

A RARE CASE OF ADRENAL GANGLIONEUROBLASTOMA WITH METASTASIZED LYMPH NODE IN A YOUNG ADULT

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ABSTRACT

Adrenal ganglioneuroblastoma is an extremely rare tumor among adrenal neoplasms in adult. We reported a 26-year-old young female patient presented with vague pain in left flank. Abdominal sonography and CTScan revealed a heterogenous solid lobulated left adrenal mass which containing central necrosis, hemorrhage, coarse calcification and multiple adjacent metastasized lymphoid nodules, hence adrenal cortical carcinoma was not excluded although testing for adrenal function were normal. The patient was performed left adrenal surgery following by chemical therapy. Histology was consistent with intermixed stroma-rich ganglioneuroblastoma. In literature, about 20 cases of adrenal ganglioneuroblastoma in adults were described, in which lymphoid metastasis was uncommon. Therefore, although adrenal ganglioneuroblastoma is obtained by histology, imaging features should be investigated further to differentiate from other neoplasms for appropriate treatment.

Keywords: *Ganglioneuroblastoma in adult, ganglioneuroblastoma, neuroblastoma.*

I. INTRODUCTION

Ganglioneuroblastoma (GNB) is usually a rare malignant neoplasm in pediatrics. In

adult, less than 50 cases of GBN was reported, in which just over 20 cases of GBN occur on adrenal gland. We report a case of adrenal GBN in 26-year-old female patient.

II. CASE STUDY

A 26-year-old female patient presented with vague pain in left flank for a long time. She had no specific medical history. Physical examination showed normal vital signs with 90/52mmHg of blood tension, vague pain when percussion in left kidney area. Blood examination showed normal range of cortisol level at 7.21ug/dl.

Sonography revealed a heterogeneous mass in left flank with prominent solid component, hyperechoic hemorrhage and several coarse calcified nodules. Abdominal CTScanner showed a lobulated poorly enhanced solid mass 48x62x68mm in left adrenal fissure with hemorrhagic and coarse calcified components, adjacent similar smaller solid masses 27x16mm which surround the left adrenal vein.

In the suspect of malignant lesion, left adrenal operation was performed to remove those masses. In gross histopathology, the main mass was jelly-like transparent, 7x7x4.5cm in size and 116g in weight, with smooth surface and central necrosis; microscopic histopathological finding confirmed a ganglioneuroblastoma, stroma rich, intermixed subtype, mitosis karyorrhexis index MKI < 100, unfavorable (26 year-old), invading to tumor capsule,

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surrounding adipose and vessels. The adjacent mass was lymph node in gross appearance while microscopic description revealed majority of Schwannian stroma rich

cells, ganglion cells and some neuroblast cells.

Thereafter chemical therapy was accomplished for 4 phases and the patient live healthy so far.

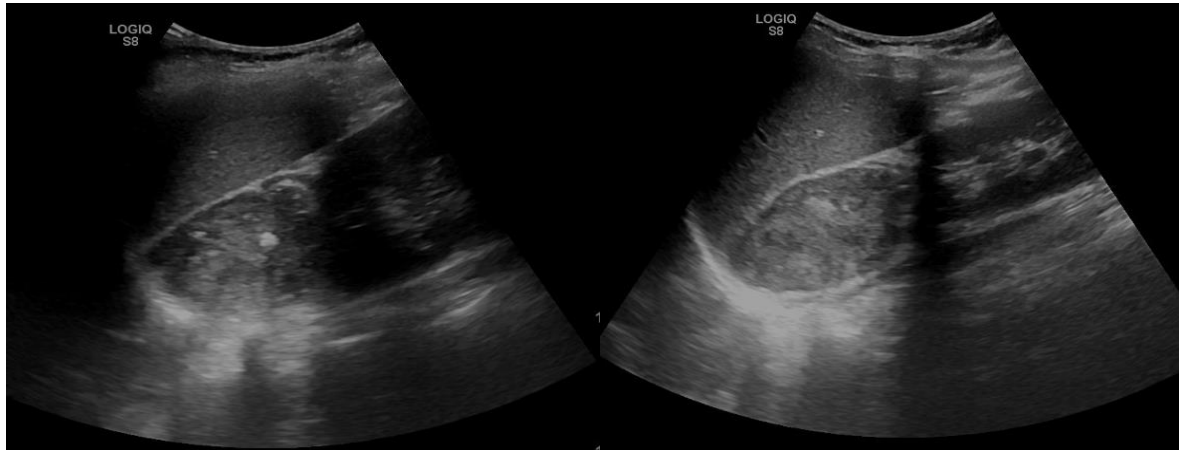


Figure 1: *Left flank ultrasound revealed a lobulated heterogeneous solid mass with several calcified nodules in left adrenal fossa.*

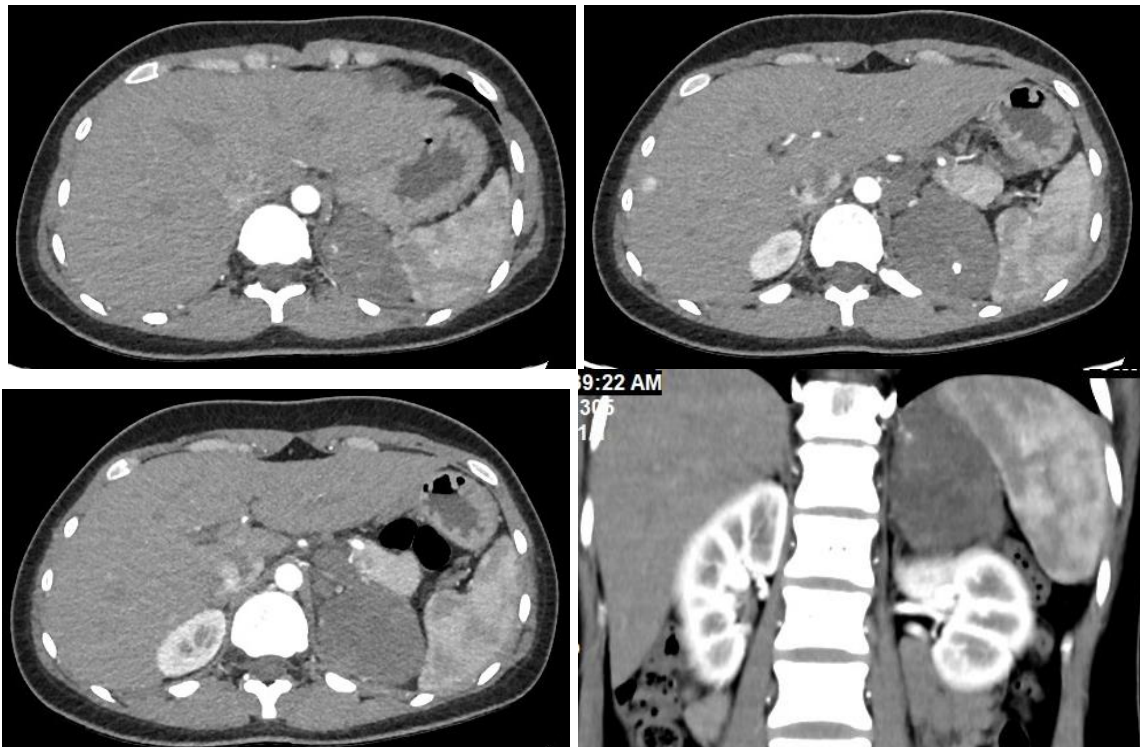


Figure 2: *Abdominal CTScan arterial phase showed a poorly enhanced mass in left adrenal fossa (A, D) with central coarse calcified nodule (B) and some similar smaller adjacent masses surrounding the anterior vessel (C).*



Figure 3: *Abdominal CTScan portal phase showed the poorly radial enhanced mass with smaller anterior nodules surrounding the left adrenal vein (D)*

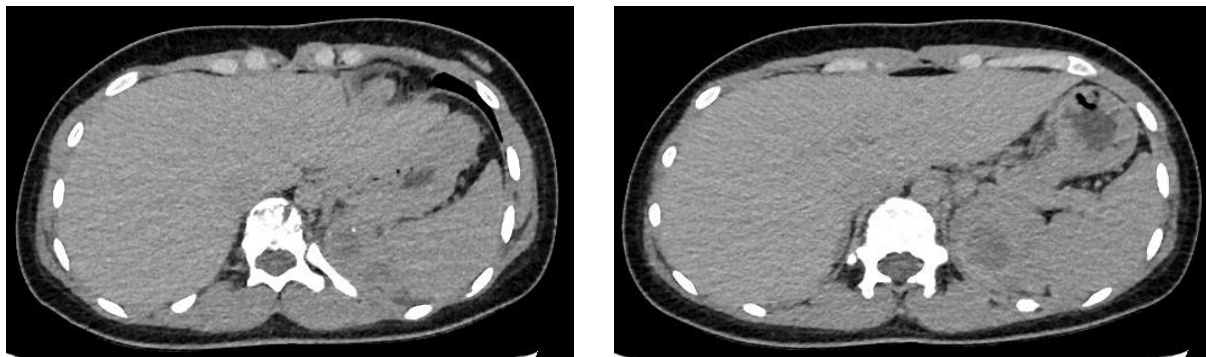


Figure 4: *Abdominal CTScan delay phase showed the necrotic portion in the upper part of this solid mass with another small punctate calcification (A).*

III. DISCUSSION

A ganglioneuroblastoma is an intermixed tumor among neuroectodermal origin neoplasm. GNB majorly appears in pediatrics and 80% of them arise within abdomen or adrenal gland [1]. Meanwhile, over 20 cases of adrenal ganglioneuroblastoma in adults

were described in literature so far [2], [3].

There is not significant dominance in gender and ethnic. Besides, despite of wide range, most of adult patient onset on the 2nd decade, our patient age was also in this decade of life [3]. Although about a third of

cases are incidentally finding, other patients complain about abdominal pain or discomfort, flank pain, epigastric pain or urinary symptoms and hypertension occasionally [2], [3]. Our patient has no hypertension but also complain about plank pain.

In pathophysiology, GNB mainly include neuroblasts and their derivatives such as ganglion cells and Schwann cells, and stroma [1]. They are actually postoperative diagnosis because GNB remains challenging to confirm preoperatively by radiological features due to their similar findings to other malignant tumors on images. Hence finding out more prominent imaging features of GNB might elevate confidence to preoperatively diagnose for radiologists.

US is usually the first imaging modality of choice, GNB is described as a heterogeneous mass with anechoic cystic or necrotic component, hyperechoic hemorrhagic component, focal or diffuse calcifications and hypoechoic solid component [1], [2], [4]. Our case performed a prominent solid component, hyperechoic hemorrhage and several coarse calcified nodules on ultrasound.

Nevertheless, CT and MRI are better imaging modalities to further evaluate the mass and staging the condition preoperatively. CT findings in GNB vary in a wide range from a cystic mass with few thin solid strands to a solid mass with few necrotic component [1] [2], [5]. They are usually poorly enhanced by contrast medium although they tend to surround major vessels without compression or occlusion. Calcification which is hallmark of neuroblastoma in children and rare in adult is typically fine and punctate in majority of case, and present in 42-60% [2], [6], however

there is no commented on calcification in over 35-year-old patient [3].

On MRI, GNB appears as an irregular contour mass with typically heterogeneous content which is relatively low signal intensity on T1-weighted images and high signal intensity on T2-weighted images, no difference in chemical shift and early enhanced by contrast medium [1], [5], [7].

Metastases were found in 36-42.5% of cases located in local lymph nodes, liver, bone/bone marrow, and para-aortic nodes [2], [3], [5], [6], in which lymph node metastases accounted for 19% [3]. Among nine out of twenty-one cases with metastasis, lymph nodes occurred in four out of the nine cases and five per nine patients with metastases were in 2nd decade [3].

On the other hand, adrenal cortical carcinoma (ACC) also has some similar findings as a solid mass with intratumoral necrosis or hemorrhage, invading into adjacent structures and vessels, calcification [8].

In order to differentiate those conditions, some typical criteria should be made. First, ACC usually presents as a solid mass while GNB presents in a range from cystic to solid mass. Secondly, besides similar intratumoral hemorrhage or necrosis, intracytoplasmic lipid is found in < 30% of ACC which could cause loss of signal on chemical shift sequence in MRI, meanwhile this feature is not reported in GNB. Thirdly, ACC shows vivid enhanced and slow washout, GNB deems to be poorly enhanced. Calcification locates centrally in form of micro-calcification to coarse in approximately 30% [8] cases of ACC, GNB otherwise is typically either punctate or fine [6].

There is no official guidelines for treatment in adult, the appropriate treatment was

derived from pediatric experience, including surgery, radiotherapy and chemotherapy and the patient is treated due to their personal staging [9], [10]. If the tumor is considered radically resectable, surgery represents the treatment of choice. Because of the high possibility of infiltration of regional lymph nodes, local lymph node dissection is recommended [6]. If the tumor is unresectable, a diagnostic biopsy should be performed and treatment options are radiotherapy or cytoreductive chemotherapy. Chemotherapy is the treatment of choice in metastatic disease [2]. Patients at low risk of metastasis or death receive minimal intervention and those at high risk receive multimodality treatment. New immunotherapeutic techniques and nuclear medicine-targeted therapies have emerged and are demonstrating promising response rates for patients at high risk [4].

In our case, the left flank mass exhibited on ultrasound and CT scanner as a solid poorly enhanced mass with intra component of hemorrhage and necrosis and some nodules of coarse calcification, the mass tends to invade the adjacent vessels; contemporarily, there are local metastases in several ipsilateral lymph nodes. Hence, those features seem to be appropriate to adrenal cortical carcinoma. The information to be confused in this patient are her young age (26-year-old) which is in the peak of GNB in adult instead of the late middle age as the common prevalence of ACC and the shortage of MRI features because of her personal reject. Therefore, without the confirm from pathophysiology, GNB was difficult to diagnose on image modalities in this patient. After undergoing operation, the patient absorbed 4 phases of chemical therapy and live healthily until now.

IV. CONCLUSION

Although neuroblastomas occur mainly in children which named Wilm's tumor, and similar findings to adrenal corticocarcinoma, peripheral ganglioneuroma in adults should not be ignored in differential diagnosis due to its risk of malignant potential. GNB remains challenging to confirm preoperatively by radiological features because of their similar findings to other malignant tumors on images, so far pathology conduce to the decisive diagnosis. Hence finding out more prominent imaging features of GNB could enhance confidence to preoperatively diagnose for radiologists.

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