

Retroperitoneal nodular ganglioneuroblastoma in adult: An unusual case report

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ABSTRACT

Neuroblastic tumor are from embryonic tumor of the sympathetic nervous system having neural crest origin and arise in the adrenal medulla, the adult population. Here, we present a case of an adult male of 23-year-old with retroperitoneal nodular ganglioneuroblastoma metastasized to the adrenal gland and regional lymph node. With the best of the knowledge, fewer than 50 cases reported in the English medical literature.

Key words: Adrenal gland, ganglioneuroblastoma, neural crest, retroperitoneum

INTRODUCTION

Ganglioneuroblastoma (GNB) was defined as “a transitional tumor of sympathetic cell origin that contained malignant neuroblastomatous and benign ganglioneuromatous element” by Robertson in 1915.^[1] Origin of neural tumors of the sympathetic chain can be found anywhere from neck to retroperitoneum, histologically represented by the categories of neuroblastoma, ganglioneuroblastoma, ganglioneuroma conceptualized as three morphological maturational manifestation of a common neoplasm. They exhibit a broad range of differentiation from neuroblastoma at one end, ganglioneuroma at the other and in between. There is a continuum between these categories, both morphologically and at the level of developmental gene expression. GNB is usually less aggressive and response to therapy with more favourable prognosis than neuroblastoma.

CASE REPORT

The present case report is about a 23-year-old male patient who was admitted to the hospital due to abdominal, lower back pain and discomfort. On admission, physical

examination revealed a left upper quadrant large abdominal mass. A multi-slice computed tomography scan of the abdomen and pelvis showed a huge mass measured (8.6 × 9 cm) in retroperitoneum anterior to the left kidney and posterior to pancreas with internal foci of calcification. Left adrenal also showed (3.4 × 2.8 × 2.0 cm) internal necrotic area suggested the possibility of adrenal metastasis. From the findings of imaging studies, we concluded this may be a huge retroperitoneal tumor. Blood level of catecholamine, renin activity, aldosterone, cortisol and dehydroepiandrosterone sulfate were not significantly elevated. Pre-operative biopsy was non-diagnostic. Patient underwent exploratory laparotomy with en bloc removal of left huge retroperitoneal mass with left nephrectomy, ipsilateral enlarged adrenal gland and regional lymph nodes. The specimen was received for histopathological examination. Perioperative and post-operative was uneventful.

Grossly tumor was well-circumscribed lobulated with the outer surface intimately adhered to left kidney pelvis area. On the cut surface mass shows variegated appearance, soft yellow and gray brown nodules, areas of necrosis and calcification [Figure 1]. On Histopathology shows small round hyperchromatic nuclei of neuroblastic cells forming nodules and various stage of maturation of it to ganglionic differentiation in a neurofibrillary background and more than 50% schwannian stroma [Figure 2a]. Homer Wright rosettes comprise of central collection of neuropil surrounded by a mantle of neuroblast [Figure 2b] and area of calcification was also seen. We sign out the report as Ganglioneuroblastoma. Immunohistochemistry showed

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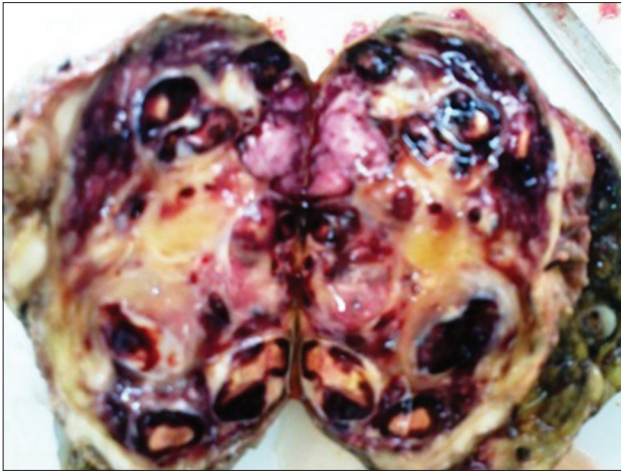


Figure 1: Gross specimen shows yellowish nodule in white fleshy surrounding hemorrhage and area of necrosis

neuron specific enolase (NSE), S-100, neurofilament protein and chromogranin positivity again support the diagnosis.

DISCUSSION

GNB is a common type of tumor in children but an extremely uncommon in adults. Histogenesis of these tumors are derived from the mantle layer of developing spinal cord and populate the primordia of the sympathetic ganglia and adrenal medulla.^[2] The incidence rate of GNB was estimated at 6.8/1 million of the white population, occurring in childhood and rarely appears in adult. More than 90% of cases are reported in children younger than 5 years of age, with a peak at 1.5 years.^[3] GNB occur most frequently in the midline along sympathetic chain, although uncommonly located in thymus, cauda equina, pineal gland and bronchus of lung.

In one series study of 80 cases of GNB, only three patients were above 20 years of age.^[4] According to Yamanaka *et al.*, only 33 cases of retroperitoneal GNB in adult have been noted in English medical literature.^[5] Mehta *et al.* in their study have reported case of bilateral intra-abdominal GNB in age of 20 years in Indian population.^[6] In an article in 1976, Kilton *et al.*, published a review of 33 cases of GNB in adult world-wide.^[7]

In adult GNB is generally discovered by accidentally or by compression symptoms, as in the present case. Radiological modality cannot distinguish definitely between pheochromocytoma, ganglioneuroblastoma, adrenal and other retroperitoneal malignancy. Pre-operative biopsy may produce result depending upon which component is biopsied, can look like a small round cell tumor or ganglionic cell component in schwannian stroma. The major differential diagnosis is extra skeletal Ewing's sarcoma/primitive neuroectodermal tumor, alveolar

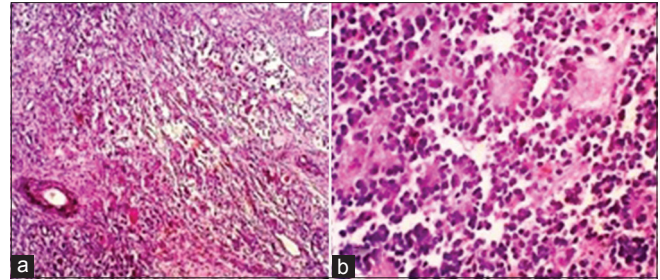


Figure 2: (a) H and E ($\times 100$) shows ganglion cell having abundant eosinophilic cytoplasm and eccentric vesicular nuclei in a schwannian stroma (b) H and E ($\times 400$) shows neuroblast forming Homer-Wright rosette with a central core of neuropil material

rhabdomyosarcoma, desmoplastic small round cell tumor, paraganglioma and adrenal malignancy.

There are two subtypes described in the literature, nodular and intermixed type, both have prognostic significance.^[2] In nodular type neuroblastic nodule surrounded by typical ganglioneuromatous stroma, whereas intermixed type show small nest of neuroblastic cell present in ganglioneuromatous stroma. The pattern of spread of the tumor is mainly regional lymph node, adjacent organ, bone marrow (78%), bone (69%), lymph node (42%), liver (20%), intracranial structures (7%), skin (2%) and testis (2%).^[8] The international neuroblastoma pathology classification uses age, neuroblastic maturation, schwannian stromal content, nodular or intermixed GNB and mitosis-karyorrhexis index in a nodule as a prognostic indicator, which distinguishes 2 prognostic subsets in this category. They are favorable and unfavorable histology. Prognosis also depends on margin of surgical resection. The overall favorable prognosis of GNB is related to the fact that most tumor are localized and have intermixed rather than nodular features.

Various ancillary test such as serum lactate dehydrogenase, serum ferritin, NSE and urinary vanillylmandelic acid/homovanillic acid ratio are helpful to predict prognosis. Amplification of hall mark oncogene N-myc, deletion of 1p or 11q, gain in chromosome 17, diploid, tetraploid status and dlk protein are associated with dismal prognosis.^[9]

Some reports suggested GNB in adults are slow growing, but almost half of the patients had locally advanced tumors or had remote metastases at diagnosis.^[7,10] With regard to prognosis, 10 out of 13 patients with distant metastases died within 1 year.^[10]

Management of this is by mainly complete surgical resection and post-operative chemotherapy. The present case is Prognostically unfavorable because of nodular subtype of GNB, late age at diagnosis, presence of regional lymph node and adrenal gland metastasis. Post-operative chemotherapy is

planned for our patient. Adjuvant radiotherapy and meticulous follow-up is needed in case of partial resection of tumor.

CONCLUSION

Retroperitoneal nodular ganglioneuroblastoma is an extremely uncommon tumor in adult. Pre-operative diagnosis by clinical, radiological and biopsy seems difficult because of an exhaustive list of tumor occurring in the retroperitoneum corresponding to the age. Surgical Pathologist should think of this tumor when dealing with adult retroperitoneal mass even though it is a rare entity.

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