Case Report

ANNALS OF MEDICINE & SURGERY

OPEN

Ewing's sarcoma arising from the right sided chest wall: a case report

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Introduction and importance: Ewing sarcoma (ES) is a malignant tumour prevalent in young adults with a reported 5-year survival ranging between 40 and 60% in most studies. Majority of the patients with ES are usually diagnosed late with significant chest wall mass, chest pain or respiratory distress.

Case presentation: Here, the authors present a case of a 21-year-old female with a diagnosis of right sided chest wall ES treated with neoadjuvant chemotherapy followed by surgical resection of the mass.

Clinical findings and investigations: The patient presented to the Surgical OPD with shortness of breath for 6 months associated with chest pain on the right side. Radiological investigations including chest X-ray and multi-detector row computed tomography chest was done. Additionally, diagnosis of ES was confirmed with histopathological examination of the mass obtained from fine needle aspiration cytology.

Interventions and outcome: She was planned for safe maximal resection of tumour with chest wall reconstruction using double prolene mesh with bone cement and the defect was sutured with adjacent ribs. Good outcome was noted on postoperative period with resolution of symptoms.

Relevance and impact: This procedure is now commonly used and is considered as an effective treatment for chest wall tumours, which was also noted in our case and the procedure is also well tolerated.

Keywords: case report, chest wall reconstruction, chemotherapy, ewing's sarcoma

Introduction

Ewing sarcoma (ES) is a malignant tumour prevalent in young adults accounting for 10–15% of all bone sarcomas[1]. ES of the chest wall have a reported 5-year survival ranging between 40 and 60% in most studies[2]. Although more common in bone, about a quarter of ESs originate in soft tissues, and about one-fourth of cases have detectable metastases at the time of diagnosis. ES most commonly involves the diaphysis or meta-diaphyseal region of long bones. However, the pelvis, ribs and spine are also often involved[3,4].

HIGHLIGHTS

- Malignant chest wall tumour due to Ewing sarcoma in young age is a rare presentation.
- Treatment includes surgical resection of the mass in conjugation with chemotherapy along with chest wall reconstruction.
- Despite adequate management , the recurrence rate of these tumours are high, thus, clinicians must follow-up the patient frequently.

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article

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Annals of Medicine & Surgery (2023) 85:3709–3713

Received 4 December 2022; Accepted 10 June 2023

Published online 19 June 2023

http://dx.doi.org/10.1097/MS9.0000000000000994

Majority of the patients with ES present with symptoms such as a mass or pain[5]. Due to this, children are usually diagnosed late with significant chest wall mass, chest pain or respiratory distress[6]. ES are distinguished by the presence of non-random chromosomal translocations producing fusion genes that encode aberrant transcription factors. The t(11;22)(q24;q12) translocation is associated with 85% of tumours and leads to EWS-FLI-1 formation, whereas t(21;12)(22;12) and other less common translocations induced EWS-ERG fusion represents the remaining 10–15% of cases[3].

Plain radiographs exhibit permeative and infiltrative destruction of the affected bone (often in the diaphysis of the long bone). Additionally, an onion skin-like appearance and spicules are suggestive of periosteal reactions. Computed tomography is helpful for delineating extraskeletal soft tissue masses, destruction of the bone cortex, and lung metastasis. On MRI, ES of the bone shows low signal intensity on T1-weighted images and high

signal intensity on T2-weighted images, and emerges as large extraskeletal soft tissue masses coming from bone[7].

Radical "en-bloc" resection remains crucial facing various challenges like infiltrations into lung, spine, pericardium or diaphragm. Simultaneous chest wall reconstruction also remains challenging when covering large defects with prosthetic materials and viable tissue to prevent instability, flailing and infections[2].

Case details

Twenty-one-year-old lady presented to Cardiothoracic and Vascular unit of Surgery Department of Dhulikhel Hospital, Kathmandu University Hospital with gradually progressive shortness of breath for 6 months associated with right sided chest pain (axillary region). Chest X-ray was done which revealed large well defined opacity involving right mid and lower zone with possible mass effect to mediastinum and hilum as shown in Fig. 1. However, there was no history of fever, cough, hemoptysis, loss of appetite and trauma. On auscultation, there was decreased airway entry on the right side.

Following this, the patient was advised for Multi-detector row computed tomography chest (Fig. 2) which showed mass on the right side. For confirmatory diagnosis, fine needle aspiration cytology from the right thoracic region was done. The findings were positive for small round cell tumours consistent with Ewing's Sarcoma (CD99 and NKX2.2 markers). Patient was then advised for neoadjuvant chemotherapy (combination of Vincristine, cyclophosphamide, doxorubicin) for four cycles. After that, her tumour size decreased from 10×5 cm (Stage IIB) to 5×5 cm as shown in chest X-ray in Fig. 3. Further management of the case was planned for right posterolateral thoracotomy with safe maximal resection of tumour with chest wall reconstruction.

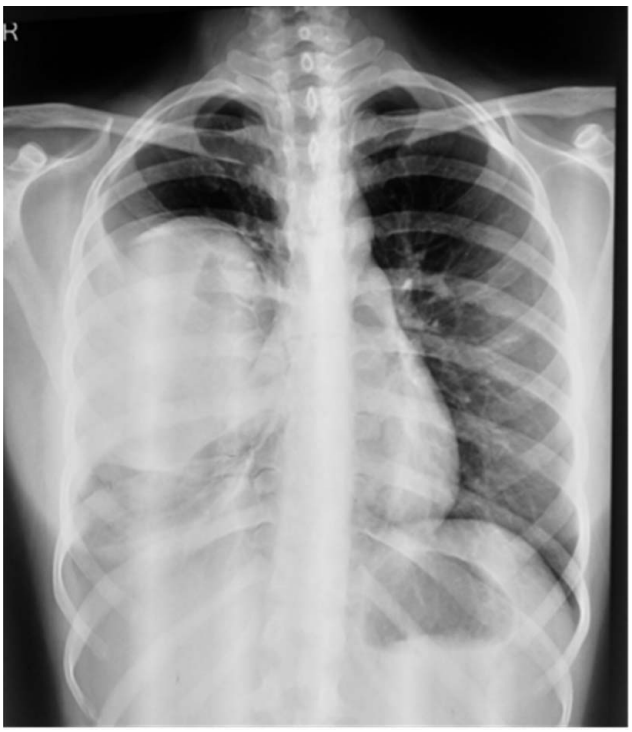

Figure 1. Large well defined opacity involving right mid and lower zone with possible mass effect to mediastinum and hilum.

Intraoperative findings were suggestive of presence of an intrathoracic tumour sized 4×5 cm with origin from right latissimus dorsi and third and fourth rib extending up to the upper pole of lower lobe of the right lung. Feeders from the subclavian vessels supplying anterior and superior part of the tumour were found. There was erosion present from the second to sixth ribs along with adhesion and attachment between upper pole of lower pole of the right lung, tumour and fourth rib. The resected tissue along with third, fourth, fifth and sixth ribs was sent for histopathological examination as shown in Fig. 4. Following resection, the defect was traced on a sterile paper. Using that measurement, two prolene mesh were cut in the same size. Bone cement was prepared and shaped to the size of the defect which was adhered with the mesh on both the sides (Fig. 5). Proper contour of the prosthesis was also given. This was placed in the defect and secured by suturing with adjacent ribs using sternal wire (Fig. 6).

Histopathological findings were suggestive of ES, as presented in Fig. 7. Two weeks after the surgery, the patient was followed up and sutures were removed. She did not have any significant complaints and was advised for follow-up chest X-ray as represented by Fig. 8. A total of 14 cycles of chemotherapy has been planned, and the patient has recently completed 7 cycles; a total of 30 days of radiotherapy has been completed as well. Currently, she has no new symptoms and is recovering well.

The case report has been written in accordance with the SCARE Guideline[8].

Discussion

We present a case of ES of right sided chest wall managed with neoadjuvant chemotherapy followed by safe maximal resection of tumour alongside chest wall reconstruction using double prolene mesh and bone cement sandwich. This treatment is considered beneficial for major chest wall defects[9].

In a retrospective study of chest wall tumours at a cancer hospital in Nepal done by Thakur and colleagues. in 31 patients; presenting complaints was mass in 96.8% and pain in 48.4%.The mass was 5cm or less in 34.4%, from 5 to 10 cm in 32.3% and more than 10 cm in 32.3%.Operative management was done with wide local excision and chest wall reconstruction using both muscular flaps and prolene mesh. There was no postoperative mortality and the rate of malignancy was 48.4%[10].

The primary treatment protocols as stated in the literature are neoadjuvant chemotherapy, radical "en-bloc" resection and chest wall reconstruction along with adjuvant chemotherapy-/radiotherapy[11,12]. By shrinking tumour mass, chemotherapy decreases vascularity and vulnerability and improves achievability of complete removal with negative microscopic margins[13]. Following neoadjuvant chemotherapy, complete "en-bloc" resection of the primary lesion and simultaneous reconstruction of the defect is compulsory. Radical surgical resection is associated with better survival rates; patients with complete resection have a higher 5-year survival rate compared to patients who had an incomplete resection[13]. Similarly, in our study, patients underwent four cycles of neoadjuvant chemotherapy followed by safe maximal resection of tumour along with chest wall reconstruction using double prolene mesh with bone cement sandwich.

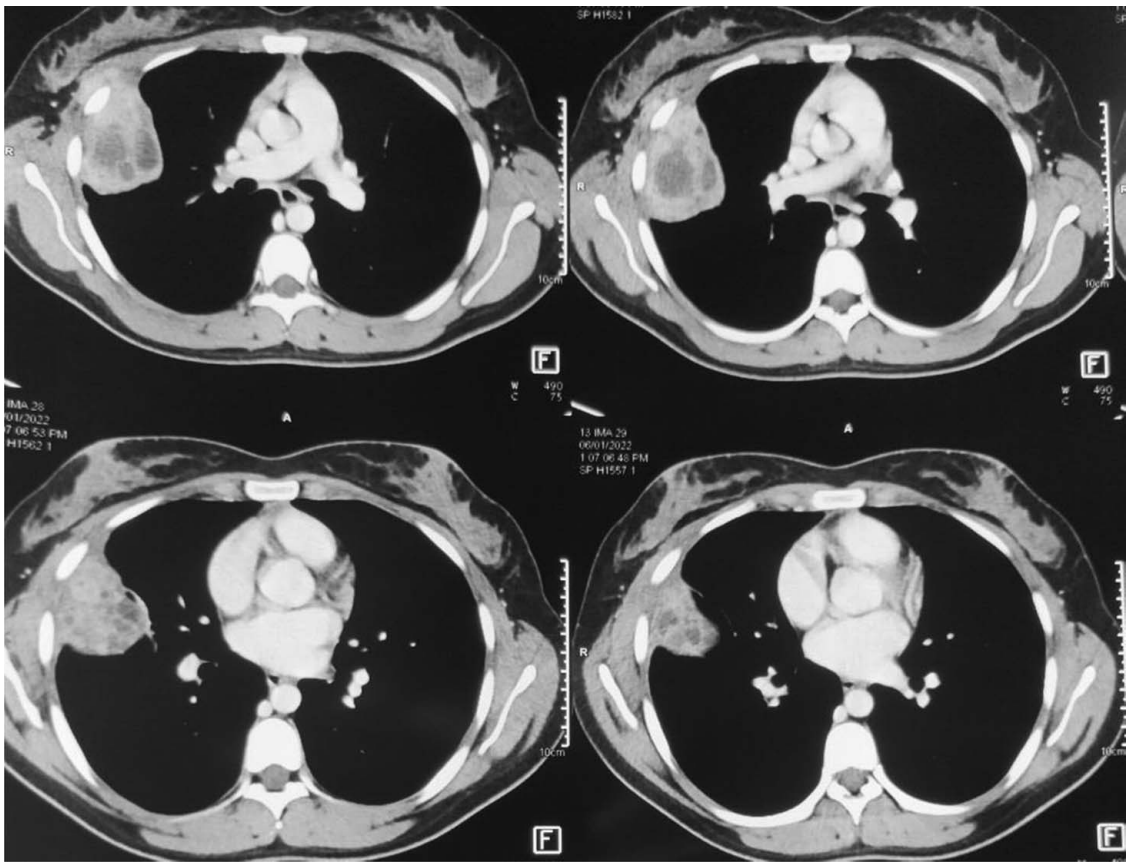

Figure 2. CT scan of the chest showing the location of the chest wall tumour. CT, computed tomography.

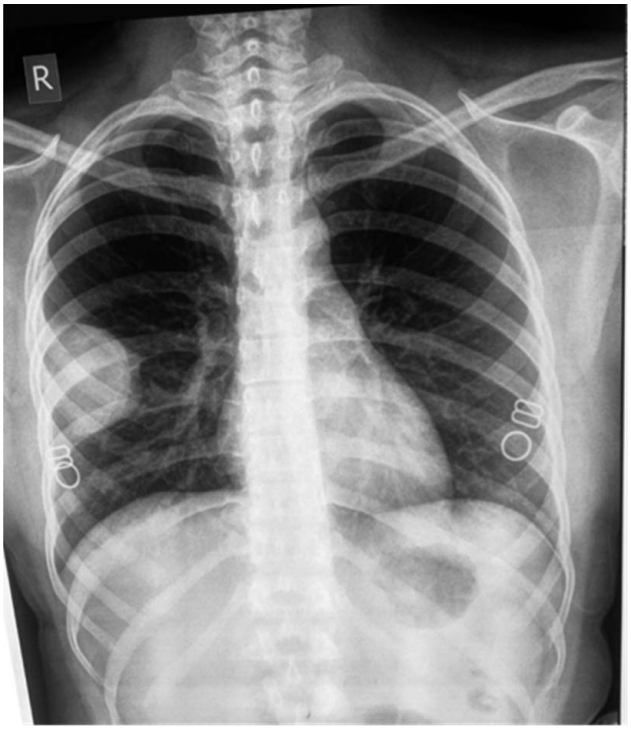

Figure 3. Chest X-ray after four cycles of neoadjuvant chemotherapy.

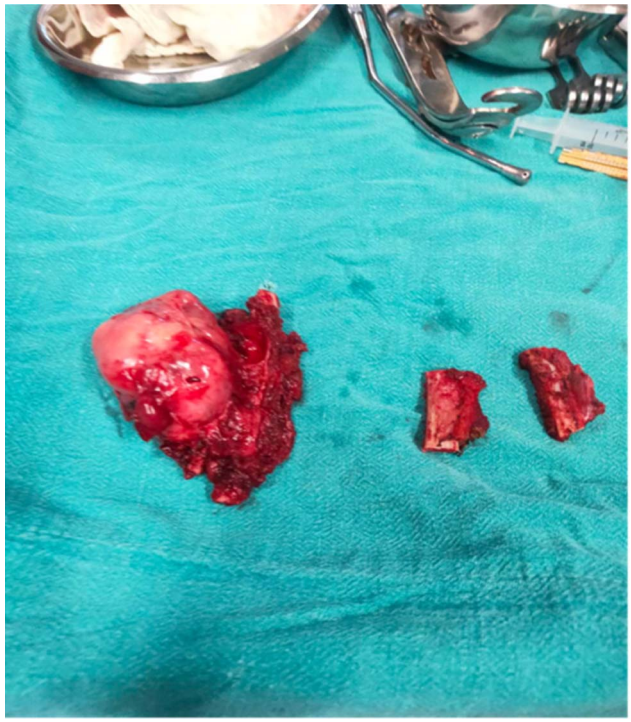

Figure 4. Resected tumour along with ribs.

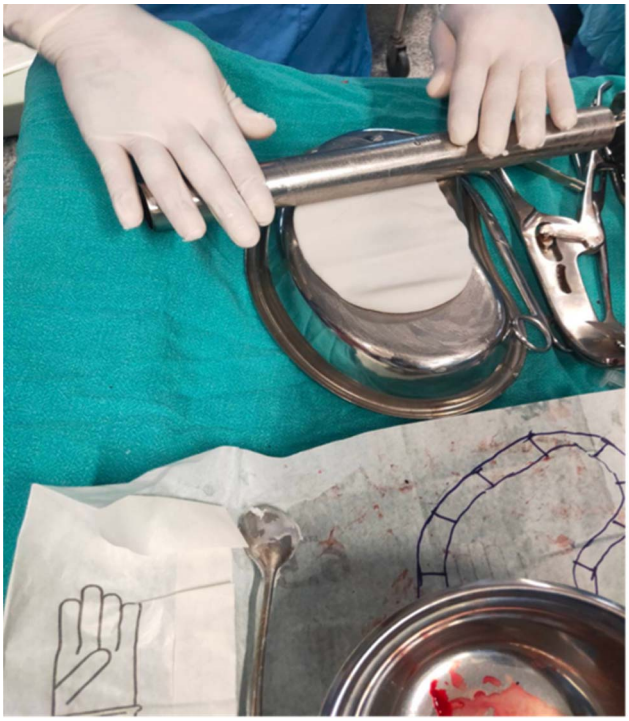

Figure 5. Preparation of double prolene mesh with bone cement.

Despite the effective treatment intervention, ES has an unfavourable prognosis. Triarico *et al.*[14] have reported a series of nine patients with "Askin tumours," and their 5-year survival was 60% at a median follow-up of 53.1 months. The known factors associated for prognosis are tumour diameters greater than 5 cm, Lactate dehydrogenase levels greater than 240U/l and late stage diagnosis[13]. In another study, patients without good

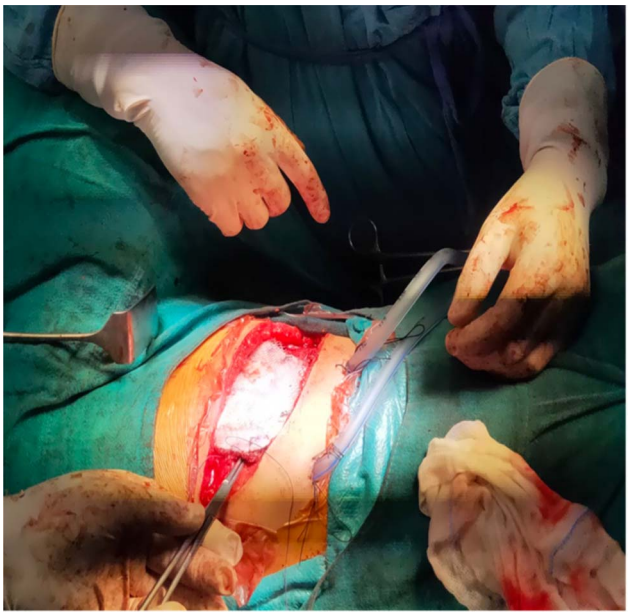

Figure 6. Chest wall reconstruction.

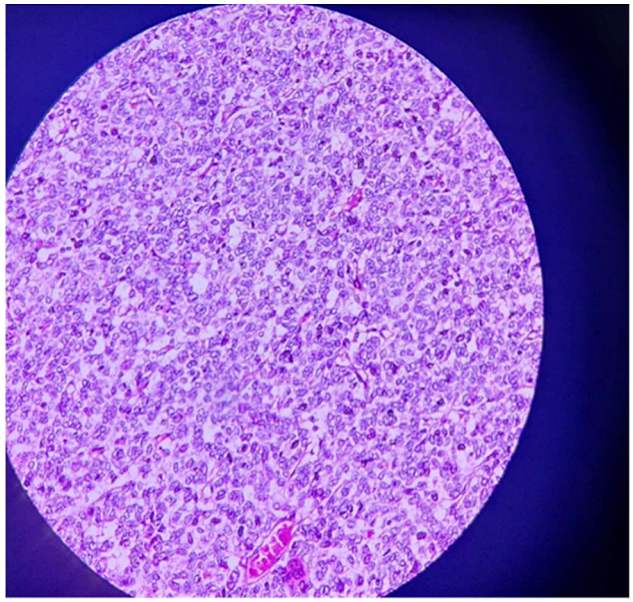

Figure 7. Section from specimen sent for histopathological evaluation after resection of tumour.

histological response to chemotherapy and positive surgical margins had significantly worse event-free survival[15]. Laskar *et al.*[16] have shown that age at diagnosis more than 18 years, poor response to induction chemotherapy, and association of

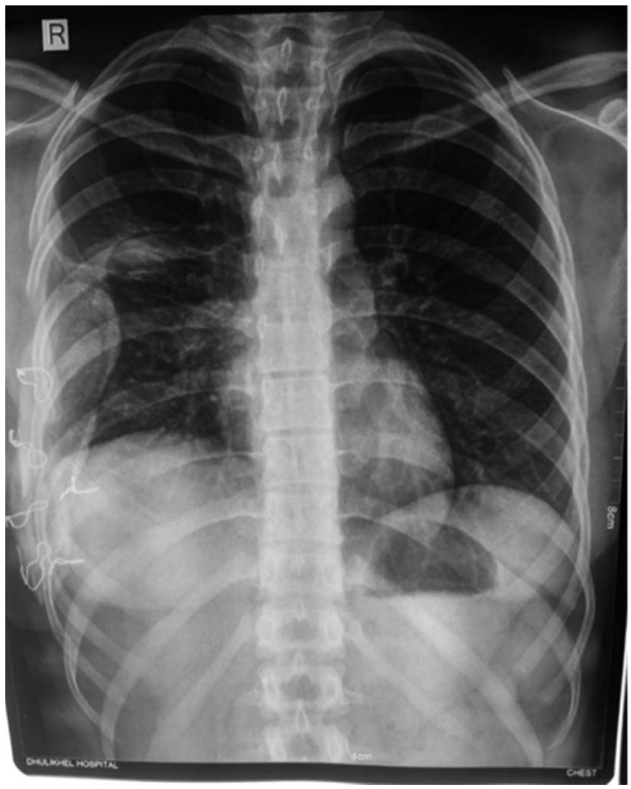

Figure 8. Chest X-ray during follow after 2 weeks.

pleural effusion are indicators of poor survival. In our case, the patient was diagnosed at an age above 18 years, had large pre neoadjuvant chemotherapy tumour size but had good response to induction chemotherapy and no pleural effusion.

In 2020, Smelt and colleagues described the successful use of personalized three-dimensional-printed chest wall prostheses made of methyl methacrylate covered by expanded polytetrafluoroethylene for patients planned for chest wall resection and reconstruction[9]. In contrast, we used double prolene mesh for chest wall reconstruction as it provides excellent stability to maintain pulmonary function[9,17]. In our case, because of unavailability and cost factor, three-dimensional mesh was not used. However, the mesh was shaped into proper contour using a dome shaped towel in which the double prolene mesh was moulded.

A study of reconstruction of chest wall using a two-layer prolene mesh and bone cement sandwich done by Manouchehr and colleagues in 43 patients showed wide surgical resection is the most effective treatment for vast majority of chest wall tumours. They studied the clinical success of chest wall reconstruction using a prolene mesh and bone cement sandwich. There were two cases of ES. The study concluded chest wall reconstruction using a prolene mesh and bone cement prosthetic sandwich is a safe and effective surgical procedure for major chest wall defects[9].

Conclusion

Surgical excision followed by chest wall reconstruction using double prolene mesh with postoperative chemotherapy yields a good clinical outcome in patients with chest wall tumour. However, ES has a poor prognosis despite adequate treatment, thus, a regular follow-up with detailed evaluation should be done to look for relapse.

Ethical approval

Ethical approval was not acquired as this is a case report and not a large scale study. However, patient consent was obtained before preparing and submitting the manuscript.

Consent

Informed consent was obtained from participant.

Source of funding

No funding was secured for this study.

Author contribution

S.V., S.K., S.P., K.K.: manuscript writing and editing. R.M.K.: surgical procedure, patient care, manuscript writing, Guarantor. B.Y., S.S., S.B.: manuscript editing.

Conflicts of interest disclosure

None.

Research registration unique identifying number (UIN)

NA.

Guarantor

Dr. Robin Man Karmacharya.

Provenance and peer review

Not commissioned; externally peer-reviewed.

Availability of data and materials

All relevant data and materials are provided within the manuscript.

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