery

CASE REPORT

Open Access

Paediatric vascular-related hereditary giant rib osteochondroma: report of a successful chest wall reconstruction

Klein Dantis[1*], Ramandeep Singh[2] and Paramdeep Singh[2]

Abstract

Background Hereditary multiple osteochondromas (HMO) are benign neoplasms that predominantly affect the bones around the knee joint, proximal humerus, wrist, pelvis, and to a lesser extent, the rib. Although rib-origin osteochondromas are uncommon and often asymptomatic, they can lead to pleural, pericardial, and diaphragmatic injuries secondary to tumour related pressure. We are presenting a unique case of a vascular-related giant osteochondroma rib that is hereditary, originating from the right second, third, and fourth ribs causing compression symptoms in a young child. The child underwent wide local excision and reconstruction using polypropylene mesh that was managed successfully with no recurrence.

Case presentation A 9-year-old male experiencing fullness and pain in the right chest for two months with a parallel medical history of swellings in first- and second-generation relatives that is hereditary in origin underwent contrast-enhanced computed tomography revealing a sessile rib osteochondroma arising from the anterior aspects of the right second to fourth ribs with an associated large cartilaginous cap. The bony growth measured 4.5 × 2.5 cm indenting the right upper lobe while, its cartilaginous cap measured 2.5 × 4.8 cm posterior-superiorly seen encasing the axillary artery and 3D volumetric reconstructive image revealed the proximity of the axillary and subclavian artery with the lesion. A biopsy was inconclusive with no malignant cells, so he underwent wide local excision of the tumour and reconstruction with polypropylene mesh. Follow-ups at one, three, six, and nine months were uneventful with normal thoracic curvature and no sign of recurrence.

Conclusion This case illustrates the surgical challenges addressed and the successful outcome of a paediatric chest wall reconstruction in a growing child, utilizing advanced imaging techniques, to underscore the importance of an individualized, innovative approach in managing rare skeletal anomalies.

Keywords Rib, Osteochondroma, Chest wall reconstruction, Mesh, Paediatric, Hereditary multiple osteochondroma, Vascular

*Correspondence:
Klein Dantis
drkleindantis86@gmail.com
[1]Department of CTVS, All India Institute of Medical Sciences(AIIMS), Bathinda 151001, India
[2]Department of Radiodiagnosis, All India Institute of Medical Sciences, Bathinda, India

Background

Hereditary multiple osteochondromas (HMO) are benign neoplasms that predominantly affect the bones around the knee joint, proximal humerus, wrist, pelvis, and to a lesser extent, the rib [1]. Although rib-origin osteochondroma are uncommon and often asymptomatic, they can occasionally cause pressure-related issues leading to injuries in pleural, pericardial, and diaphragmatic areas [2]. While many of these lesions are discovered during other medical evaluations, the vascular compromise commonly observed in the lower extremities is unusual for osteochondromas affecting the first rib potentially causing vascular or neurogenic thoracic outlet syndrome [3, 4]. With no previous reports of axillary vessel involvement, we are presenting a unique case of a giant osteochondroma that is hereditary arising from the right second, third, and fourth ribs, leading to vascular compromise, and also shall discuss its successful surgical management.

Case presentation

A 9-year-old male with height-133 cm, weight-27 kg, body mass index (BMI)- of 15.26 kg/m^2 and, chest circumference of 30 cm, presented with fullness and pain in the right chest for the past two months. Upon examination, diffuse swelling was noted over the right chest with dilated veins (Fig. 1), along with tenderness and painful right upper limb abduction beyond 90 degrees. Further examination revealed multiple localized swellings measuring 2 × 2 cm over the left seventh rib (Fig. 2), and diffuse swelling over the right thigh and left knee with no lesions over the spine. He was unable to perform a pulmonary function test (PFT). A parallel medical history in the first- and second-generation relatives confirmed a hereditary origin. His blood, renal, and liver parameters were normal except for an elevated c-reactive protein of 125.4 mg/l. The contrast-enhanced computed tomography of the thorax revealed sessile exostosis arising from the anterior aspects of the right second to fourth ribs with an associated large cartilaginous cap suggestive of sessile rib osteochondroma with its bony component measuring 4.5 × 2.5 cm indenting the right upper lobe and its cartilaginous cap measuring 2.5 × 4.8 cm (Fig. 3) posterior-superiorly seen encasing the axillary artery as well as a 3D volumetric reconstructive image revealing the proximity of the axillary and subclavian artery with the lesion (Fig. 4). The fine needle aspiration biopsy was inconclusive for osteochondroma as well as biopsied specimen did not detect any malignant

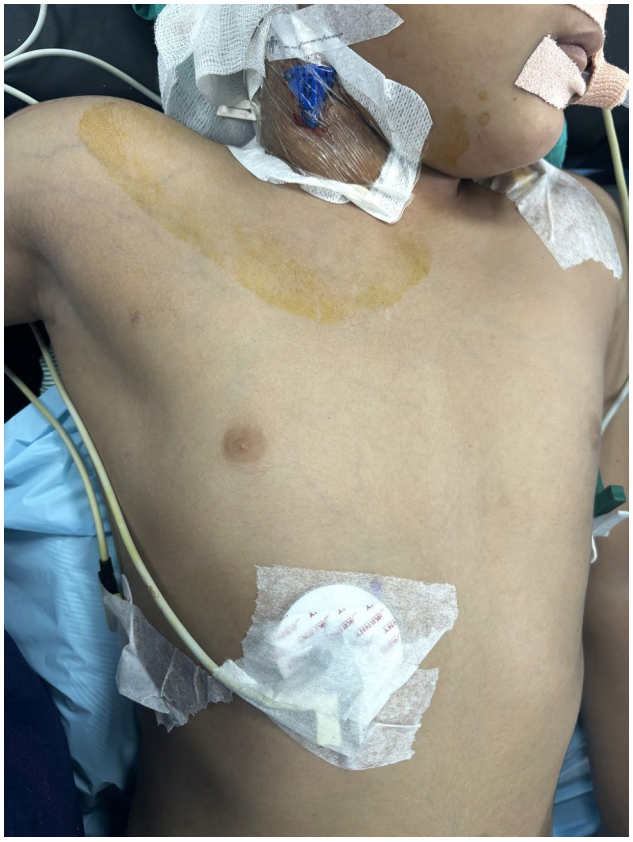

Fig. 1 Fullness in the right chest with dilated veins secondary to giant rib osteochondroma

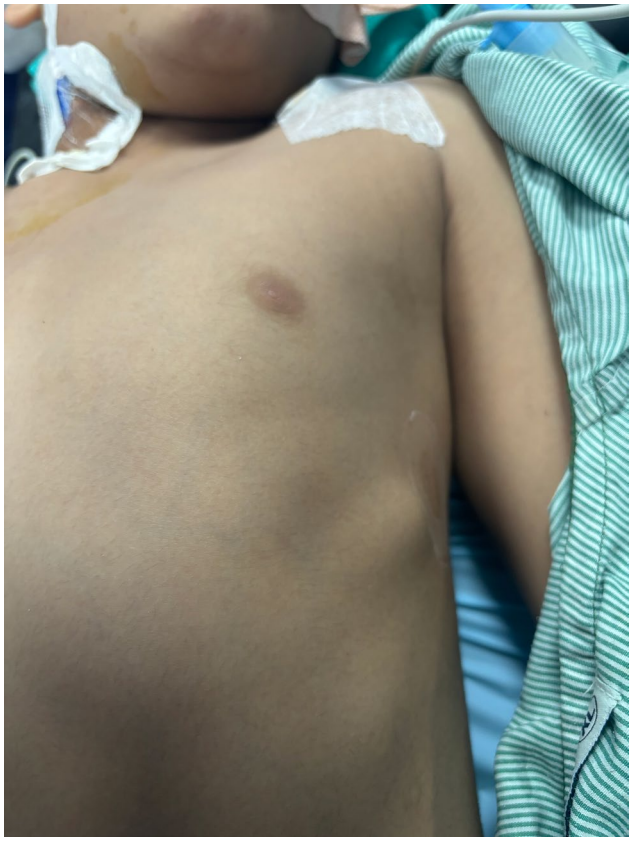

Fig. 2 Prominent bony swelling over the left seventh rib -indicating multiple in nature

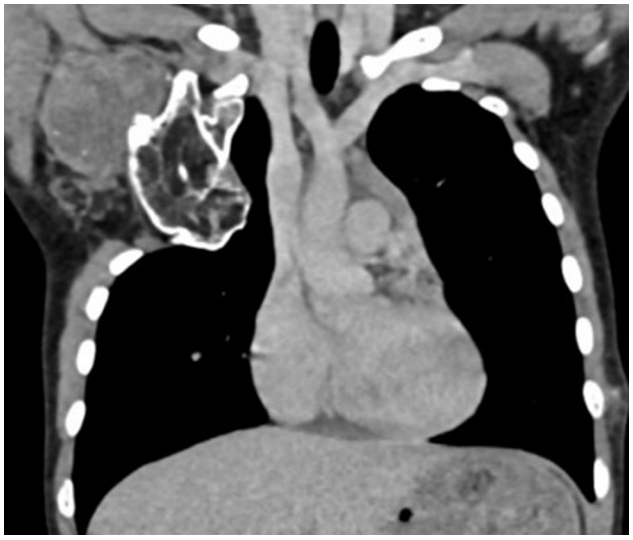

Fig. 3 Coronal view of contrast-enhanced computed tomography of the thorax in the bony window showing sessile exostosis arising from the anterior aspects of right second to fourth ribs with an associated large cartilaginous cap encasing axillary artery

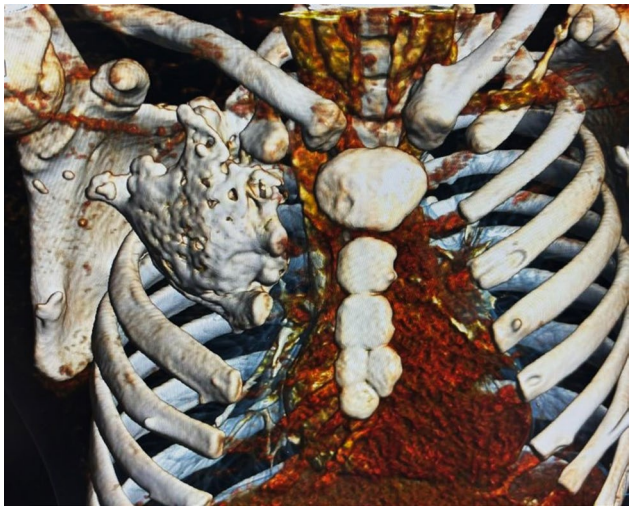

Fig. 4 3D rendered image showing sessile lesion originating from the right anterior ribs (2nd to 4th) with its close proximity to the axillary and subclavian artery

cells. As the right chest swelling was huge, painful, and interfering with daily routine activities, surgical intervention was mandatory. He underwent wide local excision of the tumour with a 2 cm margin, and reconstruction of the 6 × 4 cm defect using polypropylene mesh under general anaesthesia.

Surgical technique

A transverse incision was placed at the level of the angle of Louis from the parasternal line extending laterally for 5–7 cm (Fig. 5). Superior and inferior flaps of skin and subcutaneous tissue flaps were created followed

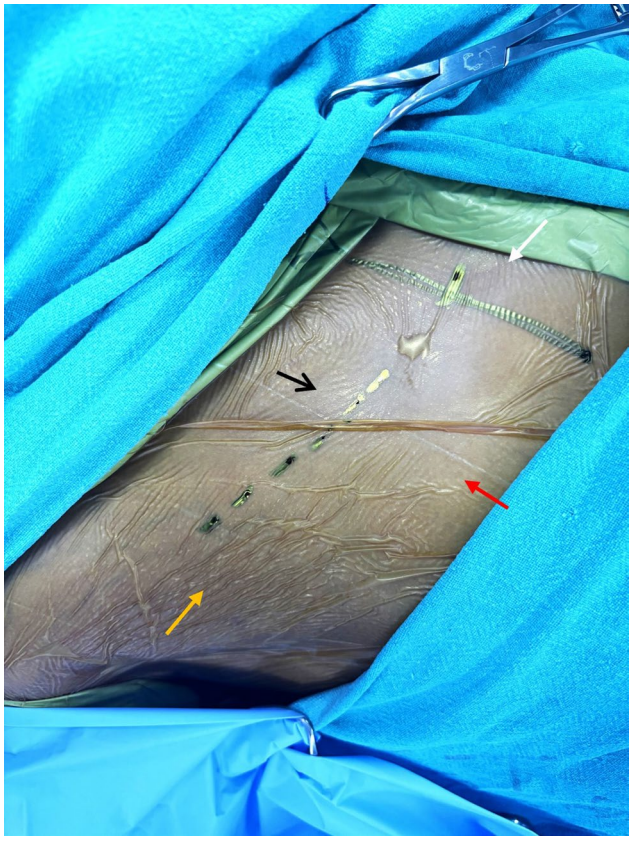

Fig. 5 Transverse incision at the level of second intercostal space extending from the parasternal line (arrows- white, black, yellow and red indicates: medial, superior, lateral and inferior)

by a pectoralis major muscle flap in a similar fashion. Bony hard swelling was identified protruding anteriorly. Medially ribs four, three, and two were scored, isolated, and cut with a bone cutter at the level of costochondral junction followed by superior and inferior aspects of the lesion along the intercostal muscle with the sufficient margin and posterosuperior aspect of the lesion being left intact to the underlying tissue. The anterior cartilaginous portion was opened and the artery traversing on the posterosuperior aspect was dissected, isolated, and safeguarded. The posterior aspect of the lesion along with the cartilaginous cap was freed from the underlying tissue and the lateral aspect of the ribs two, three, and four were scored, isolated, and cut. The entire specimen removed was sent for histopathological diagnosis (Fig. 6). The thoracic cavity was sterilized with 20% betadine and normal saline (0.9%) in a ratio of 1:4 followed by warm saline washes (Fig. 7) and a single drain placement followed by a chest wall reconstruction. The polypropylene mesh (Ethicon, Johnson and Johnson, USA, PML01,) was designed as required and placed over the defect and sutured to the intercostal muscles with prolene 3 – 0 interrupted (Fig. 8) followed by a musculocutaneous flap.

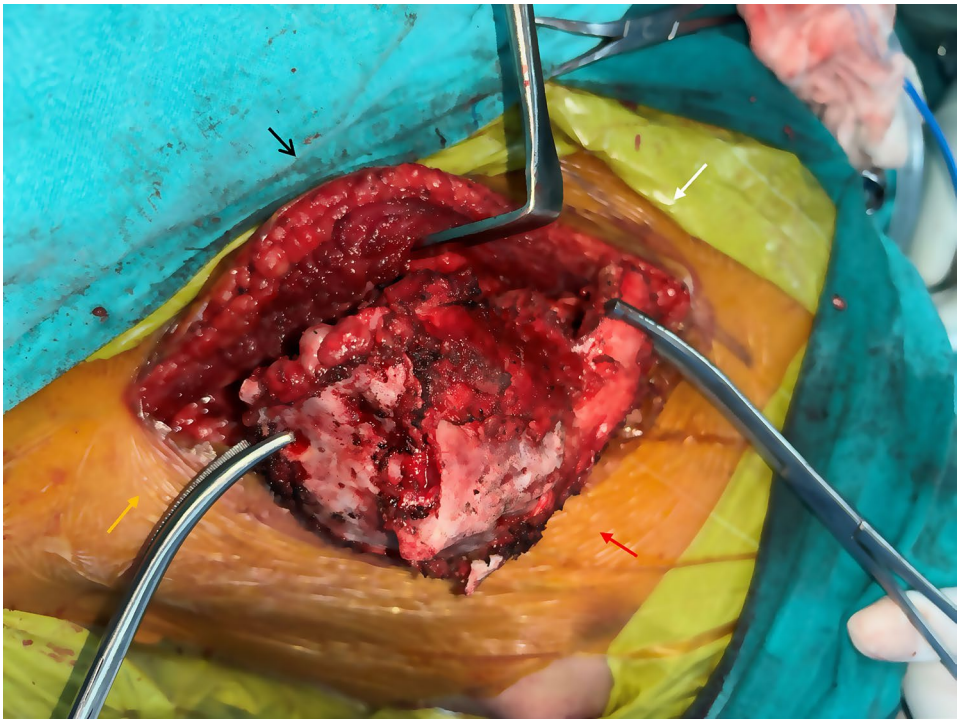

Fig. 6 Intraoperative image showing osteochondroma specimen involving second, third and fourth rib (arrows- white, black, yellow and red indicates: medial, superior, lateral and inferior)

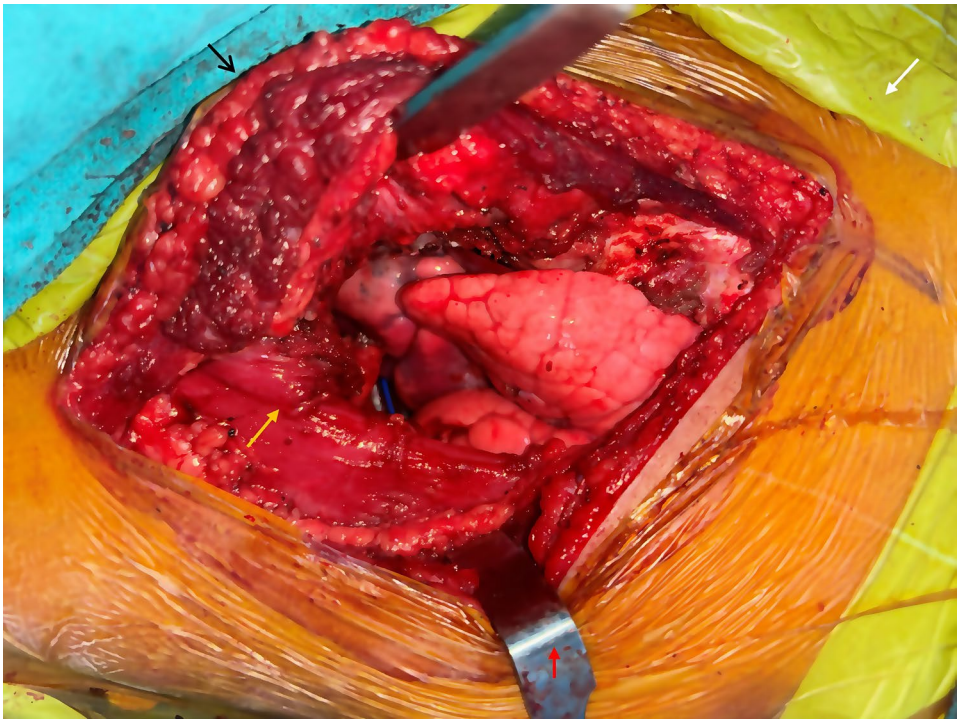

Fig. 7 Intraoperative image showing expanding lung through the intercostal space following excision and removal of the specimen (arrows- white, black, yellow and red indicates: medial, superior, lateral and inferior)

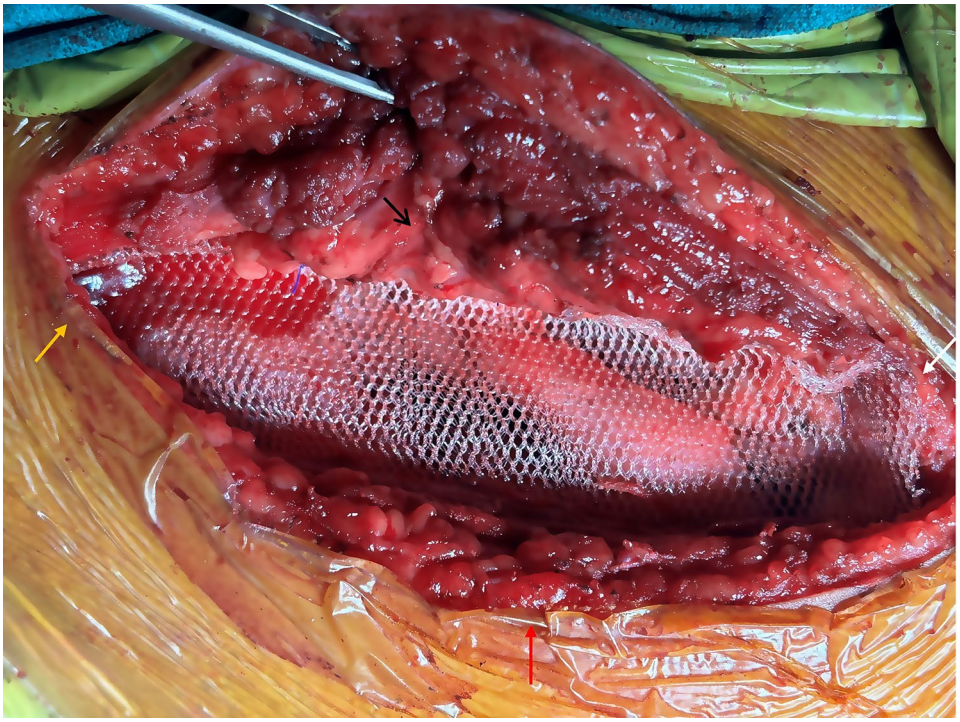

Fig. 8 Intraoperative image showing chest wall reconstruction with a polypropylene mesh (arrows- white, black, yellow and red indicates: medial, superior, lateral and inferior)

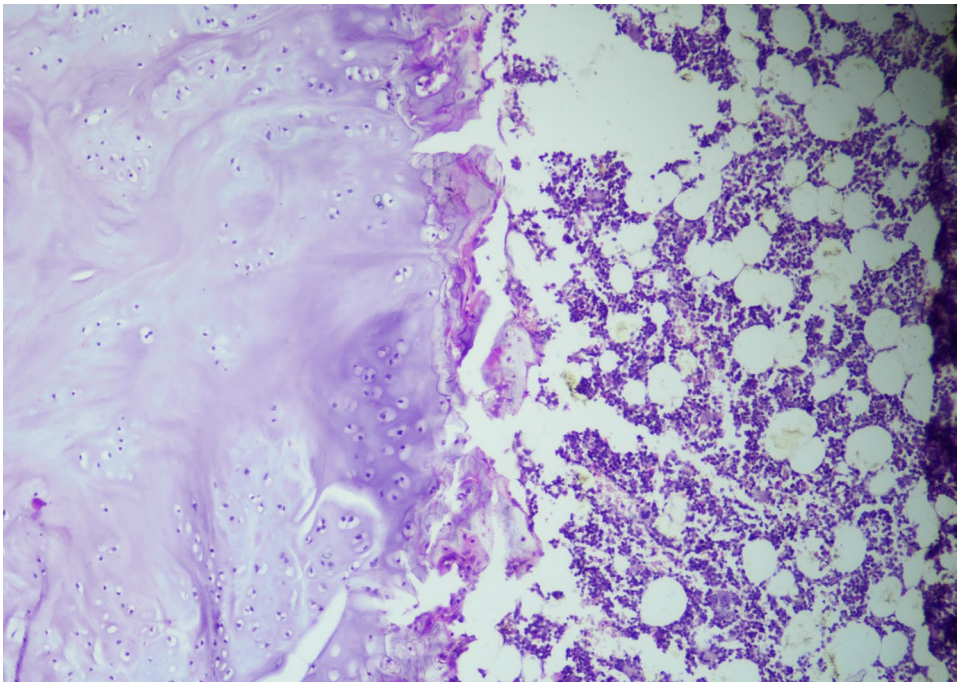

Fig. 9 Histopathology of osteochondroma (10X, haematoxylin and eosin stain) showing hyaline cartilaginous cap with an underlying stalk of mature bone. At the junction, a few areas show enchondral ossification

Although the patient experienced a postoperative fever on day 3, prompt action, including upgrading the antibiotics for 7 days, led to significant improvement with subsiding c-reactive protein. The drain was removed on day 5, and the patient was discharged to go home on day 13 after constant observation for subsiding fever. His histopathology was suggestive of osteochondroma with no signs of malignancy (Fig. 9). Follow-ups at one, three, six,

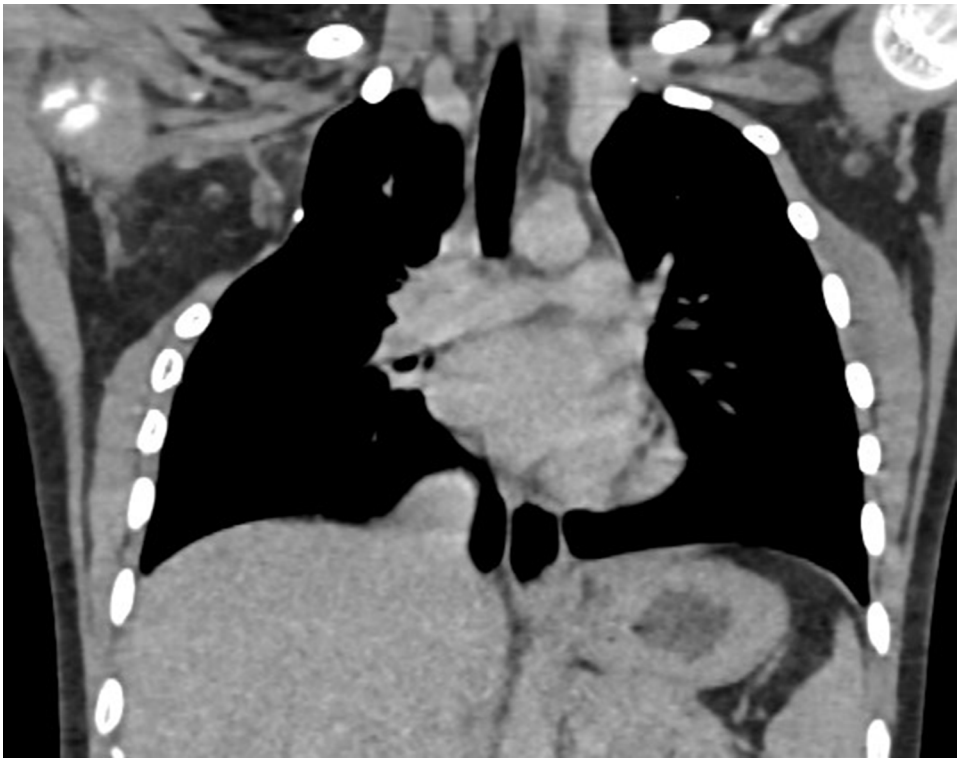

Fig. 10 Postoperative CECT thorax coronal view showing no recurrent or residual lesion at ninth month follow-up

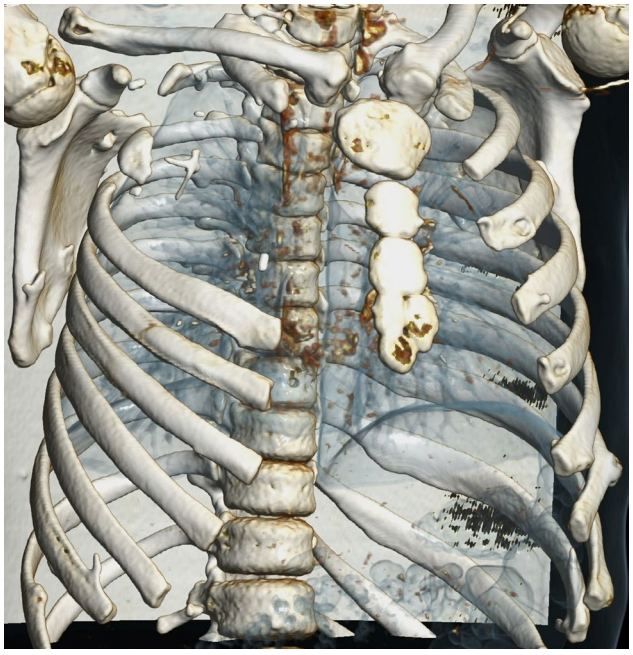

Fig. 11 Postoperative 3D volumetric reconstructive CT thorax at nine months showing no residual lesion or recurrence

and nine months with imaging for recurrence and chest wall function were uneventful with normal thoracic curvature (Figs. 10, 11 and 12) except for a hypertrophic scar developed in the seventh month, aggravated on the tenth

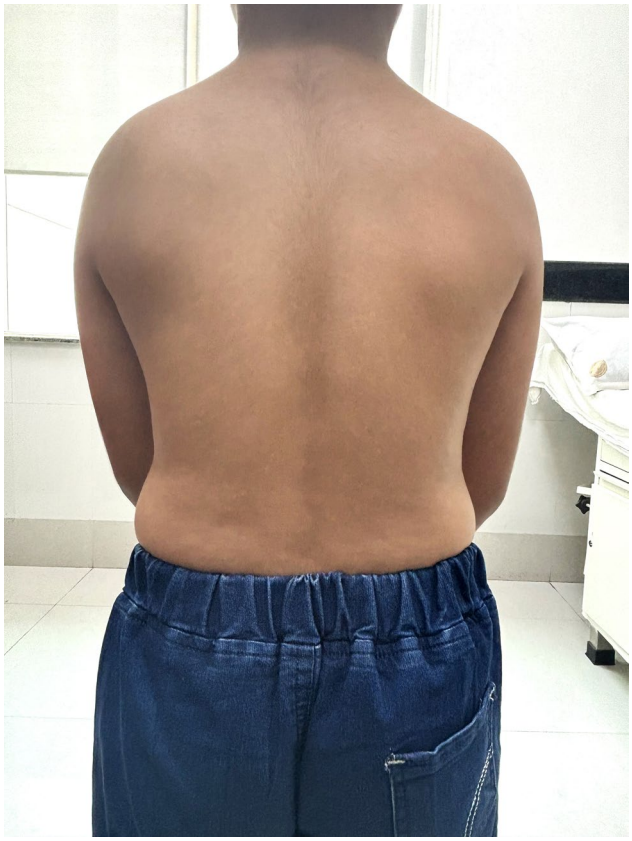

Fig. 12 Normal thoracic curvature at ninth month follow-up

month of unknown etiology for which he has been suggested to receive intralesional triamcinolone/silicone gel sheet treatment (Fig. 13). His follow-up PFT was optimal with chest circumference reduced to 27 cm from 30 cm, weight increased to 35 kg from 27 kg as well as BMI improved from 15.26 kg/m^2 to 19.79 kg/m^2 appropriate for growth and development. He can perform daily routine activities with no pain and normal shoulder movements in all directions.

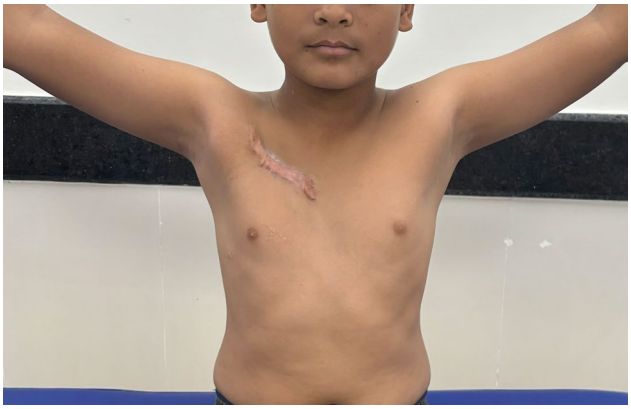

Fig. 13 Post reconstruction anterior chest wall with arms in abduction and hypertrophic scar of unknown etiology developed at seventh month and aggravated at ten months follow-up, advised for intralesional triamcinolone or silicone gel sheet treatment

Discussion

HMO is a rare autosomal dominant disease with variable penetrance of EXT1 and EXT2 genes on chromosome 8 and chromosome 11 predominantly affecting males (1.5:1) with a prevalence of 1:50000 in western countries [5]. The disease usually affects the long bones arising from endochondral ossification in the first decade of life with no extension beyond puberty [2, 5]. They induce symptoms based on localization, neurovascular compression, and associated fractures as seen in our case. Their transformation to chondrosarcoma is rare, seen in 0.5-5% of the patients only [2].

Efficient diagnostic procedures and interventions play a vital role in accurately assessing the disease's extent and overcoming technical challenges for future surgical planning. In our case, the use of 3D computed tomography proved invaluable in determining the disease's extent, identifying the involved structures and growth patterns, as well as understanding the tumour's relationship with major blood vessels thus guiding intraoperatively for better and safe surgical resection. Additionally, a video-assisted thoracoscopic approach that aids in identifying the intrathoracic lesion, however, seemed ineffective as the axillary artery was traversing the posterosuperior aspect of the cartilaginous cap and posteriorly the lesion was adherent to the underlying tissue [4].

Recently with the improvement in reconstruction technique, critical care, and pharmacological therapy, surgical resections performed are aggressive and extensive [6]. The physiological chest wall growth represents a unique challenge for chest wall reconstruction in the paediatric population. One such major concern is paradoxical respiration hence; the reconstruction must be sturdy and compatible with the chest wall's future growth to maintain intrathoracic volume [7]. Even though rigid and semi-rigid prostheses have been used in adult patients, their use in paediatric patients has led to chest wall deformation and scoliosis [6, 9]. Various materials such as methyl methacrylate, non-crosslinked biological membranes, and titanium plates have been used for large defects [8]. However, these methods have drawbacks including technical fabrication difficulties, long operative hours, postoperative fractures, and permanent rigidity leading to chronic pain and deformity, finally resulting in an unstable chest wall [7, 8]. Furthermore, the expandable prosthetic titanium rib, while allowing for serial growth, appeared bulky in relation to the soft tissues [8]. The use of Permacol membranes for chest wall reconstruction again seemed beneficial but required a bony strut underneath and a latissimus dorsi flap superiorly to replace the function of the pectoralis major muscle function [9]. A study by Glotzbecker et al. also reported that "upper chest wall resections" (above the sixth rib) were prone to scoliosis when compared to "lower chest wall resections" in paediatric patients due to the excessive force applied to the column by the rigid and semi-rigid prostheses affecting the opposite chest wall over non-rigid prostheses [10].

Although reports have discussed the benefits and drawbacks of meshes, polypropylene mesh plays a crucial role in maintaining chest wall integrity in the paediatric population, especially for larger defects over the long run [6, 10, 11]. Their low rate of local infection, material explanations, and chest wall stability have been proven in the larger study group [6, 11]. Furthermore, a simultaneous musculocutaneous flap cover over the mesh in our case prevented paradoxical respiration and led to an excellent outcome. Though composite meshes are superior in anti-adhesive properties for viscera over polypropylene meshes, no chest complaints or abnormalities on radiological imaging were reported in the present case [12].

Conclusion

This case illustrates the surgical challenges addressed and the successful outcome of a paediatric chest wall reconstruction in a growing child, utilizing advanced imaging techniques to underscore the importance of an individualized, innovative approach in managing rare skeletal anomalies.

Acknowledgements
I would like to thank Melisha Rolita Pinto (melisha.pinto@gmail.com) for editing and proofreading the manuscript.

Author contributions
Original draft preparation, conceptualization: K. DInvestigation: R.SData curation: P.SReviewed and approved by all the authors.

Funding
None.

Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
Not applicable for case reports.

Consent for publication
Consent was obtained from patient's parents to publish the data.

Competing interests
The authors declare no competing interests.

Received: 22 July 2024 / Accepted: 1 December 2024
Published online: 11 February 2025

References

1. Chakravorty A, Fong L, Grant P, Wolfenden H, Akhunji Z. Multiple rib osteochondromas: a rare cause of spontaneous haemothorax. ANZ J Surg. 2021;91(10):E666–7. https://doi.org/10.1111/ans.16705.
2. Dumazet A, Launois C, Dury S, et al. Hereditary multiple exostoses of the ribs as an uncommon cause of pneumothorax: a case report. Medicine. 2018;97(35):e11894. https://doi.org/10.1097/MD.0000000000011894.
3. O'Brien PJ, Ramasunder S, Cox MW. Venous thoracic outlet syndrome secondary to first rib osteochondroma in a paediatric patient. J Vasc Surg. 2011;53(3):811–3. https://doi.org/10.1016/j.jvs.2010.09.067.
4. Hamouri S, AlQudah M, Al-Zoubi N, Al Gargaz W, Jarboa' H, Hecker E. Rib osteoblastoma as a cause of neurogenic thoracic outlet syndrome: a case report. Am J Case Rep. 2021;22:e928548. https://doi.org/10.12659/AJCR.928548.
5. D'Arienzo A, Andreani L, Sacchetti F, Colangeli S, Capanna R. Hereditary multiple exostoses: current insights. Orthop Res Rev. 2019;1311:199–211. https://doi.org/10.2147/ORR.S183979.
6. Scanagatta P, Giogetta CE, Ravalli E et al. chest wall reconstruction in children and adolescents: recent personal experience from a case series and narrative review. AME Surg J.2022: 2;34. https://doi.org/10.21037/asj-22-22
7. Pan R, Lu X, Wang Z, Duan L, Cao D. Hemothorax caused by costal exostosis injuring diaphragm: a case report and literature review. J Cardiothorac Surg. 2022;17(1):230. https://doi.org/10.1186/s13019-022-01984-7.
8. Anderson CJ, Spruiell MD, Wylie EF, et al. A technique for paediatric chest wall reconstruction using custom-designed titanium implants: description of technique and report of two cases. J Child Orthop. 2016;10(1):49–55. https://doi.org/10.1007/s11832-015-0709-1.
9. Kane G, Orr D, Pears J, McGuinness J. A novel approach to extensive chest wall reconstruction in a child. Ann Thorac Surg. 2021;111(6):e389–91. https://doi.org/10.1016/j.athoracsur.2020.09.079.
10. Glotzbecker MP, Gold M, Puder M, Hresko MT. Scoliosis after chest wall resection. J Child Orthop. 2013;7(4):301–7. https://doi.org/10.1007/s11832-013-0519-2.
11. Stoelben E, Koryllos A. Brustwandrekonstruktion mittels polypropylennetz: eine monozentrische 8-jährige analyse [Chest wall reconstruction using polypropylene mesh: a single-center 8-year analysis]. Zentralbl Chir. 2021;146(3):329–34. https://doi.org/10.1055/a-1180-9621.
12. Dantis K, Singh R, Goel A, Garg B. An innovative reconstruction of an enbloc resected composite giant chest and abdominal wall chondrosarcoma with 3D-composite mesh. J Cardiothorac Surg. 2024;19(1):126. https://doi.org/10.1186/s13019-024-02595-0.

Publisher's note