รายงานผลภาพเอกซเรย์คอมพิวเตอร์ของมะเร็งต่อมหมวกไต ชนิดนิวโรบลาสโตมาในผู้ป่วยเด็กที่พบร่วมกับภาวะต่อมน้ำเหลืองโต ใจช่องท้องจำนวนมาก

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บทคัดย่อ

มะเร็งของต่อมหมวกไตชนิดนิวโรบลาสโตมาเป็นมะเร็งที่พบบ่อยเป็นอันดับสามของมะเร็งทุกชนิดในผู้ป่วยเด็ก และมีการเจริญพัฒนามาจากการระบบประสาทซิมพาเธติก พบได้บ่อยในกลุ่มช่วงอายุ 1 ถึง 5 ปี ทางคณะผู้เขียนได้รายงานลักษณะภาพทางรังสีของผู้ป่วยเด็กหญิงอายุ 2 ปี ที่มีอาการอึดอัดแน่นท้องและมีไข้ร่วมด้วย ตรวจด้านภายคลำพบก้อนในช่องท้องทางด้านขวา ภาพเอกซเรย์คอมพิวเตอร์พบก้อนที่ต่อมหมวกไตข้างขวาและมีหินปูนภายในก้อน นอกจากนี้ยังพบภาวะต่อมน้ำเหลืองโตในช่องท้องจำนวนมาก ซึ่งเป็นลักษณะที่พบได้บ่อยในนิวโรบลาสโตมา ผลการตรวจเลือดและปัสสาวะพบสารประกอบกลุ่มแคทเทอกอนอยู่ในระดับสูงกว่าปกติ และผลการตรวจทางพยาธิวิทยาของต่อมหมวกไตพบลักษณะที่เข้ากับโรคนิวโรบลาสโตมา คณะผู้เขียนได้สนับสนุนความเกี่ยวกับภาพทางรังสีที่มีบทบาทในการวินิจฉัยโรคนิวโรบลาสโตมา ค่าสำคัญ: มะเร็งต่อมหมวกไตในผู้ป่วยเด็ก นิวโรบลาสโตมา ภาวะต่อมน้ำเหลืองโตในช่องท้อง เอกซเรย์คอมพิวเตอร์
CT appearance of neuroblastoma with massive intra-abdominal and retroperitoneal lymphadenopathy: case report and review literatures

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Abstract

Neuroblastoma is the tumor of sympathetic nervous system that derives from primordial neural crest cells. Neuroblastoma is the third most common pediatric malignancy after leukemia and central nervous system tumors. This article reported atypical computed tomographic findings of neuroblastoma with massive intra-abdominal and retroperitoneal lymphadenopathy.

The authors reported a 2-year-old girl who presented with abdominal distension and fever. Ultrasonographic examination showed a large heterogeneous echogenic mass at right suprarenal region. Contrast enhanced computed tomography (CT) of the whole abdomen revealed an ill-defined inhomogeneous enhancing mass at right adrenal gland with internal calcification. There were massive intra-abdominal and retroperitoneal lymphadenopathy. Pathological diagnosis showed the poorly differentiated neuroblastic tumor.

This article reported atypical CT findings and reviews natural history, histological findings, biological features, and radiographic appearances.

Keyword: Neuroblastoma, lymphadenopathy, contrast enhanced CT scan

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**Introduction**

Neuroblastoma is a tumor of ganglion cell origin that derives from primordial neural crest cells which are the precursors of the sympathetic nervous system.\textsuperscript{1,2} Neuroblastoma has varied widely in spectrum of clinical presentation which depends on the site, extension of the primary tumor and presence of distant metastatic disease. The ultrasonography, computed tomography, and magnetic resonance imaging can provide determination of organ of origin, tumor extension and distant metastasis.\textsuperscript{3,4}

**Case report**

A 2-year-old girl was referred to our hospital with abdominal distension for a month and fever for a week. There was no associated with nausea, vomiting or diarrhea. The physical examination revealed a palpable large abdominal mass with tenderness at right upper quadrant region.

The ultrasonographic examination showed a 7x8-cm heterogeneous echoic mass occupying in right suprarenal fossa which could not determine whether originating from liver or right adrenal gland. Further contrast enhanced CT scan of the chest and whole abdomen were performed. There was an ill-defined inhomogeneous enhancing mass at right adrenal gland with coarse calcification inside (Figure 1a). There was direct invasion to inferior aspect of right hepatic lobe and upper pole of right kidney (Figure 1 b).

![Figure 1(a)](image1a.png)  ![Figure 1(b)](image1b.png)

Figure 1(a) Noncontrast axial CT scan of the abdomen demonstrates an ill-defined mass at right adrenal gland with coarse internal calcification (arrow). (b) Contrast enhanced axial CT scan shows heterogeneous enhancement with involvement of right hepatic lobe (arrow).
Demonstrable massive intra-abdominal and retroperitoneal lymphadenopathy along para-aortic, aortocaval, retrocrural and peripancreatic regions caused encasement of the intra-abdominal vessels (Figure 2a, b).

![Contrast enhanced axial CT scan reveals massive intra-abdominal and retroperitoneal lymphadenopathy with encasement of splenic vein (curved arrows), both renal arteries (straight arrows) and abdominal aorta.](image)

There were few 1.5-cm left supraclavicular lymph nodes. There was no extension to spinal canal. The elevated level of urine VMA (vanillylmandelic acid = 35.6 mg/24 hrs) and serum NSE (neuron specific enolase = 195.2 ng/ml) were detected.

The patient underwent core needle biopsy at right adrenal gland and left supraclavicular lymph node. The pathologic finding with immunohistochemistry of right adrenal mass showed the poorly-differentiated neuroblastic tumor, unfavorable histology group. The pathologic report of left supraclavicular lymph node revealed malignant round cell tumor which was suggestive of dissemination to distant lymph node. Our patient was defined as stage IV. There was no evidence of tumor cells in the bone marrow sample. The rarity of massive intra-abdominal and retroperitoneal lymphadenopathy in neuroblastoma makes it worth to be reported.

**Review literature and discussion**

Neuroblastoma is the second most common abdominal neoplasm in children following Wilms tumor accounting for 5-15% of all malignant tumors in children. The median age at diagnosis is 22 months. More than 90%
of cases are children age less than 5 years, with peak incidence at age of 2-3 years. Neuroblastoma is the tumor of sympathetic nervous system that may arises from anywhere sympathetic tissue naturally occurs. The common primary sites are the adrenal medulla (35%), extra-adrenal retroperitoneum such as the organ of Zuckerkandl (30-35%), and following by posterior mediastinum (20%). Less common sites are the neck (1-5%) and pelvis (2-3%).

Clinical presentation of neuroblastoma depends on the location of the primary lesion or metastatic spreading. A palpable abdominal mass is presented in 54% of patients. Large abdominal neuroblastoma may compress kidney, causing renin-associated hypertension. Pain and fever occur in one-third of patients. Cervical or apical thoracic masses result in Horner’s syndrome. Up to 60% of patients have metastasis at presentation which the common sites are the bony skeleton, regional lymph nodes and liver but spread to the lung or brain is rare.

Neuroblastoma contributes varied tumoral biologic markers which are used to determine maturity, staging and prognosis. Neuroblastoma is predilection to secrete cathecholamines (90-95%), most of which are vanillylmandelic acid (VMA) and homovanillic acid (HVA). HVA may be elevated in more maturity. On the contrary, VMA is less mature metabolite of cathecholamines. Vasoactive intestinal peptide (VIP) is produced by ganglion cells within tumor and causes watery diarrhea. Presenting of VIP shows a tendency to be more mature and have a better prognosis. As oppose to high serum levels of lactate dehydrogenase (LDH), neuron specific enolase (NSE), and ferritin are indicators of unfavorable prognosis.

Histological features consist of neuroblasts which are immature, undifferentiated small, round-shaped sympathetic cells. The secondary features are necrosis, mitosis, hemorrhage and calcification.

Neuroblastoma shows variable appearances and growth pattern on radiographic features. Ultrasonographic examination is the initial imaging modality to evaluate a child with abdominal mass which provides the excellent screening method and a baseline for follow up. By ultrasonography, neuroblastoma is usually heterogeneous solid lesion. Cystic anechoic areas usually represent hemorrhage or necrosis within the tumor. Calcification is common which has been shown as focal hyperechoic area with or without posterior acoustic shadow. Ultrasonography may depict adjacent organ involvement or distant metastasis. Computed tomography (CT) is the most common modality for assessment of tumor extension, organ of origin, regional
lymphadenopathy, vascular encasement and distant metastasis.\textsuperscript{1-5} CT of the chest and whole abdomen are standard for the diagnosis and staging. CT has accuracy of about 80\% in tumor staging, when complemented with scintigraphy or bone marrow aspiration, the accuracy has been increase to 97\%.\textsuperscript{9} Neuroblastoma usually shows heterogeneous density and 80-90\% of tumors demonstrate calcification in CT scan. The low attenuation areas within the tumor represent necrosis or hemorrhage. Direct invasion to liver, as in our finding, can be found only 5\%.\textsuperscript{10} Retroperitoneal lymphadenopathy or contiguous extension of primary tumor in retroperitoneal space occur frequently in neuroblastoma (73\%) which are usually found at renal hilum, retrocrural and para-aortic regions.\textsuperscript{1,5,6,10} Retrocrural lymphadenopathy proved to be specific for neuroblastoma (27\%) which is also seen in our patient.\textsuperscript{10}

Massive lymphadenopathy is much more likely to be found in lymphoma than neuroblastoma. Lymphoma occasionally involves adrenal glands with non-Hodgkin lymphoma being more common and Hodgkin disease.\textsuperscript{11} Lymphoma of adrenal gland can present in two forms: primary and secondary disease. Primary lymphoma is very rare which usually presents with bilateral adrenal masses without nodal involvement.\textsuperscript{12} Secondary disease is much more common which has been reported to occur in up to 25\% of all non-Hodgkin lymphoma.\textsuperscript{12} The common findings are unilateral or bilateral adrenal masses associated with retroperitoneal lymphadenopathy or other extranodal lesions.\textsuperscript{13,14}

Magnetic resonance imaging (MRI), neuroblastoma usually demonstrates heterogeneous low signal intensity on T1-weighted images, high signal intensity on T2-weighted images and enhancement after contrast administration.\textsuperscript{15} MRI is superior to CT in determining intraspinal tumor extension, relationships with blood vessels and ability to evaluate marrow involvement.\textsuperscript{2,6} The advantages of MRI are enhanced by the lack of ionizing radiation and does not require oral contrast.\textsuperscript{2,6,15} MRI has yielded high sensitivity (85-100\%) in detection of abdominal disease and distant metastasis.\textsuperscript{9}

Scintigraphic studies in neuroblastoma are performed in two aims which are identification of the primary tumor and evaluation of the bony metastasis.\textsuperscript{2} The primary tumor is depicted with a catecholamine analog (\textsuperscript{131}I- or \textsuperscript{123}I-metatolodobenzylguanidine - MIBG).\textsuperscript{131}I-MIBG scintigraphy is highly sensitive (88\%) and specific (99\%) for sympathetic tissues. The advantages of\textsuperscript{131}I-MIBG include tumor detection, evaluation of lymph node involvement, distant metastasis and monitoring of tumor after treatment.\textsuperscript{6,16} For evaluation of bony metastasis is performed
by using $^{99m}$Tc-methylene diphosphonates (Tc-MDP) bone scan scintigraphy which is much more sensitive than the plain radiographs. The drawbacks of $^{99m}$Tc-MDP are false-positivity in previous trauma and persistent in abnormal uptake for months despite of the eradication of malignant disease.\(^6\)

Current staging of the patients with neuroblastoma consists of CT or MRI for the primary tumors, bone scintigraphy for bony metastasis and bone marrow aspiration, biopsy for marrow disease.\(^2,5,6,9,15,16\)

**Conclusion**

Neuroblastoma is the tumor of neural crest in origin and most cases arise in adrenal gland. Regional lymphadenopathy are usually detected at renal hilum and porta hepatis. Massive intra-abdominal and retroperitoneal lymphadenopathy as our patient are unusual findings, however can be found. The clinical presentation depends on the site of primary tumor and location of metastasis. There are many biologic prognostic factors which are indication of the maturity of tumor and prognosis. Neuroblastoma requires multimodality of imaging which are helpful for specific diagnosis and staging.

**References**


