CASE REPORT

Bilateral Synchronous Askin's Tumor– Case Report & Review of Literature

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Introduction

Askin tumour was defined by Askin and Rosai in 1979. It belongs to Ewing's sarcoma family and originates paravertibrally in thoraco-pulmonary region [1]. This tumor is more common in children than adults, male to female ratio is 1.5:1 [2]. After extensive search in literature about the exact incidence we found that it is an extremely rare disease. In a study by Dickenson et al. the prevalence of Askin's tumour is 0.2 cases per million [3]. All of the cases in literature are unilateral, we report first case of synchronous bilateral Askin's tumor which responded to chemotherapy.

Case Report

A 15 years old young male came with swelling in left thoracic area of size 10*10 cm for 2 months which did

not respond to antibiotics and anti-inflammatory drugs. There was associated high grade fever for 1 month, dyspnea, cough. Patient lost 12 kg weight. Another swelling of size 3*4 cm in size appeared over right hemithorax posteriorly for 1 month (fig. 1). CECT thorax showed soft tissue mass eroding 11th rib on left side anteriorly and 9th rib on right side posteriorly, this mass also involved pulmonary parenchyma. Rest of the viscera was normal (Fig. 2). Wedge biopsy was taken from fungating mass on left side and histopathology showed small round cell tumor and immunohistochemistry (IHC) was positive with CD 99, confirming Ewing sarcoma (Fig. 3). FNAC done from right side lesion which showed small round cell tumor consistent with Ewing sarcoma. Patient was given vincristine, adriamycin and cyclophosphamide (VAC) based 6 cycles of chemotherapy. Patient showed complete response. Patient is now on regular follow up.

Discussion

Askin's tumor presents as soft tissue mass, often associated with dyspnea, cough, weight loss, or regional

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Fig. 1 Swellings in bilateral thoracic area

Fig. 2 CECT of thoracic cage showing Bilateral rib erosion

lymphadenopathy. Our patient had similar complaints for 2 months. Soft-tissue density mass, sometimes associated with rib erosion and/or pleural effusion, is the commonest radiographic manifestation [4]. Our patient also had rib erosion on CECT (Fig. 2). The diagnosis of Askin tumor rests on histopathological and immunohistochemistry findings. Table 1 shows diagnostic criteria for Askin's tumor proposed by Marina et al. [5].

Fig. 3 IHC Slide positive for CD99

Table 1 Diagnostic criteria for Askin's Tumor

1. Development of the tumor from a peripheral nerve;_
2. Identification of Homer-Wright rosettes;
3. Expression of NSE and Leu-7
4. Identification of t (11; 22) (q24; q12);
5. Detection of proto-oncogenes (N-myc, C-myb, C-ets-1);
6. Activity of neurotransmitter biosynthetic enzymes (tyrosine hydroxylase, dopamine B hydroxylase and acetylcholine transferase)

CECT is valuable for evaluating tumor extension at diagnosis, response to chemotherapy, and assessing tumor recurrence after surgery. MRI is may be considered complementary to CECT for very large tumors [6]. The aim of treatment for Askin tumor is to control local disease and distant metastasis. The preferred treatment option is neoadjuvant chemotherapy and surgical resection if tumor persists followed by adjuvant chemo and radiotherapy. Askin's tumor is chemosensitive and studies have shown that remission rate has improved from 26 % to 65 % over the period of time with aggressive chemotherapy [7]. Chemotherapy regimes that are used are VAC (vincristine, actinomycin cyclophosphamide), VACA (vincristine, actinomycin D, cyclophosphamide, adriamycin) and VAC alternating IE (ifosamide and etoposide). Systemic chemotherapy should be given early in the course of disease as it has high likelihood of early metastases and high recurrence rates even if the disease is organ-confined [8]. Criteria for early surgery are small initial volume of the tumor, good response to systemic therapy, an operable location and sufficient resection margins. However in our case the patient had bilateral Askin's tumor so he was given upfront chemotherapy as to avoid unstable chest wall and to prevent dissemination. The standard radiotherapy regimen was established by Miser et al. where the treatment includes exposure to 45–60 Gy plus cyclophosphamide/doxorubicin [9].

Poor prognostic indicators are advanced age, metastatic disease, extraosseous primary tumor and recurrence [10].

We conclude that Askin tumor is a important differential in small-cell tumor of the thoracopulmonary region, especially in the young age group. Patients with such tumors should be discussed in multimodality meetings and treatment should be individualized.

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Compliance with Ethical Standards

Competing Interests Authors declare that there are no conflicting and competing interests regarding this publication.

Informed Consent Informed consent was obtained from patient to report this case for educational purpose.

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