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Case Report

Infantile Bilateral Adrenal Neuroblastoma with Preauricular Lymph Node Metastasis

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Abstract

Neuroblastoma (NB) is the common extra-cranial solid malignancy of early childhood [1]. Adrenal medulla is the most common site. Thirty-five percentage of cases are diagnosed in children younger than 1 year [2]. rarely only 1% of patients have bilateral adrenal neuroblastoma [1]. Incidence of stage 4S is most common in the reported cases of bilateral adrenal NBs. To the best of our knowledge, this is the first case of bilateral adrenal neuroblastoma presenting with preauricular lymph node metastasis.

Abbreviations: NB: Neuroblastoma; FNAC: Fine Needle Aspiration Cytology; ICMR: Indian Council of Medical Research; BANB: Bilateral Adrenal Neuroblastoma; INSS: International Neuroblastoma Staging System

Introduction

Neuroblastoma (NB) is the common extra-cranial solid malignancy of early childhood [1]. Most common site is adrenal medulla. NB is derived from neural crest ectoderm. Thirty- five percentage of cases are diagnosed in children younger than 1 year [2]. Males are more commonly affected with a ratio of 1.2:1. In patients presenting with bilateral, multifocal a hereditary predisposition has been known. In such cases, the median age of diagnosis is nine months. However, rarely only 1% of patients have bilateral adrenal neuroblastoma [1]. To the best of our knowledge, this is the first case of bilateral adrenal neuroblastoma presenting with preauricular lymph node metastasis.

Case Report

An eleven-month male infant presented with swelling in right preauricular region noticed by mother for four months. He was second born to non-consanguineous parents. He was born at full term with birth weight was 2.700 kg. The infant had firm nodular swelling of 1 cmx 1 cm in right preauricular region. Systemic examination was normal.

A Fine needle aspiration cytology (FNAC) was done from the right preauricular swelling. Morphology showed lymph node infiltrated by neoplastic small round blue cells arranged in nests with homer wright rosettes with a typical mitotic figures (22/10 high power field). There was abundant neuropil. MKI (Mitotic-karyorrhexis index) was 110. Features were consistent with metastatic neuroblastoma (schwannian stroma poor), differentiating type, intermediate MKI-favourable histology. Immunohistochemistry was positive for synaptophysin and chromogranin. N-MYC amplification was negative.

Contrast CT abdomen and pelvis showed irregular heterogeneously enhancing lesion involving left adrenal-4.4x1.4 cm (APXTR), abutting adjacent spleen and stomach with loss of fat planes. Right adrenal also showed well defined heterogeneously enhancing lesion with areas of coarse calcification measuring 3.6x2 cm (APXTR). Another well-defined heterogeneously enhancing soft tissue density with coarse calcification was found in left paravertebral location with extension T8-T10 measuring 2.7x1,8x3.7 cm (APXTRXCC). A few heterogenous lesions were noted in pre-aortic and para-aortic

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location- largest measuring 3.7x3.1x5.3 cm. It was seen to partially encase abdominal aorta and superior mesenteric artery. Impression was bilateral adrenal neuroblastoma with left paravertebral and retroperitoneal lymph node involvement. Urine VMA/creatinine ratio was 116.045 mg/g creatinine (normal biological reference range for 6 months-3 years:<13). A Ga-68 DOTANOC PET scan for staging showed similar abdominal findings. A pleural based nodule in anteromedial basal segment of left lower lobe lesion of 19x27x55 mm was also noted. Therefore, final impression was metastatic bilateral adrenal neuroblastoma.

Though metastatic disease, considering age and N myc status, child was stratified as intermediate risk as per ICMR (Indian Council of Medical Research) guidelines. He was treated with neoadjuvant chemotherapy- six cycles of carboplatin, etoposide alternating with cyclophosphamide, adriamycin and vincristine. A reassessment after neoadjuvant chemotherapy with PET scan showed regression in size and uptake in bilateral adrenal with resolution of metastases. He underwent left adrenalectomy with right adrenal sparing surgery. Post operative histopathology was suggestive of cytodifferentiation towards ganglioneuroblastoma. Hence, child was kept on observation. Currently, child is one year post therapy and disease is in remission.

Discussion

Neuroblastoma was initially referred by Dr. Rudolf Virchow as an abdominal "Glioma" [3]. The term NB was coined by James Homer-Wright in 1910 [4]. The cell of origin is primitive sympathetic ganglion cells (neural crest cells) [1]. Constitutes 8–10% of all childhood cancers [1]. Forty-six percentage of NB arise from adrenal gland [5]. Only one percent of patients have bilateral adrenal neuroblastoma. Bilateral adrenal NB is mostly synchronous at presentation. Heterochronous involvement of bilateral adrenal NB is reported only in three cases in literature [1]. Antenatal mothers with intrauterine NB infants have had hypertension, headache, sweating and flushing. This is key for perinatal detection [6]. But in our case, there was no history of maternal hypertension.

Neuroblastoma has been called "the great mimicker" because of its varied clinical manifestations related to the site of the primary, metastatic sites and its tumor markers [7]. Patients with Bilateral Adrenal Neuroblastoma (BANB) are usually younger with distant metastasis. Inspite they have a favourable outcome [8,9]. Multicentric growth of neuroblastic nodules is regarded as pathogenesis for multifocal neuroblastoma [7].

Staging for bilateral adrenal neuroblastoma was carried out in accordance with the International Neuroblastoma Staging System (INSS), in which it is recommended to define the stage separately for each adrenal gland. Common stage of the disease is determined by the most affected adrenal gland. In the diagnosis to indicate a bilateral adrenal lesion should be indicated by the stage Arabic numerals with the letter "M", which means multifocal tumor.

In management, bilateral adrenal resection is associated with the definite complication of adrenal insufficiency. So, bilateral adrenalectomy is generally deferred. Prognosis of is good even in case of progression in bilateral disease. In a study by Andreeva et al, in 29 cases of BANB, the three-year event-free survival was 86 ± 6.4%. The three-year overall survival rate was 100%. The shows that they have a more favourable course of the disease [10]. Child is on follow up for a period of one year and currently in remission.

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