THORACIC NEUROBLASTOMA WITH SPONTANEOUS REMISSION

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Introduction

Neuroblastomas are tumours of infancy and childhood. They arise from cells of neural crest origin in the adrenal medulla and sympathetic ganglia. Neuroblastomas are one of the most common malignant tumours of childhood, with 40% arising in the adrenal glands and 15% in the chest [1]. They generally have good prognosis. They have propensity to occasionally undergo spontaneous regression. An incidentally detected, histologically confirmed case of neuroblastoma, which showed spontaneous remission is presented.

Case Report

A six week old female infant was admitted with history of cough and running nose of five days duration. She was born as full term normal delivery (FTND) with birth weight of 2.25 Kg to G2P1 mother with no postnatal complications. Chest radiograph PA and lateral projections revealed a posterior mediastinal mass. She was subjected to CT scan chest, which confirmed a soft tissue density mass measuring 3.3x3 cm in the posterior mediastinum to the left, paraspinal in position, lifting the left main bronchus and the left pulmonary artery anteriorly without evidence of invasion of adjacent vertebral body, rib or surrounding lung tissue, no intraspinal extension was appreciated suggesting a diagnosis of neuroblastoma (Fig-1).

Routine investigations were within normal limits. Bone marrow biopsy and urinary catecholamine metabolites were within normal limits, CT scan guided biopsy aspirate showed a highly cellular smear with small round to oval cells with minimal cytoplasm hyperchromatic nucleus lying in sheets. In few areas occasional pseudo rosettes of tumour cell were seen in the centre of which there was fibrillar pink material and diagnosis of neuroblastoma was confirmed.

Cough and running nose responded to symptomatic treatment. The child was discharged and parents were advised to bring back the child for review after three months or earlier in case she developed any symptoms. Child remained asymptomatic and CT scan chest was repeated on review after three months. The posterior mediastinal mass showed spontaneous regression to a 8 mm

rounded residual soft tissue density with a faint rim calcification without involvement of surrounding tissues, with left main bronchus reverting to its normal position (Fig-2). CT scan chest was once again repeated after another four months which showed findings same as Fig-2 suggesting spontaneous remission.

Fig. 1 : CT scan chest, section at the level of main bronchi demonstrates neuroblastoma lifting the left main bronchus and the left pulmonary artery which is seen as a small nodule

Fig. 2 : CT scan chest section at the same level as fig. 1 shows spontaneous regression of the lesion

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Discussion

Neuroblastoma is a malignant tumour of infancy and childhood with 75% cases presenting before four years of age. Extra-abdominal tumours (chest, neck, and pelvis) do better in terms of survival than tumours arising from upper abdomen [2]. A minority of children with thoracic neuroblastoma will have dumbbell tumours suggesting intraspinal extension [3]. In two third of cases, tumour is widely disseminated at the time of diagnosis. The metastatic spread is usually to the skeleton, bone marrow and lymph nodes. It has some very interesting peculiar features of good prognosis, even with disseminated disease. It has a propensity to occasionally undergo spontaneous regression [4]. However, no figures have been quoted in available literature. It also has ability to undergo spontaneous or induced differentiation to a benign ganglioneuroma. Neuroblastoma may originate anywhere along the sympathetic nervous system chain. The most common site of primary tumour is, however, within the abdomen either in the adrenal gland or in paraspinal ganglions. Kaneko et al [4] treated 36 patients of less than 12 months of age between 1982 and 1997 of which 7 had no surgery, 21 underwent partial tumour resection with or without chemotherapy, remaining had complete resection and are alive and doing well without tumour, indicating that complete resection is not required in patients with neuroblastoma under 1 year of age.

Slovis et al [3] observed that in thoracic neuroblastoma chest radiograph was 100% sensitive in suggest-

ing the diagnosis. MR imaging was 100% sensitive in predicting enlarged lymph nodes, intraspinal extension, and chest wall involvement. CT scan was 88% sensitive for intraspinal extension but only 20% sensitive for lymph node enlargement. CT scan was 100% sensitive in detecting chest wall involvement. Neither test was able to detect remote disease, as noted by bone scan.

Extra abdominal neuroblastomas show better prognosis in first year of life with ability of spontaneous regression. MR imaging appears to be single best imaging modality following the chest radiograph. Child should be kept under observation. Surgery should be contemplated only if the child is symptomatic and complete resection is not required in patients under one year of age [4].

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