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Posterior Mediastinal Neuroblastoma in 4years old child - a Case Report

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Abstract*: A 4years old malechild presented with cough, dysponea and recurrent pulmonary infection. The CT and MRI Chest revealed a huge right side posterior mediastinal mass with no evidence of metastasis. He underwent surgical exploration with successful enblock removal of the mass, the biopsy was suggestive of neuroblastoma. He was discharged on the 6th post operative period and is in regular follow up.*
Keywords*: posterior mediastinum,Neuroblastoma,dysponea,recurrent pulmonary infections*

I. Introduction

Neuroblastoma is an embryonic tumor of the sympathetic nervous system. It is derived from neural crest cells, arising within the adrenal gland or anywhere along the peripheral sympathetic nervous system. It is the most common cancer of infancy and the fourth most common type of cancer in children.2/3rd cases presents in children younger than 5 years. Patients with localized neuroblastoma (stage 1) have excellent event-free survival (EFS) rates with surgical excision. Five-year overall survival rate for children with neuroblastoma is about 85% for infants, 55% for children aged 1-5 years, and 40% for children older than 5 years.

II. Case Report

A 4 years old male male child presented with productive cough, progressively worsening dysponea, and recurrent pulmonary infections for last 6 months. There was no history of tuberculosis. Haematological investigation showed increased TLC. The chest X-ray showed opacity in the righthemithorax and collapse of the right lung. Pulmonary function tests were impaired: FVC 1.16 (37.2% of predicted) and FEV1 1.04 (42.3% of predicted). CT and MRI Scan revealed a hugemass originating in the posterior mediastinum and occupying almost whole of the righthemithoraxcompressing the heart, great vessels and hilar structure with mediastinal shift towards the left side . The mass showed heterogeneous density. Urine examination revealed elevatedhomovanillic acid (HVA) and/or vanillylmandelic acid (VMA) levels. The patient underwent Right thoracotomy with en-block removal of the mass. Postoperative period was unremarkable The biopsy showed a mass of 14 x 8 x 2.5 cm weighing 1.5 kg with variegated appearance . Multiple sections revealed encapsulated lobulated tumor composed of closely packed sheet of hyperchromatic cells separated by thin incomplete fibrous septae.OccassionalHomer-Wright type of rosettes were also present. All the findings were consistence with neuroblastoma. He was discharged on 6th postoperative day and is in our regular follow up.

III. Discussion

Neuroblastoma is the most common malignant extracranial tumor of childhood (accounting for 8-10% of all childhood cancers). In infants (<12 months) neuroblastoma is the most common type of all malignancies (58/1,000,000 infants per year). Peak incidence is at 2-3 years[1,2].Prenatal neuroblastoma is occasionally picked up on maternal ultrasonography.Male to female ratio is 1.3:1. Thoracic neuroblastomas (posterior mediastinum) may be asymptomatic and are usually diagnosed by imaging studies obtained for other reasons. Presenting signs or symptoms may be insignificant and involve mild airway obstruction or chronic cough. Thoracic tumors extending to the neck can produce Horner syndrome. Tumors that arise from the paraspinal sympathetic ganglia may result in the presence of neurologic symptoms, including weakness, limping, paralysis, and even bladder and bowel dysfunction.However, patients may also present with unexplained fever, weight loss, irritability, and periorbital ecchymosis secondary to metastatic disease to the orbits. The presence of bone metastases can lead to bony pains and pathologic fractures.[2]

Approximately two thirds of patients with neuroblastoma have abdominal primaries. Symptoms produced by the presence of the mass depend on its proximity to vital structures and usually progress over time. Tumors can develop in the abdominal cavity (40% adrenal, 25% paraspinal ganglia) or other sites (15% thoracic, 5% pelvic, 3% cervical tumors, 12% miscellaneous) In 60-70% of cases the disease is not diagnosed until it has already metastasized. More than 90% of patients have elevated homovanillic acid (HVA) and/or

vanillylmandelic acid (VMA) levels detectable in urine [3].CT Thorax confirms the location and extent adjacent organ involvement, or vascular involvement. [4,5]CT demonstrates calcification in up to 90%. MR imaging is the investigation of choice for better delineation of full extent of mass, extradural/intraspinal extension and chest wall invasion.[5,6] Treatment strategies are tailored to risk category (low, intermediate, or high) assigned on the basis of age, stage, *MYCN* gene amplification, DNA ploidy, and histology.[7]Response to treatment (which consists of surgery, chemotherapy, and radiotherapy) depends on the age of the patient, the extent of spread, tumor histology, and the genetic and molecular characteristics of the tumor cells .The International Neuroblastoma Staging System (INSS) is used for clinical purposes. Patients with localized tumors (regardless of age) have an excellent outcome (80-90% 3-year event-free survival [EFS] rate), with surgical excision of tumor only. Adjuvant chemotherapy is generally not needed for this group of patients.For patients with regional or metastatic disease,adequatesamples(biopsies) are essential for planning of chemotherapy /radiotherapy. Postchemotherapy,second-look surgery can be used to attempt a complete resection.[8]

Follow-up care includes physical and clinical examination, monitoring of urinary catecholamines, and diagnostic imaging. Because most recurrences occur during the first 2 years following treatment, most protocols recommend close follow-up care during this interval.Patients who remain free of recurrent disease for 5 years are considered cured.[8,9]

IV. Conclusion

Neuroblastomais a chemosensitive pediatric malignancy.Treatment (in the form of surgery, chemotherapy, or radiotherapy) depends on the age of the patient, extent, tumor histology, genetic and molecular characteristics of the tumor cells.Surgical resection should be considered forlocalisied disease as it provides excellent outcome and survival rate.

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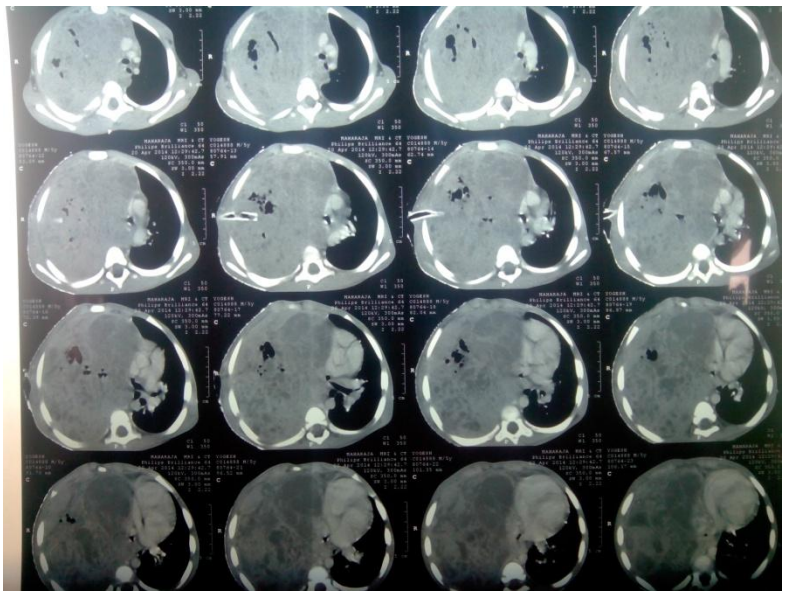

Figure 1. CT Thorax showing posterior mediastinal mass

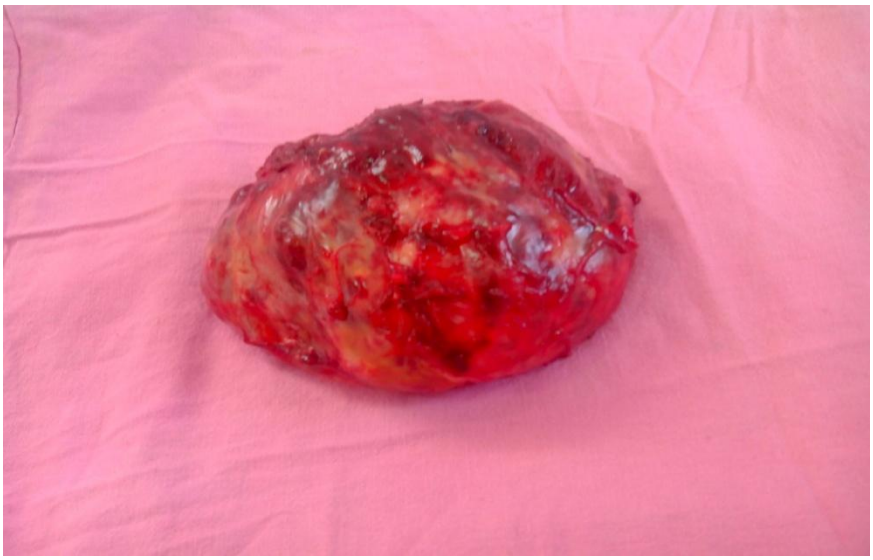

Figure 2: En-Block Resected specimen of the posterior mediastinal mass

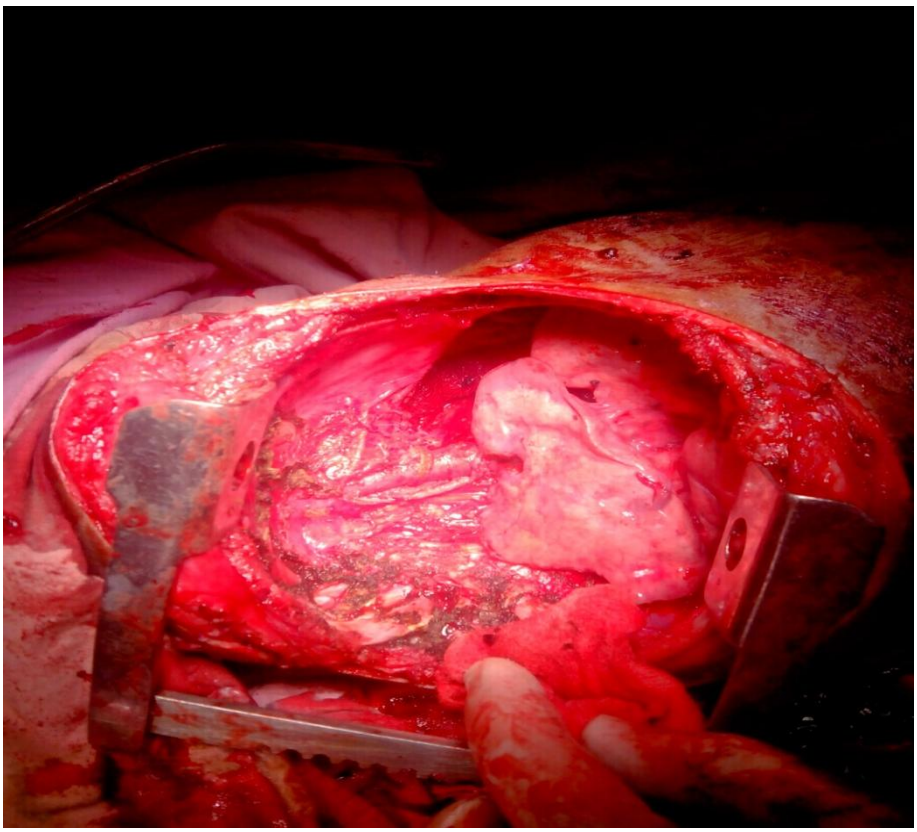

Figure 3 : intra operative picture showing the thoracic cavity after complete removal of the mediastinal mass