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Retroperitoneal Ganglioneuroblastoma in 10 years Old Patient: a case report

KEYWORDS: Ganglioneuroblastoma, tumor, retroperitoneal

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ABSTRACT *Ganglioneuroblastoma is a rare tumor arising from undifferentiated neural crest cells, with malignant potential. It occurs equally in both sexes with usual presentation in young children. It may present as metastatic disease requiring histological diagnosis, with CT and MRI being modalities of choice. Treatment remains surgical excision with adjuvant therapy.*

INTRODUCTION:

Neuroblastomas are embryonal cancers of peripheral sympathetic nervous system[1]. Neuroblastomas are most common extra-cranial solid tumor in children & the most commonly diagnosed malignancy in infants[2]. Ganglioneuromas, ganglioneuroblastomas and neuroblastomas differ from each other in terms of neuroblast maturation. Ganglioneuroblastomas are less mature form and regarded as more aggressive tumors. The median age of children at diagnosis is 22 months & 90% of cases are diagnosed by 5 years of age, rarely develop in children over 10 years[3]. These tumors may develop at any site of sympathetic nervous system tissue. Approx. half of these tumors arise in adrenal glands & other common sites are paravertebral, retroperitoneum, posterior mediastinum, pelvis & cervical area. Clinical features vary widely and are frequently nonspecific (asthenia, sweating, fever, pain related to the primary tumor or metastatic lesion & abdominal distention). Children under age of 2 years, generally present with a large abdominal mass, fever & weight loss. In older children, the cause may not be recognized until metastatic disease produces bone pain, respiratory problems & gastrointestinal disturbances[4].

Here, we present the case of 10 years old previously healthy female who was diagnosed to have retroperitoneal ganglioneuroblastoma.

CASE REPORT:

A 10 years old female patient presented in surgical out patient department with complain of distention of abdomen since 2 months. The patient was examined clinically and large lump was found in left hypochondrium & left paraumblical region which was non mobile, non tender, not moving with respiration, not fall forward in knee-elbow position & cystic in consistency. USG abdomen and IVU showed left kidney was displaced inferiorly & suppressed superiorly along the superior pole of a large intra-abdominal cyst located in left flank. CECT abdomen showed significantly large sized relatively well defined encapsulated non-infiltrating intra-abdominal/ retroperitoneal cyst (10×11.7×14.2 cm) within left flank with multiple internal cysts separated by variable sized thickened septae, located between spleen and left kidney & is significantly compressing and displacing the spleen superiorly and kidney inferiorly. The adjacent bowel loops are displaced to the right aspect of the abdomen. Left kidney shows normal excretion of contrast and no obstructive changes seen. The patient was managed surgically in the form of exploratory laparotomy with excision of cyst. Intraoperatively a large (approx. 15×12 cm) encapsulated cyst in left supra renal region which on cut section revealed hemorrhagic fluid with multiple septae through the cyst was found. (Fig1,2).

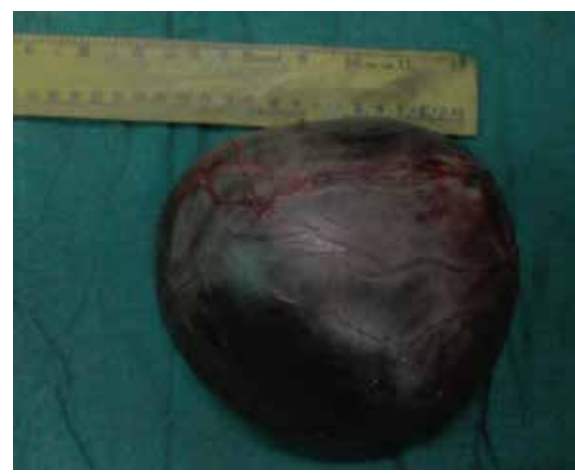

Fig 1: Excised Specimen

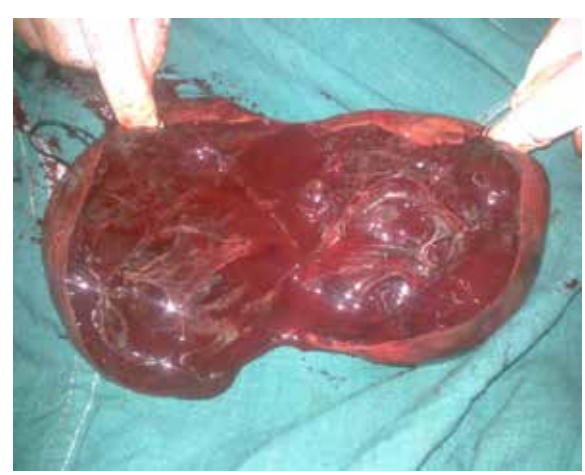

Fig 2 : Cut section of the specimen.

Histopathology showed ganglioneuroblastoma, intermixed

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type with no evidence of metastasis. The patient underwent two cycles of chemotherapy with regular clinical and imaging assessments. The patient was regularly followed every 3 months.

DISCUSSION:

Ganglioneuroblastoma is a peripheral neuroblastic tumor which is found very rarely. It arises from undifferentiated cells of neural crest. They have intermediate malignant potential between neuroblastoma and ganglioneuromas. Ganglioneuroblastomas occur in both genders equally. It is found more commonly in young children upto 10 years of age[5]. They occur most commonly in adrenal medulla, extraadrenal retroperitoneum and posterior mediastinum. They produce catecholamines which are important diagnostic markers. These tumors may regress spontaneously. The prognosis of ganglioneuroblastoma is relatively good. Neuroblastomas can produce metastatic disease. Most commonly it metastasize to liver, lung, bones. Children younger than a year old may present with multiple skin metastasis associated with bluish discoloration (bluberry muffin syndrome)[6]. Patients with this tumor usually present with pain, either by primary tumor or by metastatic disease.

Histologic confirmation is required for definitive diagnosis. Ultrasonography is first line method for identifying this tumor. CT & MRI are imaging modalities of choice[7]. The prognosis of patients with localized disease and younger age is better. Outcome of favorable or less aggressive neuroblastoma is worse in older children[8]. Mediastinal neuroblastoma are better in prognosis than abdominal neuroblastoma as the former tend to present earlier when the size is small and hence complete resection of tumor is possible[9].

CONCLUSION:

The rare occurrence of ganglioneuroblastoma along with its presentation in pediatric population result in a great deal of suspicion when younger child present with symptomatic mass as in our case. So, it should be kept in one of the differential diagnosis.

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