Pediatric Neuroblastoma and Anemia (Jehovah’s Witness Case Report)

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Abstract

Introduction: Neuroblastoma is the most common extra cranial solid tumor in young children and is derived from embryonic neural crest cells (i.e. is a neuroendocrine tumor) that can have variety of presentations including failure-to-thrive and gastrointestinal symptoms.

Case: A 13 month old whose parents are Jehovah’s Witnesses presented with generalized edema and diarrhea. Laboratory evaluation and upper endoscopy results showed iron deficiency anemia and protein losing enteropathy. Additional imaging of abdomen and pelvis revealed a mass that encased the celiac vessels. Elevated VMA/HVA and CT and MIBG scan diagnosed neuroblastoma without metastases. Since no other sites of disease were present on scans or in the bone marrow, the classification was intermediate risk. Indications, risks, and alternatives of various treatment options were discussed in detail with the parents who wanted to avoid transfusions. Anemia was treated with intravenous and oral iron and darbopoetin. The toddler was treated with 10 cycles of risk-adapted chemotherapy which did not require transfusion (vincristine, cisplatin, and oral cyclophosphamide) followed by surgical removal of the mass. The patient remains in complete remission 4 years after diagnosis.

Discussion: Diarrhea, iron-deficiency anemia, and failure-to-thrive in an infant or toddler can be a presentation of neuroblastoma. Anemia associated with solid tumors in a Jehovah’s Witness can be treated with intravenous iron sucrose, oral ferrous sulfate+Vitamin C, and darbopoetin. In intermediate risk cases, an effective chemotherapy regimen can be used without major reduction of red cell or platelet production to avoid red cell and/or platelet transfusions.

Keywords: Neuroblastoma; Anemia in Jehovah’s witness; Darbopoetin; Tumor; Risk-adapted chemotherapy

Abbreviations: CT: Computed Tomography; MIBG: Metaiodobenzylguanidine; VMA: Vanillylmandelic Acid; HVA: Homovanillic Acid; INRG: International Neuroblastoma Risk Grouping

Introduction

Neuroblastoma is the most common extra cranial solid tumor in children, accounting for 7-8% of all childhood cancers; outcomes after reduced chemotherapy for intermediate-risk neuroblastoma are good [1]. It usually presents as an abdominal mass, but sometimes may present with other systemic symptoms like vomiting, fever, malaise, lethargy, bone pain secondary to metastases. Rarely, neuroblastoma can cause chronic diarrhea caused by effects on the GI system of the various substances secreted by the tumor [2]. Iron deficiency is also a known cause of protein losing enteropathy [3].

We share our experience with an unusual presentation of neuroblastoma in a child with diarrhea and generalized edema, protein losing enteropathy, and severe iron deficiency anemia complicated by the family being Jehovah’s Witnesses for whom blood transfusions are contraindicated because of religious beliefs. In this case transfusion was avoided; anemia was treated with iron sucrose, followed by oral ferrous sulfate + Vitamin C and darbopoetin. The child’s intermediate-risk neuroblastoma was also successfully treated.

Case Presentation

A previously healthy 13 month old girl presented with abdominal distention and dependent edema for 2 days. She was in her usual state of good health until 1 week prior to presentation, when she developed acute watery diarrhea with 6-8 stools/day without blood in stool. On physical examination, she was alert, a febrile with minimal distress secondary to the abdominal distention. Her vital signs were within normal limits for age; she had gained 2 pounds within the last 3 days. Laboratory testing showed normal values for complete metabolic panel other than low protein and albumin (2.9/1.5 gm/dL). Total WBC count with differential was normal. UA was negative for proteinuria, abdominal ultrasound showed mild ascites. She continued to have lower extremity edema and ascites, most prominent in the morning upon waking.

Over the next 2 weeks, she became less playful and stopped walking. Her oral intake decreased and diarrhea worsened. Repeat labs showed microcytic anemia (MCV 75) with a...
Neuroblastoma is an embryonal malignancy derived from the primitive cells of the sympathetic nervous system [4]. It is the most common extra-cranial solid tumor occurring in childhood and has a diverse clinical presentation and course depending on the tumor biology. The tumor can present itself anywhere in the entire sympathetic nervous system; however, adrenal gland and abdominal (extra-adrenal) are the most common sites which have improved 5-year overall survival to 50% [5].

Intermediate-risk disease consisting largely of difficult to resect and symptomatic Stage 2/3 disease and infants with Stage 4 disease have good outcome with cycles of chemotherapy followed by surgical resection. The role of radiotherapy in intermediate risk tumors is related to extent of resection, location, and risk of long term effects. The most commonly used chemotherapeutic agents in intermediate risk disease have been vincristine, cyclophosphamide, cisplatin, doxorubicin, carboplatin, topotecan, and ifosfamide. High-risk disease includes older children with Stage 4 disease and MYCN amplified tumor [6]. Advances in therapy for patients with high-risk disease include intensive induction chemotherapy and myeloablative chemotheraphy, followed by the treatment of minimal residual disease using differentiation therapy and immunotherapy; these have improved 5-year overall survival to 50% [5].

Surveillance studies during and after treatment are able to detect asymptomatic and unsuspected relapse using urinary VMA/HVA, 123I-MIBG scan, and/or CT or MRI. A multidisciplinary team is involved in the treatment, supportive care and follow-up of these children with neuroblastoma. Timely management of a potential problem detected during therapy and follow up visits will maximize health and the quality of life on therapy and after therapy. Our patient became healthy and has stayed healthy.

Our case is somewhat unique in that the high survival rate in intermediate risk disease would not warrant a court order for myelosuppressive therapy and surgery when acceptable alternatives for a Jehovah’s Witness are possible. We achieved an excellent outcome following principles of care: reduction of size of tumor with risk-adapted chemotherapy, followed by careful surgical resection.

**References**


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