Locally Advanced Askin's Tumour in a Child—a Rare Case Report and Review of the Literature

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Abstract Askin's tumor is a primitive neuroectodermal tumor (PNET) developing from the soft tissues of the chest wall and mainly occur in children and adolescents. The management approach is complex and requires a multidisciplinary team. We report a case of locally advanced Askin's tumour in a child because of its rarity and also to highlight the importance of keeping the possibility of PNET tumours in mind while dealing with cases of chest wall tumour in children.

Keywords Askin's tumour · Primitive neuroectodermal tumour · Thoracopulmonary region

Introduction

Askin's tumour is a rare tumour of childhood. Askin's tumor is a primitive neuroectodermal tumor (PNET) of the thoracopulmonary region described by Askin [1]. It develops from the soft tissues of the chest wall, particularly in the paravertebral region. Ewing's sarcoma, Askin's tumour and PNET are all considered as members of the Ewing's family of tumours; and when localised to the thoraco-pulmonary region, these are termed as Askin tumours [2]. We report a case of locally advanced Askin tumor in a child managed with multimodality approach.

Case Report

A 5 years-old boy presented in the oncology department with history of cough for 3 months and swelling in the right upper chest wall for 2 weeks which was gradually increasing in size and associated with mild pain. He had no history of fever. Physical examination revealed a mass measuring 3x3cm, firm, nontender and irregular margin in the right 2nd and 3rd intercostals space in the upper chest wall. On auscultation, breath sounds were decreased in the right chest. Laboratory investigations were normal except increased serum lactate dehydrogenase (LDH). His past history and family history were unremarkable. Chest X-ray revealed a homogenous large opacity in the right hemithorax with the deviation of the trachea and mediastinum to the left. The mountoux test was negative and none of his family members had tuberculosis. Contrast enhanced computerized tomography (CECT) of the chest showed a homogenous mass without any calcification measuring 10.4×8.6×8.1 cm in the right hemithorax arising from 2nd and 3rd ribs, involving the anterior mediastinum and

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encasing the ascending aorta, SVC and compressing the right bronchus and trachea (Fig. 1a). The mass was also extending anteriorly infiltrating into pectoralis minor muscle (Fig. 1b). Ultrasound guided core needle biopsy of the mass revealed small round cell tumour with scanty cytoplasm and relatively large nuclei. The IHC study of the tissue was strongly positive for PAS and CD99 but was negative for leucocyte common antigen (LCA), desmin, S-100 and PLAP (Fig. 2). Considering the site and histology, diagnosis of Askin's tumor was made. Metastatic workup in form of CECT abdomen-pelvis and bone marrow examination was negative. The child was administered six cycles of chemotherapy with Vincristine, Ifosfamide, Etoposide and Doxorubicin. The tumour regressed significantly with 6 cycles of chemotherapy. Post chemo CECT showed $4.6 \times 4.2 \times 1.7$ cm residual mass in the anterior mediastinum towards the right side without any extrathoracic extension (Fig. 1c and d).

In view of predominantly anterior based residual tumor with involvement of 2nd and 3rd ribs anteriorly in a young child, median sternotomy approach was planned as opposed to the conventional posterolateral thoracotomy approach to have a better access for the ribs and wider excision with disease free margins. Subsequently, patient underwent median sternotomy and the tumor was excised along with the 2nd and 3rd rib, intercostal muscles and part of pectoralis minor. Chest wall reconstruction was done using a 7×11 cm prolene mesh and the mesh was sutured to the defect margin taut enough to avoid any paradoxical movement in respiration. Intraoperatively, the mass was well encapsulated with adherent to the 2nd and 3rd ribs. The ipsilateral lung was compressed but was free from tumor invasion. Post operative

recovery was uneventful. The histological examination of the mass showed small residual tumor with sheets of small round cells and frequent Homer-Wright rosettes. The margins and ribs were free of tumour. Postoperatively, he received two cycles of adjuvant chemotherapy with ifosfamide and etoposide. Presently, he is clinically and radiologically disease free post 6 months of treatment completion. He is on regular three monthly follow-up with six monthly radiological evaluation till 2 years.

Discussion

In children, Ewing's sarcoma is the most common chest wall tumour. PNET of the chest wall is a rare malignant tumour seen in children and young adults [3]. Dickinson et al. in a 45-year database review found the prevalence of Askin's tumour to be of 0.2 cases per million [4]. Askin's tumour is now considered the same as PNET as no histological difference has been noted between Askin and non-thoracopulmonary PNET [3].

The common presentation in patients with PNET of the chest wall is chest pain, respiratory distress or a chest wall mass as in the index case [3]. A chest wall soft-tissue density mass, sometimes associated with rib erosion and/or pleural effusion, is the commonest radiographic manifestation [5]. In our case the mass was adherent to the 2nd and 3rd rib but not associated with pleural effusion.

The characteristic CT picture in the patient with Askin tumour/PNET is that of a heterogenous mass of chest wall origin with areas of necrosis and haemorrhage with or without

Fig. 1 **a** CECT showing a large homogenous mass in the right hemithorax involving the anterior mediastinum and encasing the ascending aorta, SVC and compressing the right bronchus and trachea. **b** CECT showing the mass extending anteriorly infiltrating into pectoralis minor muscle. **c** Post Chemotherapy CECT showing significant reduction in the mass. **d** Post Chemotherapy CECT showing significant reduction in the mass

Fig. 2 **a** Microphotograph showing diffuse sheets of small round cell tumour with scanty cytoplasm and relatively large nuclei. (Haematoxyline and Eosin stain). **b** IHC with PAS. **c** IHC with PAS with diastase showing cells sensitive to diastase. **d** IHC with CD99

an intrathoracic component [6]. Expansion of the tumour may cause compression of the underlying lung or may directly invade it. In our case the mass appeared homogenous without any cystic or necrotic components.

Ewing's sarcoma, PNET, rhabdomyosarcoma, neuroblastoma and lymphoma are small round cell tumours encountered in children and young adults [7]. IHC with PAS and CD99 helps in establishing the diagnosis. In our case, PAS and CD99 was strongly positive.

The accepted protocol for the management of this tumour is neoadjuvant chemotherapy followed by surgical excision of the tumor followed by post operative chemotherapy with or without radiotherapy [1–4]. The neo-adjuvant chemotherapy leads to better local control of the disease, less extensive surgery and treats the distant microscopic metastasis. Surgery with wide margins is ideal, but infrequently possible in patients with chest wall tumor. These are the patients who require post operative radiation. Neoadjuvant chemotherapy reduces the size of tumour significantly and can help in post operative clear margins, as in the index case. In view of post-operative negative margins, radiotherapy was not given in the index case. Sawin et al. demonstrated that pre-operative chemotherapy resulted not only in a reduced tumor volume, but also in prolonged survival rates [8]. Veronesi et al. summarized the benefits of pre-operative chemotherapy, including the reduction in the risk of intraoperative tumor rupture and tumor cell dissemination, the increase in the probability of R0 resection, the enhanced probability of post-operative function preservation by a more conservative surgical approach, the

decrease in any occult distant spread and the provision of a pathological and clinical evaluation of the response that favors the choice of the best post-operative regimen of chemotherapy and radiotherapy [9].

The prognosis of Askin's tumor is very poor as it is diagnosed at an advanced stage and the 5 year survival rates ranges from 10 to 60 % [3, 4, 8, 10]. The best prognosis can be provided by surgical treatment with wide resection. Indicators of poor prognosis include advanced age, metastatic disease, extraosseous primary tumour and recurrence [10].

Conclusion

Askin's tumour, a primitive neuroectodermal tumour of the thoracopulmonary region, is a rare tumour presenting in childhood. The diagnosis requires immunohistochemical workup supported by imaging investigations. The management of Askin tumor is a paradigm of cooperation between clinicians, surgeons and pathologists from establishing diagnosis to organizing the therapeutic strategy.

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REFERENCES

1. Jürgens H, Bier V, Harms D, Beck J, Brandeis W, Etspüler G et al (1988) Malignant peripheral neuroectodermal tumors—a retrospective analysis of 42 patients. *Cancer* 349–357
2. Askin FB, Rosai J, Sibley RK, Dehner LP, McAlister WH (1979) Malignant small cell tumor of the thoracopulmonary region in childhood. *Cancer* 2438–2451
3. Berg H, Rijn R, Merks J (2008) Management of tumors of the chest wall in childhood: a review. *J Pediatr Hematol Oncol* 214–221
4. Dickinson J, Watts AC, Robb JE (2009) Extra-osseous Ewing's sarcoma. *J Bone Joint Surg* 215
5. Sabati JM, Franquet T, Parellada JA, Monill JM, Oliva E (1994) Malignant neuroectodermal tumour of the chest wall (Askin tumour): CT and MR findings in eight patients. *Clin Radiol* 634–638
6. Winer-Muram HT, Kauffman WM, Gronemeyer SA, Jennings SG (1993) Primitive neuroectodermal tumors of the chest wall (Askin tumors): CT and MR findings. *AJR* 265–268
7. Linnoila RI, Tsokos M, Triche TJ, Marangos PJ, Chandra RS (1986) Evidence for neural origin and PAS-positive variants of the malignant small cell tumor of thoracopulmonary region ("Askin tumor"). *Am J Surg Pathol* 124–133
8. Sawin RS, Conrad EU III, Park JR, Waldhausen JH (1996) Preresection chemotherapy improves survival for children with Askin tumors. *Arch Surg* 877–880
9. Veronesi G, Spaggiari L, De Pas T, Solli PG, De Braud F, Catalano GP et al (2003) Preoperative chemo-therapy is essential for conservative surgery of Askin tumors. *J Thorac Cardiovasc Surg* 428–429
10. Baldini EH, Demetri GD, Fletcher CDM, Foran J, Marcus KC, Singer S (1999) Adults with ESE/PNET: adverse effect of older age and primary extraosseous disease on outcome. *Ann Surg* 79–86