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Case Report

Primary pancreatic neuroblastoma presenting with opsoclonus–myoclonus syndrome

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ABSTRACT

Although neuroblastoma is a common solid organ malignancy in children, primary pancreatic neuroblastoma is a rare entity in children, with very few cases reported in the literature. The case discusses the presentation of a 21-month-old female presenting to the neurology clinic with ataxia and erratic eye movements. Our case illustrates the computed tomography, ultrasound, and scintigraphic findings of primary pancreatic neuroblastoma presenting as opsoclonus–myoclonus syndrome. Computed tomography and ultrasound demonstrated a vascular, enhancing mass in the pancreatic body clearly separate from the adrenal gland. Metaiodobenzylguanidine scan demonstrates focal intense uptake in the pancreatic body. The patient's diagnosis was confirmed with biopsy, and her malignancy responded well to conventional chemotherapy. The case is important in that it demonstrates the unusual imaging appearance of a primary pancreatic neuroblastoma.

Introduction

Neuroblastoma is the third most common pediatric malignancy and the most common extracranial solid organ malignancy in children [1]. Approximately 30-35% of neuroblastomas are extra-adrenal in origin and located in the retroperitoneum. However, these tumors can arise from neural crest cells anywhere in the body. Although a common tumor in general in children, primary pancreatic neuroblastoma is quite rare in children and very few cases have been reported in the literature since 1969, several of which represent pancreatic involvement following relapse or disease progression [2–8]. We discuss a rare case of neuroblastoma arising primarily from the pancreas and illustrate the findings on computed tomography (CT), ultrasound, and Meta-iodobenzylguanidine (MIBG) scan imaging.

Case report

A full-term 21-month-old female presented to the pediatric neurology clinic with chronic ataxia. The symptoms began 9 months before presentation in the neurology clinic. Nine months before presentation, the patient was admitted to the neurology service and was diagnosed with acute cerebellar ataxia. Per the family, the patient never recovered, and the patient did not relearn to walk until 15 months. The patient

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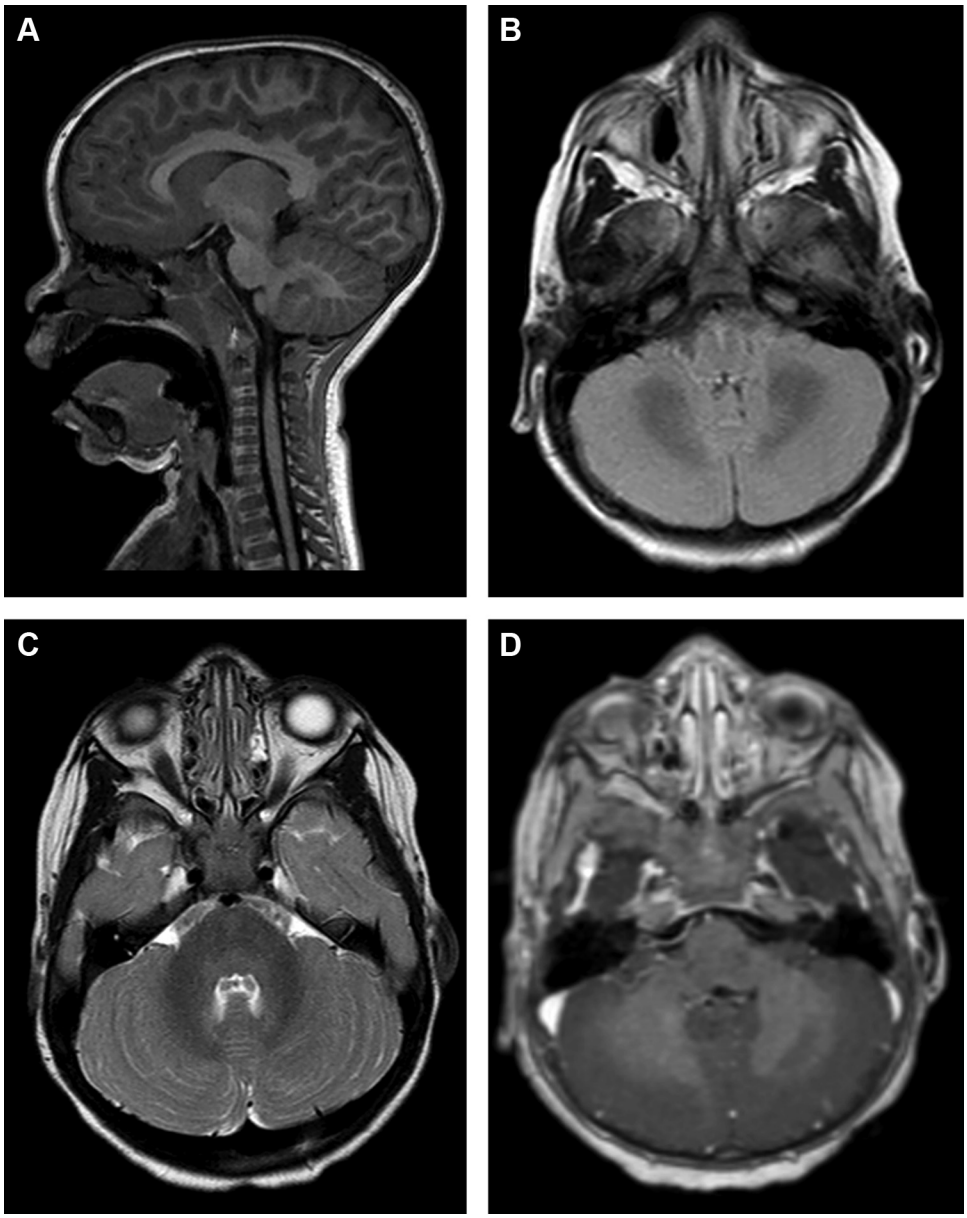

Fig. 1 – (A-D) Sagittal T1 (A), axial T2 fluid-attenuation inversion recovery (FLAIR) (B), axial T2 turbo spin echo (TSE) (C), and axial postcontrast T1 (D) images of the brain demonstrate a normal appearing cerebellum.

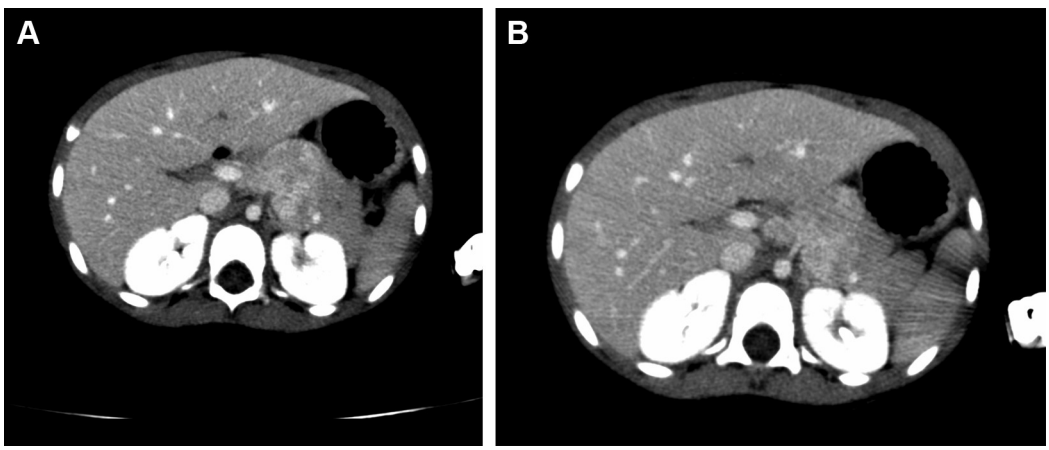

Fig. 2 – (A and B) Axial computed tomography image of the abdomen demonstrating a heterogeneously enhancing mass in the pancreatic body and tail (A) with encasement of the celiac axis (B).