Case Report

Recurrent Askin's Tumor:Ewing's Sarcoma/Primitive Neuroectodermal Tumor of Chest Wall

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A case of recurrent Askin's tumor is presented occurring in a 13 year old boy. Askin's tumour is a rare primitive neuroectodermal tumor of chest wall and belong to Ewing family of soft tissue sarcoma. It is extremely malignant with a high frequency of both metastatic spread and of local recurrence. Multi-drug chemotherapy as well as local disease control with surgery and/or radiation is indicated in the treatment of all patients.
Key words: Askin's tumor, primitive neuroectodermal tumor, Ewing's sarcoma, chest wall tumor.

Pediatric soft tissue sarcomas are a group of malignant tumors that originate from primitive mesenchymal tissue and account for 7% of all childhood tumors[1]. Rhabdomyosarcomas, tumors of striated muscle and undifferentiated sarcomas account for more than one half of all cases of soft tissue sarcomas. The remaining non-rhabdomyosarcomatous soft tissue tumors account for approximately 3% of all childhood tumors[2]. Amongst these one sub-group is Ewing's family of tumors (EFTs). EFTs includes Ewing's tumor of bone, extra-osseous Ewing's sarcoma, primitive neuroectodermal tumors (PNET or peripheral neuroepithelioma) and Askin's tumor (PNET of chest wall). Studies using immunohistochemical markers, cytogenetics, molecular genetics and tissue culture indicate that these tumors are derived from the same primodial stem cell[2]. In some studies Askin's tumor and soft-tissue Ewing's sarcoma are considered as a single entity, malignant small- round-cell tumors[3].

Case Report

A 15-year-old boy presented in out- patient department at THQ Hospital Bhalwal, with a large swelling over left chest for the last six months. A smaller swelling at the same site was operated about one year back at a private hospital. The detailed examination revealed a swelling 15cm x 12cm, extending from just below the left clavicle to the 7th intercoastal space in midclavicular line and from left sternal border to the posterior axillary line in transverse plane. The nipple was displaced laterally; previous scar was visible and skin over the swelling showed areas of discoloration and ulceration. The swelling was non-tender, had bosselated surface with solid and cystic areas with tethering of the skin over the middle part of the swelling. It was not mobile. No axillary or cervical lymph nodes were palpable [Fig.1]. The histopathology report after the previous surgery was available showing a cellular neoplasm composed of sheets of small round cells, individual cells have scant cytoplasm, round hyperchromatic nuclei with fine granular stippled chromatin, no mitotic figures / necrotic areas identified.

Immunohistochemical stains were used, MIC-2 was diffusely positive; Desmin was negative. Diagnosis of Ewing's Sarcoma/PNET group of tumors was made. The patient, unfortunately, was not referred to any oncology department for further management after the first operation.

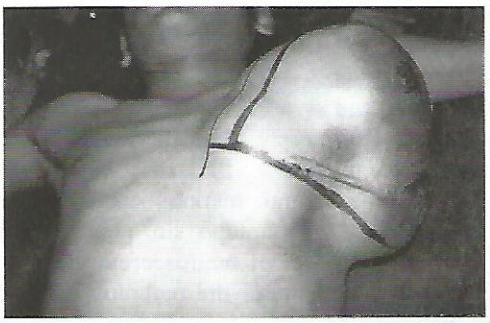

Figure 1: Pre-operative photograph showing the tumor and marking for the skin flaps.

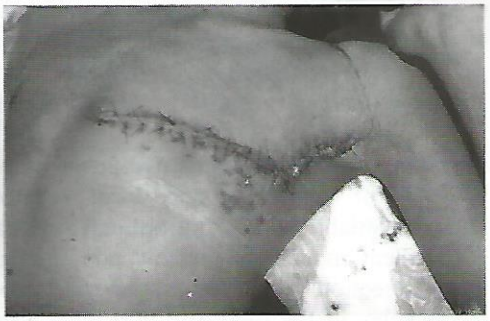

Figure 2: Post- operative photograph after one week showing adequate skin closure.

CT scan was done. It showed solid and cystic areas replacing all the soft tissues, encroaching on the chest wall and erosion of the ribs anteriorly, in the lower part of the swelling. No mediastinal, pleural or subphrenic abnormality was seen. The bronchovascular pattern was normal. The patient was prepared and Wide Excision of the tumor was performed. The tumor was very vascular with few solid and large cystic and necrotic areas replacing

the pectoral muscles. Part of the tumor was adherent densely to the precardial area where it had to be shaved. The intercoastal muscles were spared. Primary skin closure was done [Fig.2]. Histopathology showed recurrent Ewing's sarcoma / Askin's tumor. The patient made uneventful recovery; he was referred to Shaukat Khanum Cancer Hospital for chemo and radiotherapy. Regular follow-up is carried out.

Discussion

EFTs occur most frequently in the second decade of life and account for 4% of childhood and adolescent malignancies. The incidence in boys is slightly higher than in girls (ratio 1.1:1)[4]. Extraosseous Ewing's (EOE) and PNET comprise less than 40% of EFTs[5]. Common sites for PNET are chest 44%, abdomen/pelvis 26%, extremities 20%, head & neck 9% and all other sites 4%[6]. Rare sites such as Dura[7] and Esophagus[8] are also described in literature. The usual presentation is localized pain or a mass present for many months. There may be generalized symptoms such as fever, anemia and malaise and these are more common in patients with disseminated disease[9]. The MIC 2 gene product (CD99) is a surface membrane protein that is expressed in most cases of Ewing's sarcoma/ PNET family of tumors and is useful in the diagnosis of these when the results are interpreted in the context of clinical and pathologic parameters[1,10]. PNETs also show variable staining with some neural markers including neuron-specific enolase, leu-7, synaptophysin, neurofilament and S100[6]. The demonstration of neurosecretory granules by electron microscopy enhances the pathologist's ability to make the diagnosis of PNET[11]. In Ewing's sarcoma and related PNET, a t (11:22) translocation or a (21:22) rearrangement is associated with hybrid transcripts of the EWS gene with the FL[11] or ERG gene[12]. A molecular test (RT-PCR and restriction analysis of PCR products) currently available on a research basis only, now offers the opportunity of markedly simplifying the definition of EFTs[13,14].

Askin's tumors are extremely malignant with high frequency of both metastases and local recurrences. Limited data is available addressing optimal surgical and oncologic treatment modalities for this rare tumor. Age and surgical resection were found to be important prognostic variables in the treatment. No other variables such as tumor size, location, stage of disease or radiation therapy were found to improve survival. Surgical resection should be considered for all patients with Askin's tumor[15]. The successful treatment of patients with Askin's tumor/EFTs require the use of multi-drug chemotherapy, in addition to radiation therapy and / or surgical therapy to the primary tumor[4,5,16,17,18,19]. The prognosis for patients with recurrent or progressive disease is poor. The selection of further treatment depends on many factors, including the site of recurrence and prior treatment as well as individual patient considerations. Aggressive attempts to control the disease, including myeloablative regimens, may be warranted[20].

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