MULTIMODAL APPROACH IN ADOLESCENCE ASKIN TUMOR

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INTRODUCTION

Askin tumor is a primitive neuroectodermal tumor (PNET) developing from the soft tissues of chest wall and is part of Ewing's sarcoma family of tumors[1].
It is a rare tumor predominantly occur in children and adolescents mainly 15-18 years of age[2]. Sex distribution is still unclear [2].

AIM

We report a case of adolescence with Askin tumor successfully treated by surgery following neoadjuvant chemotherapy and postoperative radiotherapy with no early local recurrence.

CASE PRESENTATION

12 years old girl presented with 2 weeks history of cough, shortness of breath and weight loss. Chest examination revealed a decreased breath sound and stony dullness over left hemithorax.

Chest imaging (CXR, CECT and MRI Thorax) showed solid lesion at lateral 3rd rib 11.4x8.1x10.6cm encasing the rib with massive pleural effusion. Bone scintigraphy demonstrated increase tracer uptake over left 2nd-5th rib. USG guided biopsy showed small round blue cell tumor histomorphology and immunochemical studies compatible with Ewing sarcoma.

Post 5 cycles of neoadjuvant chemotherapy repeated MRI showed localised and more than 50% reduction of tumor mass.

She underwent left thoracotomy, en-bloc resection of left pleural mass, thoracic wall together with 3rd and 4th ribs Thoracic wall defect repaired with composite mesh. Postoperative recovery was uneventful.

Chemotherapy restarted on day 7 of surgery. HPE reported complete resection of the tumor with clear margin. She received radiotherapy 36 Gy in 20 fractions. Post-operative MRI thorax showed no evidence of locoregional tumor recurrence.

DISCUSSION

Askin tumor is a subset of Ewing's sarcoma which arise from chest wall. Askin tumor is characterized histologically by features of small round blue cell tumors and needs to distinguish among other tumors such as Ewing's sarcoma, rhabdomyosarcoma and neuroblastoma by immunochemical stain and cytogenetic studies as described by Askin et al. in 1979 [3]. Askin tumor are highly aggressive and local recurrence are common. The prognosis is poor with 2- and 6-year survival rates 38% and 14% respectively[4]. Mean survival rate is reduced to 11 months after local recurrence[4]. Due to large size masses at presentation, the tumor is usually non resectable primarily[5,6]. Sawin[5] proved that preoperative chemotherapy not only reduced tumor volume but also prolonged survival rates. This patient was treated according to AEWS 0031 chemotherapy regime. Complete surgical resection is also an important prognostic factor for survival as suggested by Christiansen and colleagues[2].

CONCLUSION

Askin tumor has a very poor prognosis. An aggressive multimodal treatment consisting of chemotherapy , radical surgical resection and post-operative irradiation may improve patients survival rate. Long term follow up is needed to asses the efficacy of the above approach

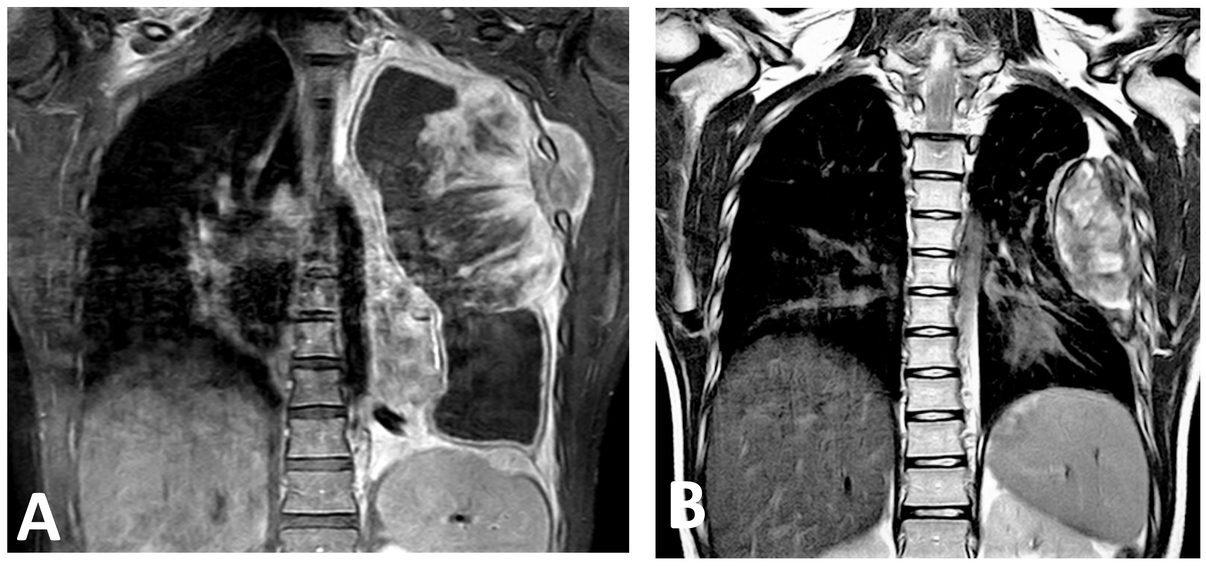

Figure 1. T1 Fat saturated post gadolinium coronal image before (A) and after (B)neoadjuvant chemotherapy showing localize mass with no pleural enhancement and resolved pleural nodule at lung base.

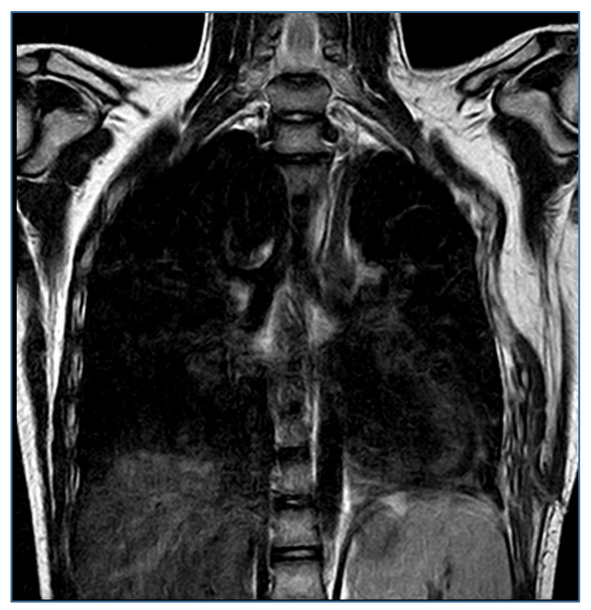

Figure 2. T1 Fat saturated post gadolinium coronal image postsurgery with no evidence of locoregional tumor recurrence.

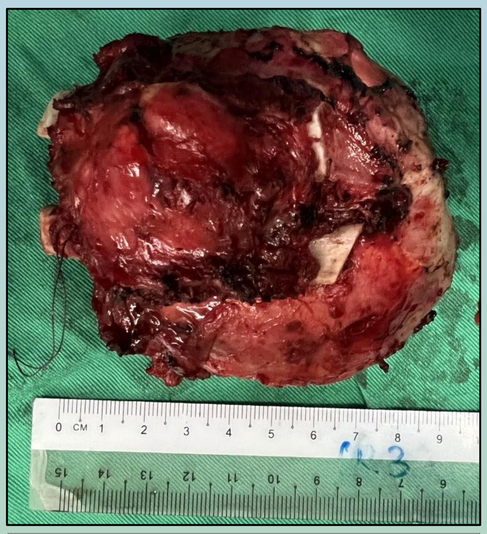

Figure 3. Surgical specimen showing subpleural extension of lesion into left 3rd and 4th ribs.

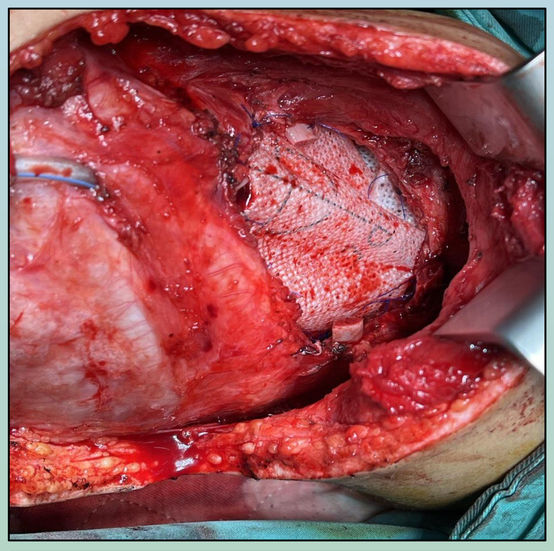

Figure 4. Chest wall reconstruction with composite mesh

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