Case Report

Askin's tumour- A rare case presentation

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Abstract

Askin's tumor is form of primitive neuroectodermal tumor developing from the soft tissues of the chest wall[1]. Here, we present a case of Askin's tumour in a 5-year-old male child who had presented with swelling and pain over left anterior chest wall. Small biopsy of the mass was done which confirmed the diagnosis.

Key Word: PNET. Askin's tumor.

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Received Date: 04/06/2019 Revised Date: 14/07/2019 Accepted Date: 21/08/2019
DOI: https://doi.org/10.26611/10141614

Access this article online	
Quick Response Code:	Website: www.medpulse.in
	Accessed Date: 15 October 2020

INTRODUCTION

Askin's tumor is a peripheral primitive neuroectodermal tumor within the thoracopulmonary region, which primarily occurs in children and young adults. It is a highly misdiagnosed and rare disease with a lack of clinical and pathological morphology in nature, which is easily confused with other small round-cell tumors[2].

CASE PRESENTATION

A 5 year old male child presented with swelling over left anterior chest wall in the 4th ICS and fever since 15 days. Swelling was associated with pain and was gradually increasing in size. He had no history of cough, breathlessness, hemoptysis, trauma. On examination, the patient was afebrile with pulse rate of 96/min, RR of 42/min and BP of 100/66 mmHg. Mild pallor was present and SpO2 was 97% at 3 litres of O2. Grade I clubbing was present. On Local examination, a tender hard mass of size 4x4 cm was palpable superolateral to left breast. On Auscultation, Breath sounds were reduced in left infraclavicular and mammary area. Bronchial breath sounds were heard in left mammary area. Lab investigations included CBC which revealed Hb- 11.1, WBC- 11300, Platelets- 4,67,000, ESR-40 (raised). LFT, RFT and Electrolytes were normal. LDH- 639 (raised). Chest X-ray revealed mass in between left 4th and 5th ICS with pleural effusion and lung collapse. Diagnostic pleural tapping was done which revealed hemorrhagic fluid. CT chest with contrast revealed left pleural effusion with left lung collapse. Mass lesion of size 5cm was noted in pleural space extending to extra-thoracic muscle plane. Small biopsy- of mass of left chest revealed malignant small round cell tumor suggestive of Askin's tumor (PNET). The patient was referred to higher centre for further management.

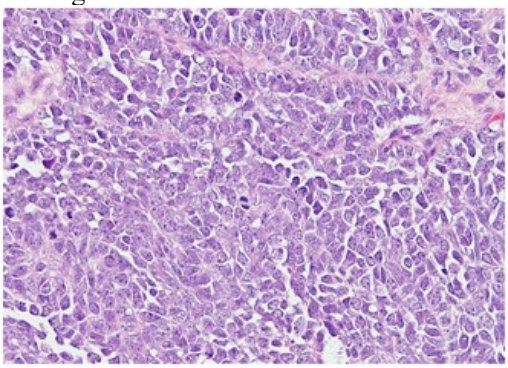

Figure 1: Histology Slide- Askin's tumor showing small round cells

DISCUSSION

Peripheral primitive neuroectodermal tumor (PNET) is a type of soft tissue sarcoma, described as arising intracranially[2]. PNETs arise from the primitive nerve

cells of the nervous system, but they can also occur outside the central nervous system (peripheral PNETs) in the chest wall, pelvis, extremities and so on[3]. PNETs of the chest wall were originally reported by Askin *et al* in 1979, since then, a PNET that occurs within the thoracopulmonary region is named as Askin's tumor. It has been reported to primarily occur in children and young adults[2]. It is a subset of Ewing sarcoma which arises from the chest wall. Histologically, it is distinguished by typical small round blue cells of monomorphous appearance. It is more frequent in males than infemales (1.5:1)[5]. This disease primarily occurs in the soft tissue of the chest wall, rib periosteum, chest and lung[2]. Askin's tumor usually presents with common respiratory symptoms. It may include symptoms like cough, chest pain, fever and breathlessness[5]. The commonest radiographic manifestation is a chest wall soft-tissue density mass, sometimes associated with rib erosion and/or pleural effusion[4]. Treatment of Askin's tumor consists of radical surgery, neo-adjuvant or adjuvant chemotherapy and radiotherapy. The best prognosis can be provided by surgical treatment with wide resection. As local recurrences after resection and metastases are frequently seen in Askin tumor, it has a poor prognosis and a short survival[6].

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Source of Support: None Declared
Conflict of Interest: None Declared