Askin Rosai Tumour

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Introduction

In 1979 Askin and Rosai reported a rare clinicopathological entity of malignant small-cell tumour of the soft tissues involving the chest wall in childhood and adolescence [1,2]. It was later recognized as the chest wall counterpart of the primitive neuroectodermal tumour (PNET) of the soft tissues [3]. The significance of recognition of this entity is its ominous connotation for poor survival [4].

Case Report

A 14 year old male presented with fever, weight loss and swelling right side of chest. Clinically, a diffuse lump was felt extending from the right posterior chest wall to the renal angle. Computed tomographic (CT) scan abdomen revealed a large lobulated mass measuring 7.7 x 4 cm on the right lower rib cage with erosion of the twelfth rib, involvement of intercostal muscles, right sided pleural effusion and right paraspinal soft tissue.

A CT guided fine needle aspiration showed a cellular smear composed of discohesive small round cells with scant cytoplasm, hyperchromatic nuclei and inconspicuous nucleoli. At places they formed Homer Wright rosettes around a delicate pink material. Mitoses were frequent (Fig. 1). A CT guided biopsy revealed sheets of small round hyperchromatic cells, infiltrating fibrocollagenous tissue with evidence of crushing and mitoses. The differential diagnosis on morphology included primitive neuroectodermal tumour (PNET), embryonal rhabdomyosarcoma and lymphoma. On immunophenotyping, the tumour cells expressed vimentin and MIC-2 (CD 99) (confirming the diagnosis of PNET). They were negative for desmin, muscle-specific actin (MSA) and leucocyte common antigen (LCA) (thus ruling out the other two diagnosis). A diagnosis of PNET of the chest wall (Askin Rosai tumour) was made (Fig. 2). Staging work-up included CT thorax which showed pleural involvement. Bone scan revealed multiple skeletal metastases and marrow metastases were detected on bone marrow aspiration (Fig.3). The disease

was staged as stage IV and induction chemotherapy was started which included etoposide, ifosfamide, vincristine, adriamycin and cyclophosphamide. He received four cycles of chemotherapy to which the response was dramatic. A repeat CT scan showed a reduction in the tumour size to 2x1.6cm with persistent rib erosion and a small pleural lesion. Bone marrow was negative. A repeat bone scan revealed persistence of disease hence the option of radical surgery was abandoned.

Discussion

Askin's tumour is a primitive neuroectodermal tumour presenting in children as a chest wall mass involving ribs. Establishing an accurate preoperative diagnosis of Askin's tumour poses a diagnostic challenge and is imperative both for instituting specific therapy and for prognosis [5]. In most instances it is diagnosed as sarcoma [2,6]. In our case, fine needle aspiration cytology (FNAC) and biopsy conclusively proved the diagnosis supported by immunophenotyping. The differential diagnosis includes embryonal rhabdomyosarcoma and lymphoma, which are excluded by attention to finer details like rosette formation and absence of rhabdomyoblasts. In recent times immunophenotyping has made the distinction more objective with demonstration of membrane-pattern positivity for MIC-2 (CD 99) (whereas muscle and lymphoid markers are negative). The pattern of staining is important as CD 99 cytoplasmic positivity can be seen in small cell variant of synovial sarcoma. A small proportion of rhabdomyosarcomas may be positive, hence positivity for a second neural marker (S-100, synaptophysin) and negativity for muscle markers makes the diagnosis more objective [7]. Its similarity with PNET is further established by detection of the EWS-FLI1 chimeric gene [3].

CT scan is valuable for tumour diagnosis, assessing the effects of chemotherapy and recurrence after

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Fig. 1 : CT-guided fine needle aspiration showing small round cells forming a rosette around neurofibrillary material. Cells have large nuclei, some showing nucleoli (Papanicolaou x 40).

Fig. 2 : CT-guided pleural biopsy revealed sheets of small round cells with hyperchromatic nuclei showing crushing, infiltrating fibrocollagenous tissue (haematoxylin & eosin x 10). Inset: Tumour cells show membranous pattern of MIC-2 positivity (IHC with DAB x 40).

surgery. Magnetic resonance imaging can determine chest wall muscle and marrow involvement. Neither is adequate for adjacent lung invasion. Bone, bone marrow and lung are the most frequent sites of metastasis.

Treatment includes radical surgical resection, neoadjuvant and adjuvant chemotherapy and radiation. Patients with Askin's tumour treated with aggressive pre-resection chemotherapy have smaller tumours to resect (less than 100 cc by volume) with improved survival [8]. In our case, there was gross reduction of the primary tumour after chemotherapy, but multiple bony metastasis persisted hence radical surgery was not performed. Overall the prognosis is poor. Occasionally intraoperative radiotherapy has been helpful [9]. In a large series of 975 cases of Ewing's sarcoma (bone counterpart of this tumour), the worst prognosis was seen in patients with bony metastases. Lung metastases alone carried a better prognosis [10]. The case has been

Fig. 3 : Tumour cells in cohesive clusters with moulding in bone marrow aspiration: note a megakaryocyte at the arrow (Leishman x 4). Inset: Tumour cells showed rosette formation at places (Leishman x 10).

presented to draw attention to its diagnostic dilemmas, since therapy and prognosis are influenced by it.

Conflicts of Interest

None identified

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