IJBC 2015; 7(3): 197-169

Askin Tumor Presenting as Superior Vena Cava Syndrome

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Submitted: 10-03-2014 , Accepted: 19-06-2015

Keywords: Askin tumor, primitive neuroectoderma tumor, superior vena cava syndrome, children

Introduction

primitive neuroectodermal tumor (PNET) of the chest wall (also known as Askin tumor) is a malignant tumor arising from the migrating cells of the neural crest which belongs to Ewing sarcoma family tumors (ESFT) [1-3]. Askin tumor with a high incidence of local recurrence and distant metastasis generally presents with symptoms of respiratory distress and painful mass in the chest wall [2-4].

Presentation of Askin tumor with superior vena cava (SVC) syndrome should be considered as an extremely rare manifestation and oncologic

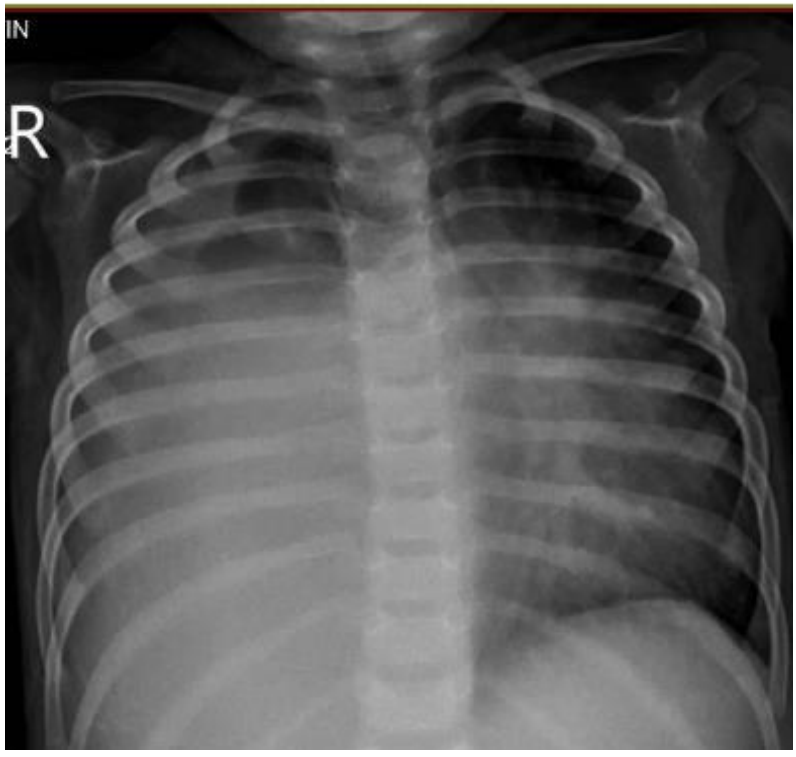

Figure 1: Chest radiography showing a homogeneous opacity in the right hemithorax

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emergency which could have a fatal outcome with complete blockage. To the best of our knowledge, this is the first case of Askin tumor in chest wall presenting with SVC syndrome in children.

A 2-year-old girl was admitted in our ward due to progressive respiratory distress and marked facial edema. Chest radiography showed a homogeneous opacity in the right hemithorax (figure 1).

Contrast-enhanced computed tomography of the chest revealed right upper mediastinal mass which was compressing the SVC and collateral vessels (figure 2). Thorough radiologic evaluation detected no metastasis. Mediastinal biopsy on the basis of morphologic characteristics and immunohistochemistry was compatible with the diagnosis of PNET. Therefore, the patient was scheduled to receive combination chemotherapy followed by surgical resection.

Histologically, PNETs are characterized by a sheet of small round blue cells that react positively to glycoprotein CD99. Homer–Wright pseudorossettes are a characteristic histological finding in PNET [3-6].

Figure 2: Contrast-enhanced computed tomography of the chest showing right upper mediastinal mass which was compressing the SVC and collateral vessels

Review of literature shows only one case of pulmonary Ewing's sarcoma described in a child less than ten years of age who presented with SVC syndrome caused by the intrathoracic mass compressing the SVC [3].

Gupta and colleagues have reported lymphoma as the most common cause of SVC syndrome 6. Another study demonstrated all the cases of SVC syndrome had a malignant etiology, the most common of which was T-cell acute lymphoblastic leukemia followed by lymphoma [7].

Our patient presented with SVC syndrome showing typical histological and immunohistochemistry features of Askin tumor.

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Despite its rare incidence, SVC syndrome due to Askin tumor should be kept in mind as a differential diagnosis of SVC syndrome in which early detection and prompt treatment can be lifesaving.

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