

Patient with neuroblastoma complicated by giant thrombus of an inferior vena cava with intracardiac extension

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ТӨМЕНГІ ҚАҚПА ВЕНАСЫНАН ОҢ ЖАҚ ЖҮРЕКШЕГЕ ДЕЙІН СОЗЫЛҒАН ГИГАНТТЫ ТРОМБПЕН АСҚЫНҒАН НЕЙРОБЛАСТОМАСЫ БАР НАУҚАС

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ПАЦИЕНТ С НЕЙРОБЛАСТОМОЙ ОСЛОЖНЕННОЙ ГИГАНТСКИМ ТРОМБОПРОТЯЖЕННОСТЬЮ ОТ НИЖНЕЙ ПОЛОЙ ВЕНЫ ДО ПРАВОГО ПРЕДСЕРДИЯ

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Introduction

Neuroblastoma is the most common extracranial solid tumor in childhood. Before the age of one year neuroblastoma occurs in 50% of diagnosed neoplasms, moreover by age 5 it occurs in 90%. This malignancy was described by James Wright in 1910, and was named because cells were associated with fibrils in arrangements similar to neuroblasts. Neuroblastoma arise in tissues of the sympathetic nervous system and can be in different part of body. In all cases neuroblastoma is diagnosed by imaging (MRI), also in 90% patients have catecholamine acids in urine and high level of Neuron-specific enolase. Case with we had been presented is a tremendous clinical challenge.

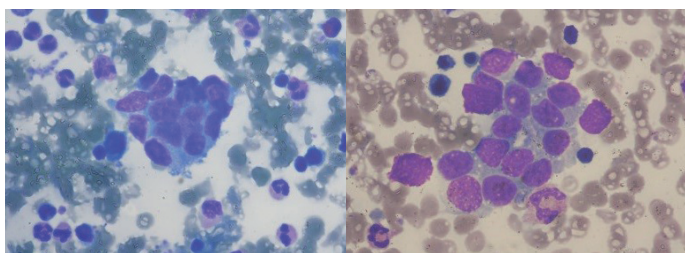
Description of clinical case

3 years old girl had admitted to National research center of mother and child health

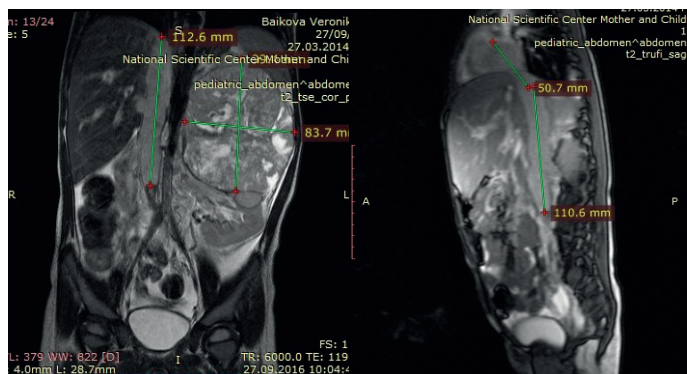
Astana, Kazakhstan in September 2016 with fever, weight loss, diffuse abdominal pain, loss of appetite. During physical examination revealed palpable and motionless mass in the upper left side of abdomen. On ultrasonography investigation solid tumor of left retroperitoneal space had been detected, size was 119x95x112 mm. An abdominal MRI had shown the formation an irregularly-oval shaped solid structure with precise uneven contours about 109x86x93 mm, in the projection of the left adrenal gland, compressing left kidney but without invasion on it. Also there was revealed thrombus growing from inferior vena cava and filling the cavity of right atrium, with size 5 cm

in width and 13 cm in length, prolapse. Laboratory findings: NSE -139.40 ng/ml (normal value: 1-15 ng/ml), AFP 3,12 MU/ml (normal value: 1-5 MU/ml), BMA 29,13 mg (normal value: 1-5 mg). Myelogram: accumulations of small blue round tumor cells, typical for neuroblastoma. With the aim of decreasing of sizes of tumor 3 cycles non-adjuvant chemotherapy N5/N6 according to NB2004 was provided. After these courses of chemotherapy tumor significantly decreased in sizes, it becomes 46x37 mm in diameter. Giant thrombus of IVC and RA was also decreased in sizes 10x11 mm (previous sizes 3,35x5,07x3,67). IVC is dilated till 5,5 x 5,9 mm (previous sizes 1,6 cm). In January 2017 radical operation of tumor resection was made. Resection margins were clear. Histology: Morphological picture corresponded to a poorly differentiated neuroblastoma with pathomorphosis 3rd degree. Child was carried out more 3 cycles of adjuvant chemotherapy. During all period of treatment patient took additional anticoagulant therapy by low doses of Heparin and Warfarin. The EchoCG after 6 courses of chemotherapy (N5/N6) had shown absence of IVC thrombosis, and thrombus in the cavity of RA significantly decreased in sizes 8,0x6,3 mm. Further treatment of this child contained high dose chemotherapy with autologous stem cell transplantation according to NB 2004 protocol and external beam radiotherapy on the tumor bed (30Gy). Now patient is in clinical and radiological remission.

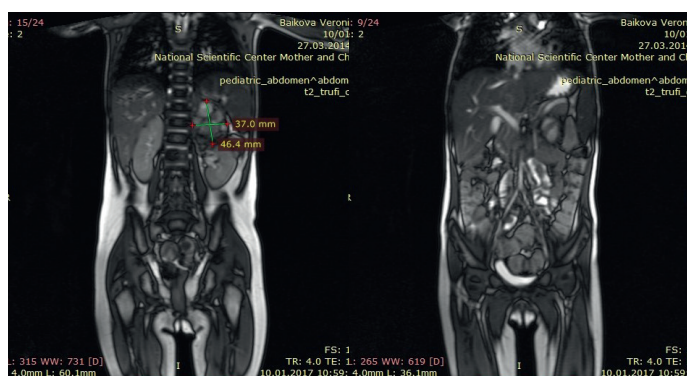
- Histology findings: Homer Wright rosettes are a type of rosette in which differentiated tumor cells surround the neuropil



- MRI at the moment of diagnosis

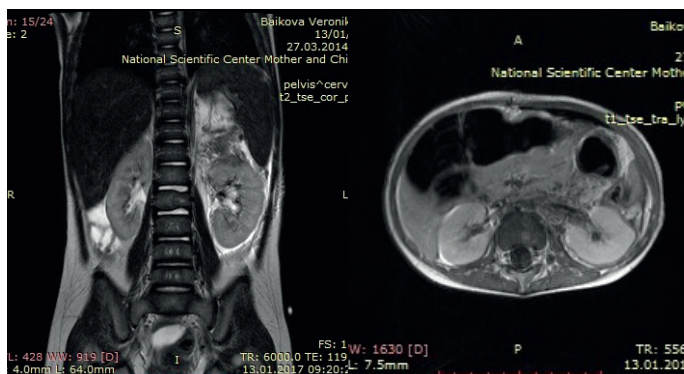
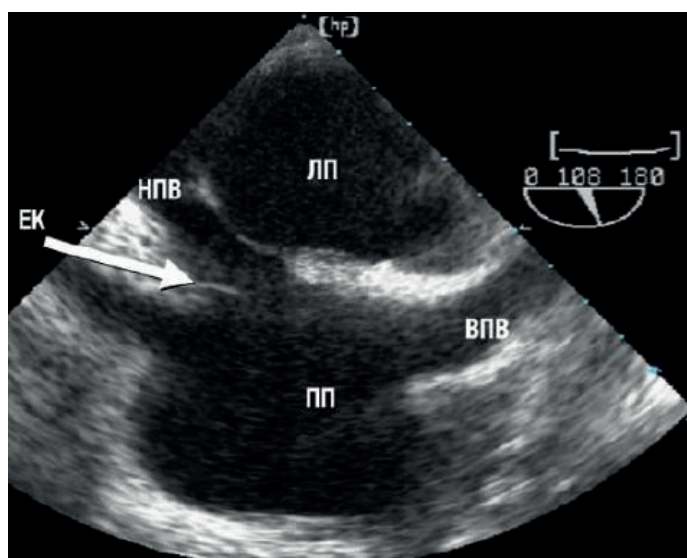


- MRI after neo-adjuvant chemotherapy, before operation



- EchoCG at the moment of diagnosis

In the region of right atrium there is tissue mass (thrombus), sizes 3,35x5,07x3,67, arising from IVC, have prolapse in the cavity of left atrium. IVC is dilated till 1,6 cm, filled by thrombus.

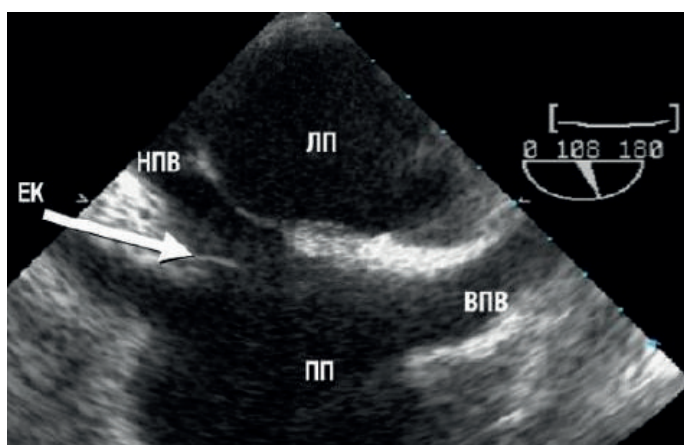


- MRI after operation(condition after total operation)

EchoCGafter 6 courses of N5/N6

- IVC is normal, thrombus is absent

• In the cavity of right atrium there is tissue mass (thrombus), sizes 8,0x6,3mm (previous sizes 3,35x5,07x3,67). IVC is normal, thrombus is absent (previous sizes 1,6 cm).



Discussion

Wilms' tumor presenting with extension into the inferior vena cava and right atrium is thus rare and renders the affected child with additional cardiovascular complications and operative risks. As a result of the uncommon occurrence, a consensus on management based on prospective study would be difficult. The present report is supportive of the use of preoperative chemotherapy in the initial management of advanced Wilms' tumor extending into the right atrium[2]. In literature survey there was found the review of the charts of 155 children with WT treated between 1983 and 2005, and analysis of 16/155 (10.3%) children with WT who presented cavaatrial tumor extension, being 8/16 IVC and 8/16 atrial thrombus. Preoperative chemotherapy was able to reduce thrombus extension in 8/11 (73%) treated patients and cardiopulmonary bypass (CBP) was avoided in 2 patients with atrial thrombus. Surgical resection of tumor and thrombus was successful in all cases, receiving or not preoperative chemotherapy and overall survival was similar in both groups[3]. Therefore two main strategies of treatment of these nephroblastoma patients present nowadays: active surgical resection with CBP or preoperative chemotherapy, which reduce sizes of thrombus significantly, and it gives opportunity to cure patients without aggressive surgery.

Conclusion

1. Neuroblastoma is one of the frequent solid tumors in pediatric oncology, but extremely rare it can be complicated by thrombosis of inferior vena cava with intracardiac extension. This process probably is due to the disorders in rheologic properties of blood because of huge number of tumor cells circulating in the bloodstream, and we regarded it as proto-oncogenic thrombus.

2. Differential diagnosis between nephroblastoma and neuroblastoma in preoperative period should be based not only

on the radiology findings, but also on the results of specific neuroblastoma markers.

3. Neuroblastoma in high risk group patients is very sensitive to chemotherapy, therefore the proto-oncogenic thrombus after treatment by intensive neo-adjuvant chemotherapy in combination with anticoagulant therapy can dissolve thrombosis without aggressive surgical intervention.

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