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Unexpected Tumor Rupture in a Case of Treatment-naïve Neuroblastoma - A Mortality Experience from a Tertiary Medical Institution

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Abstract

Neuroblastoma (NBL) is one of the most common extracranial neoplasms in children. Mortality is often attributed to treatment-related adverse events, sepsis secondary to immunocompromised status, and multi-organ failure resulting from advanced illness. Cases of NBL initially presenting with life-threatening events are rare. Here, we present a fatal NBL case that initially manifested profound anemia and coagulopathy, which later progressed to hemorrhagic shock due to tumor rupture.

Keywords: Mortality, neuroblastoma, oncology, pediatrics

Introduction

Neuroblastoma (NBL) is a major contributor to extracranial neoplasms in children. Mortality is linked to treatment-related adverse events, serious bacterial infection secondary to immunocompromised status, and multi-organ failure resulting from advanced illness. Rarely do patients succumb to extensive disease before the initiation of treatment or during the disease diagnostic stage. We present a mortality case of treatment-naïve NBL to emphasize the urgency of a precise diagnosis.

Case Reports

A 1-year and 7-month-old boy was admitted to the hospital because of a pale appearance. One month prior to hospitalization, the patient appeared pale and was brought to the outpatient department of pediatric hematology for a survey. In addition to the pale appearance, a distended abdomen was reported. Laboratory evaluations revealed profound anemia and coagulopathy, prompting admission for detailed examinations.

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During hospitalization, computed tomography (CT) revealed a right heterogeneous mass (size: 10.9*10.3*8.0 cm) located in the retroperitoneal region (**Figure 1**). To differentiate from the tumor nature, 24-h urine vanillylmandelic acid (VMA) and catecholamine were collected. Bone marrow (BM) aspiration was performed and was negative for malignancy. However, on the second day of hospitalization, immediately after completing urine sampling, an event of hypotension with a disturbed conscious status occurred. The prompt survey revealed evidence of anemia and coagulopathy progression, thereby diagnosing uncontrolled tumor bleeding. Life-saving management, including fluid resuscitation, blood component transfusion, and intravenous coagulation factor 7a infusion, was performed. Because of his critical condition, he was referred to the pediatric intensive care unit for intensive treatment.

Upon stabilization of vital signs, an emergent CT scan revealed profound hemoperitoneum (**Figure 2**). A salvage operation was performed for bleeding control and tumor resection. Under laparotomy, a huge tumor encapsulated the kidney, adrenal gland, and inferior vena cava. Dissection was performed cautiously. However, refractory hypotension and massive bleeding persisted despite vasopressor use and transfusion of blood components. Cardiopulmonary resuscitation (CPCR) was initiated when cardiac arrest occurred. Unfortunately, his vital signs were unresponsive to high-quality CPCR. His family decided on palliative care after an explanation of his bleak prognosis. The patient succumbed to the illness on the third day of hospitalization. Further histology of the resected tumor and urine catecholamine report confirmed the diagnosis of NBL.

Discussion

NBL is one of the most common extracranial neoplasms in children. Embryologically, NBL originates from primitive ganglion cells, which transform into the sympathetic nervous system and adrenal glands. Based on embryological origination and development, NBL can be recognized in areas of the abdomen, paraspinal sympathetic chains, adrenal glands, and rarely the renal parenchyma. Misdiagnosis of Wilms tumor (WT) as NBL may occur. Both NBL and WT can initially present as abdominal distention or mass; thus, laboratory and/or radiological evaluations are warranted. An accurate preoperative diagnosis is crucial because surgical timing and planning differ significantly.

In our case, a precise distinction between NBL and WT was challenging because of similarities in clinical and radiologic presentation. Previous studies have revealed that the presence of constitutional symptoms and intramural calcification are more likely to be associated with a diagnosis of NBL.[1,2] However, these characteristics were not observed in our case. Urinary VMA analysis possessed diagnostic value in differentiating NBL from WT. Unfortunately, we failed to disclose the result of the urinary VMA test before the progression of his disease. Serum neuron-specific enolase (NES) can be used as a rapid resulting marker. However, the sensitivity of NES is debated, and such testing is not available at our institution. BM involvement was reported to be more prevalent in cases of NBL than in WT. Nevertheless, BM sampling in our case was negative for malignancy. After a cautious and retrospective review of the CT scan at presentation, a small and hyperdense lesion could barely be traced. However, the challenge of distinguishing between calcification and minimal tumor bleeding remains.

The mortality in our case was mainly attributed to massive bleeding from a large tumor volume and

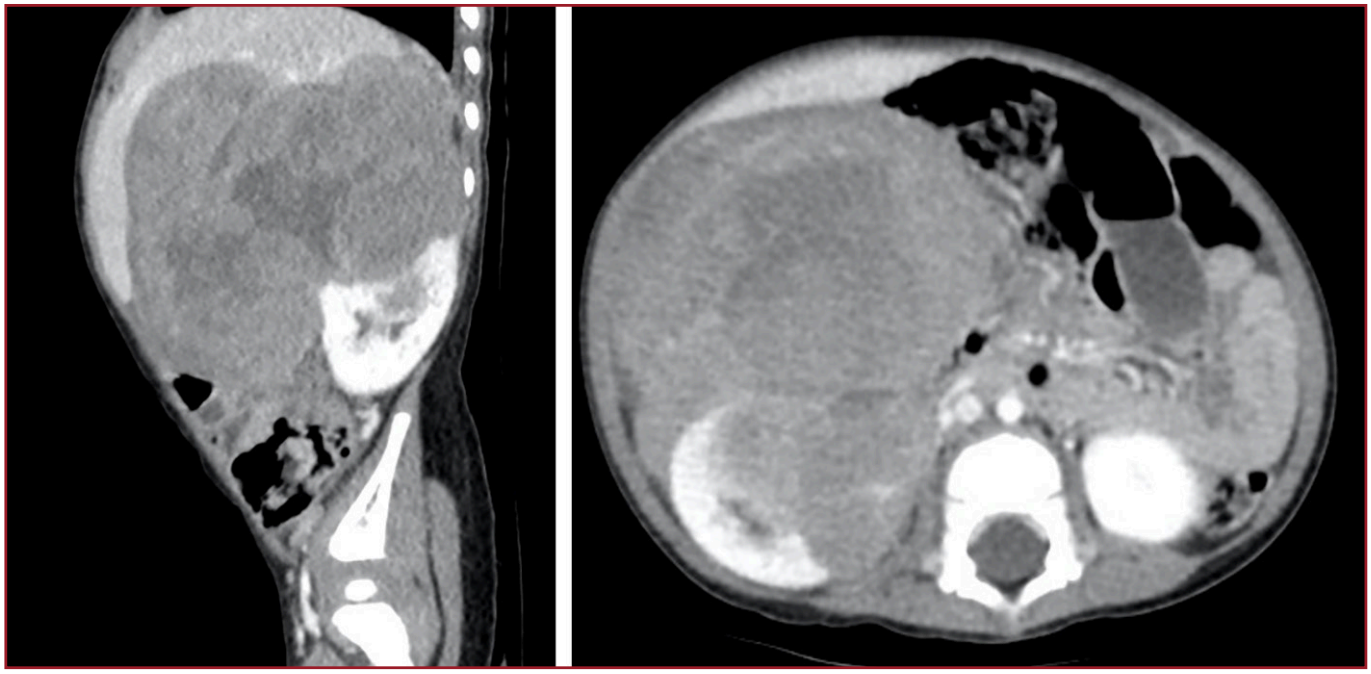

Figure 1. A computed tomography scan of the abdomen at presentation. A) A heterogeneous tumor at retroperitoneal space encases the adrenal gland and partial kidney, B) A heterogenous tumor displaces the inferior vena cava.

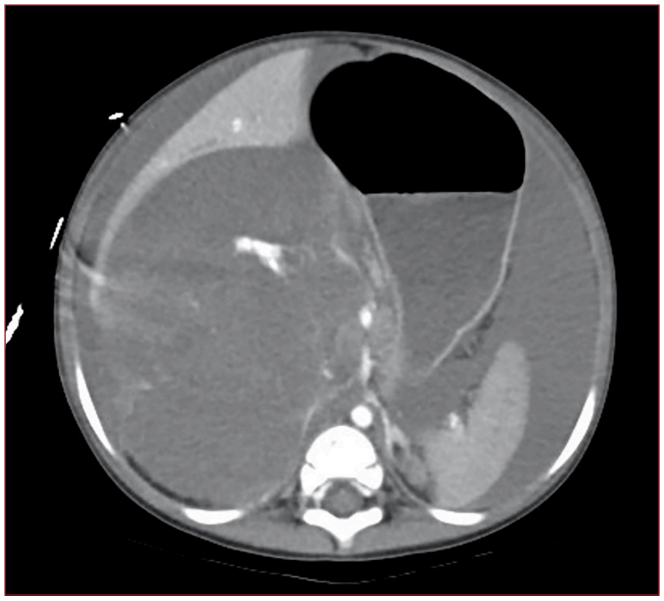

Figure 2. A computed tomography scan of the abdomen on the second day of hospitalization. The image reveals massive ascites with a small abdominal aorta, which is compatible with the status of hemoperitoneum.

coagulation dysfunction. This case highlights the importance of early and accurate diagnosis of NBL. Prompt cancer management decreases the tumor burden, resulting in a desirable disease outcome. In addition to typical differences in radiologic appearance, some studies advocated the use of different modalities to assist in the differential diagnosis. A lower apparent diffusion coefficient (ADC) value in diffusion-weighted imaging of magnetic resonance imaging was reported to be associated with the diagnosis of NBL relative to that of WT.[3] A positron emission tomography based CT scan was also reported to be helpful in the accurate localization of disease.[4] If clinically trapped in a diagnostic dilemma, physicians could turn to different imaging modalities for a precise diagnosis.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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