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Case Report

The diagnostic challenge of a rare malignancy called neuroblastoma: a clinical case report

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ABSTRACT

Neuroblastoma is a paediatric tumor that originates from neuroblasts. An interesting case report from Grodno of a child with mixed etiology showed some findings for neuroblastoma and some for Ormond's disease because the patient also has retroperitoneal fibrosis. Although both could be connected or can be separate diseases. Disease started with congenital anomaly of grade 2-3 left hydronephrosis and space occupying lesion in the retroperitoneal space (neuroblastoma). Then additional soft tissue tumor-like mass found near abdominal aorta and in the retroperitoneal space (Ormond's disease). This could be neuroblastoma of retroperitoneal space. Although neuroblastoma can be a separate disease or it could be in association with another disease, this article discusses the abnormal clinical presentation of neuroblastoma.

Keywords: Neuroblastoma, Retroperitoneal fibrosis, Left hydronephrosis grade 2-3

INTRODUCTION

Neuroblastoma is a type of rare pediatric tumor that occurs in 1 of 8000 live births with the median age of diagnosis of 17-18 months and only 5% of patients older than 10 years develop neuroblastoma.[1] Neuroblastoma is an extracranial solid tumor that can arise and can arise anywhere along the sympathetic nervous system.

Neuroblastoma is a pediatric malignancy that affects neural crest cells called neuroblasts. It is a tumor of the peripheral nervous system. In most common ways it affects adrenal medulla and sympathetic ganglia but sometimes it can affect kidneys, it can wrap around kidneys or can affect abdomen as well.[2,3] Clinical presentation of neuroblastoma is highly dependent on location and it can vary by age. Neuroblastoma can secrete catecholamines and we can identify their metabolites in urine and plasma.[4,5] Neuroblastoma is a paediatric form of cancer that affects children. Treatment and diagnosis of neuroblastoma is still challenging because of its clinical and biological heterogeneity. Neuroblastoma is a complex and from the genetic point of view there can be numerous chromosomal abnormalities.[2-6] It is a most common extracranial solid tumor for pediatric patients.[3] Disease progression and treatment varies widely among patients. Patients with MYCN gene amplification show worse prognosis and it manifests as very high-risk neuroblastoma.[1]

CASE REPORT

A 12-year-old boy on his first visit came to the grodno hospital for treatment of left hydronephrosis grade 2-3. The patient had congenital malformation of the left kidney. The surgeons did abdominal reconstructive surgery for correcting congenital malformation of the left kidney and elimination of obstruction of left ureter. While doing surgery surgeons noted intersections of embryonic adhesion around the left ureter. After that the surgeon did

drainage of the left retroperitoneal space. Primary diagnosis noted as left hydronephrosis grade 2-3 and space occupying lesion in the retroperitoneal space (neuroblastoma). There was a tumor-like mass found in the peritoneum and left retroperitoneal space. For surgical therapy surgeons did stenting of the left ureter, corrected kidney malformation (reconstructive surgery) and removed the tumor-like mass in the peritoneum and left retroperitoneal space. Draining of retroperitoneal space. After some months the ureter stent was removed. After therapy and all surgical management after a year the patient came for follow-up examination and in examination on the anterior and lateral wall of the abdominal aorta additional soft tissue mass were noted. Additional tissue was in paraaortic and along in the retroperitoneal cavity. With a biopsy it showed fibrosis like. So, diagnosis of Ormond's disease (retroperitoneal fibrosis) was also made.

Initial instrumental studies

These studies were done when the patient first visited the hospital. ECG was performed after stability from operation and there was sinus tachycardia, heart rate was around 97 BPM with normal position of EOS and alpha angle +62 with increased electric activity of heart. PQ interval 0.157 and QRS complex was 0.079, about QT interval it was 0.289. Dimension of right kidney 96x39 mm and left kidney 100x43 mm. In the colour doppler imaging mode the vascular pattern on the subcapsular region and angioarchitecture was unchanged. There was dilation of left renal pelvis with normal position and right kidney was unchanged (normal). On the left side dilation of renal pelvis, ureter and also of kidney calyces. Conclusion was left side calico pyelo ureter ectasis.

MRI report

It showed diffuse infiltration of tissues on the anterior abdominal wall post prior to surgery. Moderate infiltration of tissues of the mesentery and fatty tissue on the left and paranephric tissue and accumulation of free fluid under the wall of anterior abdomen up to 2 mm thick. Around subdiaphragmatic and near intestine loop also accumulation of fatty tissues were noted. Along the abdominal aorta (retroperitoneal) additional tissue component was found spreading along anterior and lateral walls of the vessel measuring 26x8 mm, 72 mm long up to aortic bifurcation with reduced intensity on T2. The liver was unchanged. With no structural and function change. Normal cystic and hepatic duct and as well as normal gallbladder structures. The main thing about adrenal gland is also presented without any structural changes.

Conclusion of MRI studies

Left renal pelvis and ureter was dilated, right kidney remains unchanged. Moreover, only fatty infiltrative changes along mesenteric and additional tissue components around the aorta are retroperitoneally found. Willms tumour or any other kidney related pathology is already excluded.

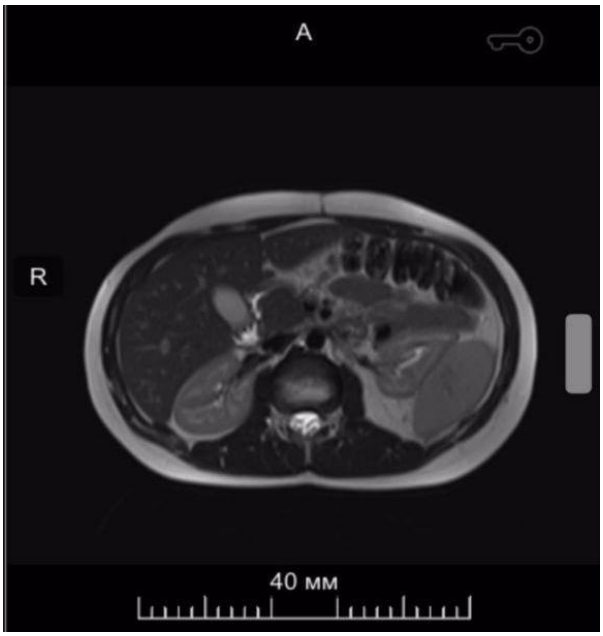

Figure 1: Axial view of MRI of abdomen.

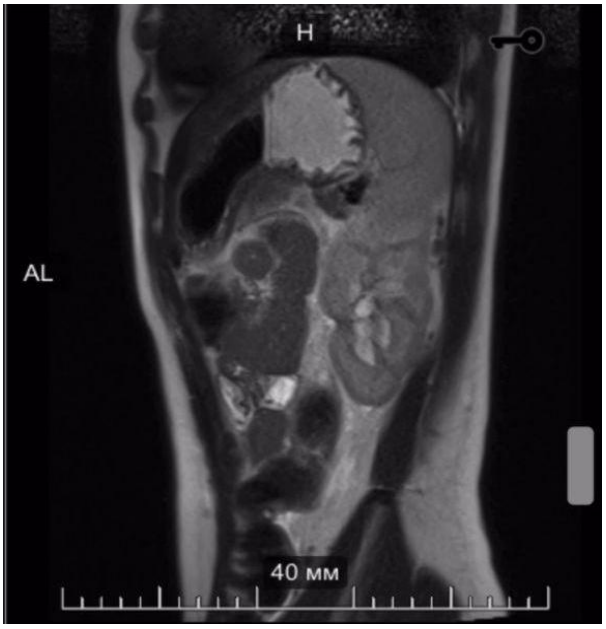

Figure 2: Anterolateral view of MRI of abdomen.

With the use of computed tomography Cyst of right sided kidney and left hydronephrosis grade 2 noted. The kidneys were equal in size and in the lower pole of the right kidney found avascular cyst up to 8 mm. The pelvis of the left kidney is 19x17mm and left calyx is 10 mm. The left ureter is in the upper third at a distance of about 25 mm from the ureteropelvic region to 10 mm where it makes a band and there is the ureteric cyst. The liver was clear and smooth; the structure of parenchyma is homogenous without any foci of pathology. The pancreas was not enlarged. The

pancreatic tissue was unchanged and without any obstruction or dilation. The adrenal gland was U shaped.

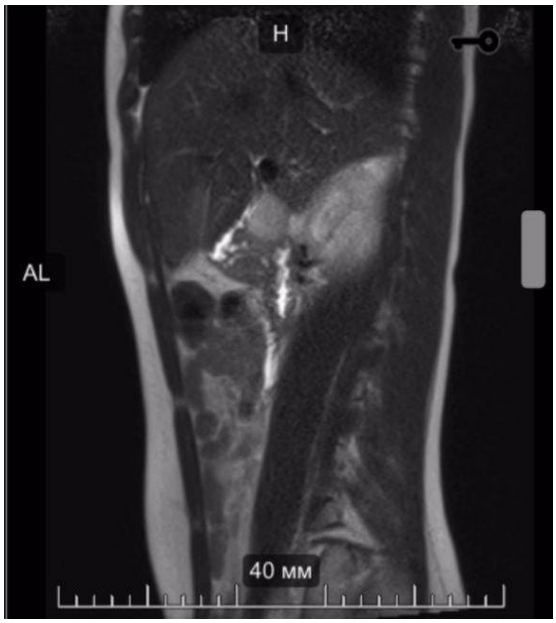

Figure 3: Anterolateral view of MRI of abdomen.

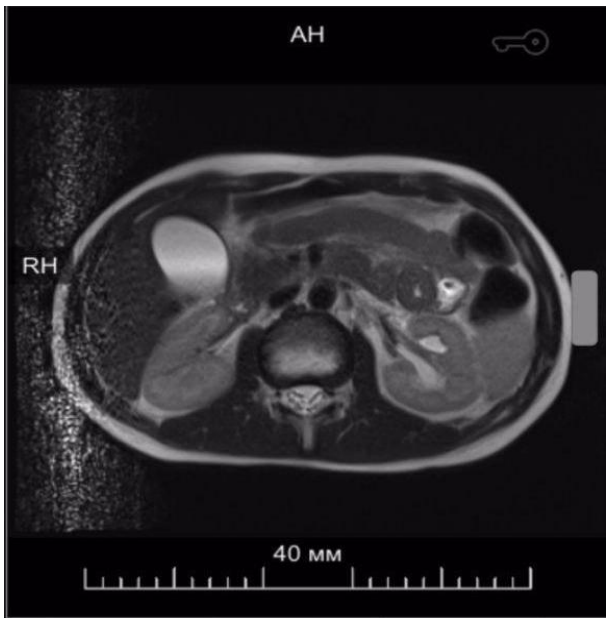

Figure 4: Axial view of MRI of abdomen.

Examination of lungs through X-ray was done at the intensive care unit. That showed no abnormalities. No mass, not any dilation and without any infiltrative changes. The domes of the diaphragm were clear. A crescent shaped air layer (SAL) was detected only in the right suprahepatic space.

Initial laboratory studies

These laboratory studies were done at the presentation of a patient when he first showed up in the hospital. The patient has blood type (A) and he is a rhesus factor positive. There was not any anti-erythrocyte antibody in the blood. Urinalysis showed normal yellow colour with acidic urine (pH:5.0) and urine density was 1015. No protein, glucose or ketone in the urine. 1-2 erythrocytes and 3-4 leukocytes were present. The coagulation panel determined activated partial thromboplastin time 28.2 sec and APTT were 0.97, prothrombin time was 12.1 sec. Prothrombin complex activity 87.1%, international normalized ratio 1:1 and fibrinogen was 3.67 g/l. Blood biochemistry showed total protein 76 g/l, urea 3.8 mmol/l. Normal creatinine level and glucose level. Total bilirubin and liver function tests AST and ALT were also in the normal ranges. Electrolytes levels are sodium 141 mmol/l, potassium 4.97 mmol/l and chlorine 109 mmol/l. Other investigation and analysis like liver function and urine analysis, coagulation panel and blood biochemistry as well as electrolytes all were normal. Gene analysis like for MYCN gene and for any other were not done. The patient was advised to have a regular visit for ultrasound control for every 3 months and for treatment and after some months he was referred to have stent removal of left ureter.

DISCUSSION

Paediatric tumours like neuroblastoma should be diagnosed early and treated early to increase chances of good prognosis. Neuroblastoma mainly occurs in children less than 5 years old and affects mainly adrenal medulla and sympathetic ganglia.[6] But it's not always true. It can also occur in the abdomen, chest, pelvis and even in the neck. The heterogeneity in clinical presentation and clinical course is challenging for both diagnosis and treatment.[7]

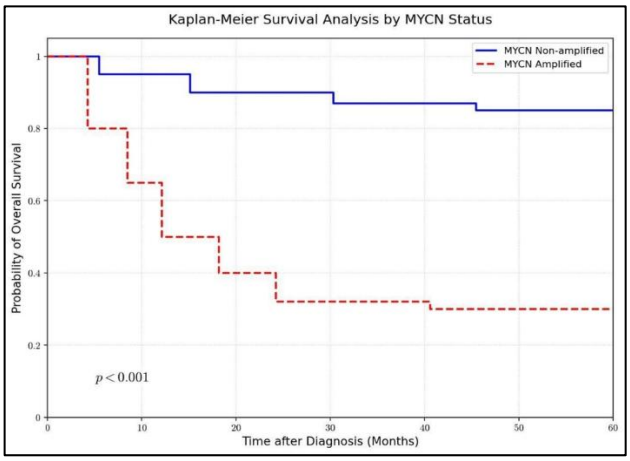

Figure 5: MYCN status on neuroblastoma.

MYCN gene is the main gene that promotes development of pluripotent stem cells from ectoderm in embryonal period then it further develops into neural crest cells and MYCN gene also plays a major role in differentiation of those cells. SO, overexpression of the MYCN gene leads to development of high-risk tumors like neuroblastoma.[1] For diagnosis of neuroblastoma sometimes biochemical

tests can play an impactful role. Neuroblastoma is a tumor of neural crest cells (neuroblast) and this type of tumor cells often secretes catecholamines. These catecholamines metabolites can be detected in plasma serum or urine. Studies showed that neuroblastoma displays a relative lack of the catecholamine storage vesicle characteristic of mature chromaffin cells and also their PPGL derivatives. Because of these reasons neuroblastoma does not cause hypertension but metabolites of catecholamines like homovanilic acid and vanillylmandelic acid can be detected in urine or plasma.[5] These tests along with instrumental studies like CT(computed tomography) and MRI (magnetic resonance imaging) are also impactful like laboratory analysis.[8] So with a combination of diagnostic tests we can detect severe tumors like neuroblastoma and can start treatment early.

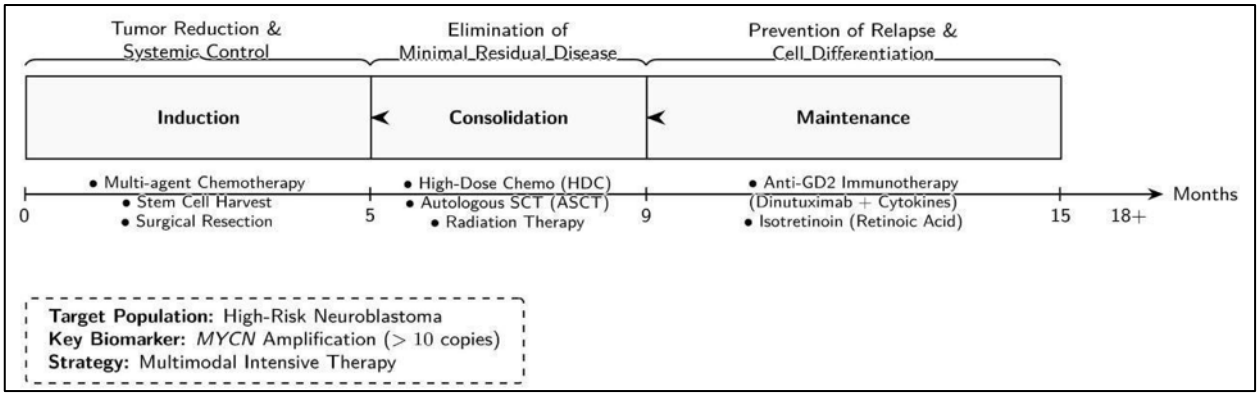

Figure 6: Risk-based treatment of neuroblastoma.

Treatment of neuroblastoma is risk-based and MYCN amplified neuroblastoma can be treated with MYCN inhibitors.[9]

For mild-risk neuroblastoma or localized lesions of neuroblastoma can be treated with excision of tumor and chemotherapy after, the most common surgical approach is posterolateral thoracotomy for thoracic neuroblastoma.[10]

CONCLUSION

Neuroblastoma is very complicated and challengeful for diagnosis as well as for treatment. In this article, a 12-year-old boy, it is relatively very rare and uncommon. The main goal of the article is to acknowledge physicians about this rare type of malignancy and it is recommended to go MYCN gene testing to know prognosis status.

Surgical intervention played a central role in local disease control and histologic diagnosis. Complete gross resection, when safely achievable without unacceptable morbidity, contributed to improved local control in this patient. This case also emphasizes careful preoperative planning (detailed imaging and multidisciplinary coordination) and intraoperative decision-making to balance extent of resection with preservation of organ function.

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