Radiological Sciences Proceedings

UCLA Health

Extra-Adrenal Neuroblastoma in an Infant: A Case Report

Bhupathy SA, MD[1] | Pierce J, MD, PhD[2] | Goldstein J, MD[2] | Ghahremani Koureh S, MD[1]

Author Affiliation: [1] Department of Radiological Sciences, David Geffen School of Medicine at UCLA
[2] Department of Pathology, David Geffen School of Medicine at UCLA

Corresponding Author: S.B. (sbhupathy@mednet.ucla.edu)

UCLA Radiol Sci Proc. 2026;6(1):1-11

Abstract: Neuroblastoma is the most common extracranial pediatric solid tumor and arises most often from the adrenal glands. However, this tumor can occur anywhere along the sympathetic nervous system. Its clinical presentation is variable and depends on the location and extent of the mass. We present the case of an infant who initially had a palpable mass on his back, later developed lower extremity weakness, and was subsequently diagnosed with a paraspinal neuroblastoma. MRI was key in visualizing compression of the lower thoracic spinal cord by the tumor, explaining the patient's symptoms and necessitating tumor debulking. The course of the patient's treatment was complicated by a tumoral hemorrhage that required emergent embolization by an interventional radiologist. This case highlights options for the management of neuroblastoma, including chemotherapy, embolization, and surgical resection, with an emphasis on interdisciplinary decision-making.

Keywords: *neuroblastoma, magnetic resonance imaging, metaiodobenzylguanidine*

Introduction

Neuroblastoma, an embryonal tumor of the autonomic nervous system derived from incompletely differentiated neural crest tissues, is the most prevalent extracranial solid tumor in childhood and a leading cause of cancer-related death in children, usually developing within the first five years of life.[1–4] Arising from tissues of the sympathetic nervous system, neuroblastoma is most commonly located in the adrenal medulla and paraspinal ganglia.[4,5] The clinical presentation is highly variable and depends on the site of origin and on paraneoplastic syndromes and bone marrow infiltration.[6] We present the unique case of an infant with neuroblastoma that was first detected on an ultrasound for a palpable back mass and subsequently characterized by MRI. The patient's course was complicated by the neuroblastoma's

Key Points

- Neuroblastoma is the most common extracranial pediatric tumor and most often arises from the adrenal glands. Extra-adrenal thoracic and cervical tumors generally carry a favorable prognosis; however, they can present with symptoms of spinal cord compression.
- Imaging modalities including ultrasound, magnetic resonance imaging (MRI), and nuclear medicine studies are key in establishing the diagnosis of and determining the prognosis in neuroblastoma cases. In particular, metaiodobenzylguanidine (MIBG) scans can be used for evaluating metastatic disease.
- The management of neuroblastomas must consider tumor size and extent, as well as complications such as hemorrhage, and requires a multidisciplinary approach that may include hematology, oncology, interventional radiology, and surgical teams.

Abbreviations
ALK: anaplastic lymphoma kinase
CT: computed tomography
DWI: diffusion-weighted imaging
MIBG: metaiodobenzylguanidine
MRI: magnetic resonance imaging

paraspinal location, spinal canal invasion, and resultant cord compression, as well as by a hemorrhage of the tumor that required embolization. Chemotherapy was used to reduce the tumor's size, and the tumor was eventually resected. This case report was prepared following the CARE guidelines.[7]

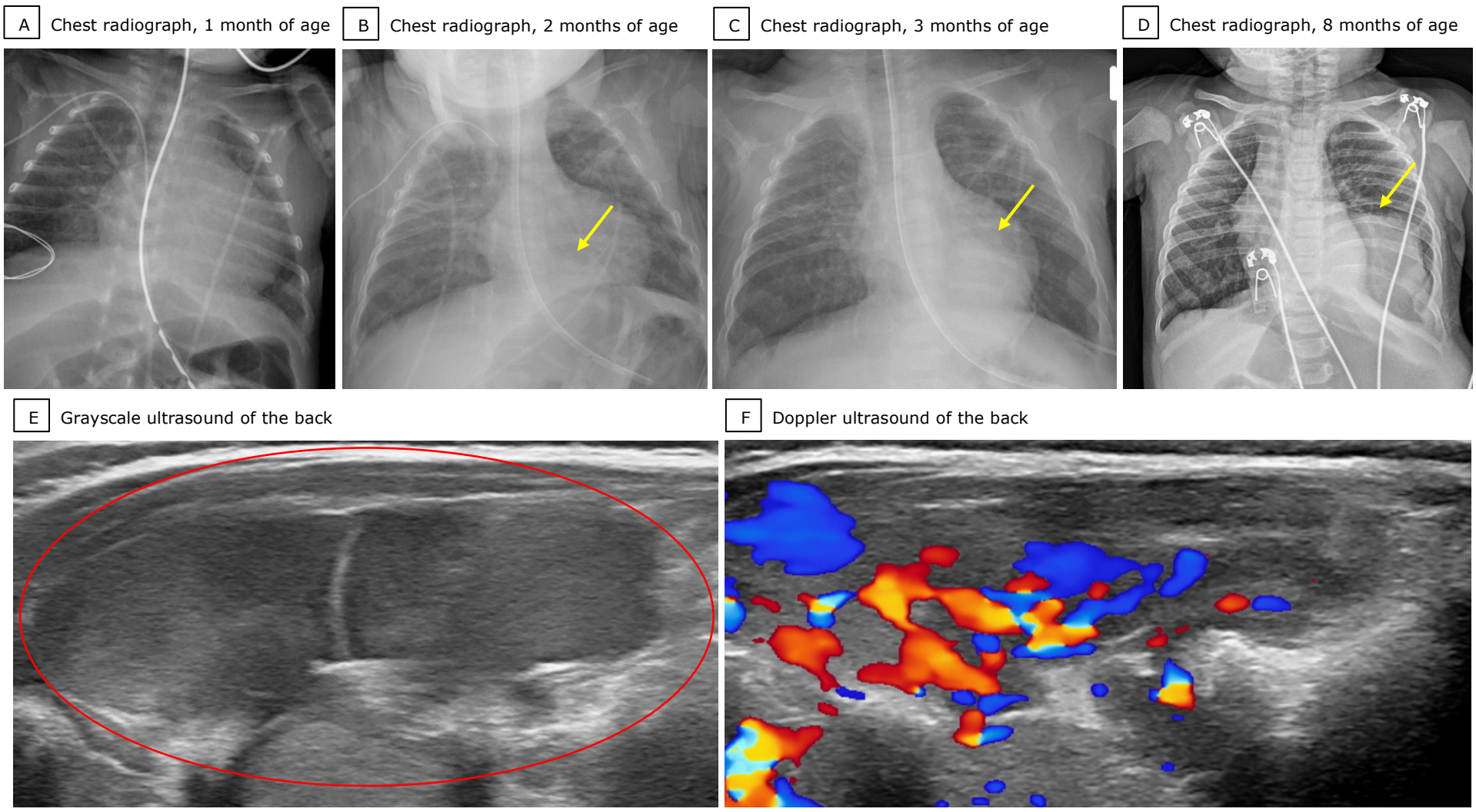
Case Presentation

A newborn boy with a prenatal history complicated by severe fetal growth restriction and maternal diabetes was born at approximately 26 weeks' gestation via cesarean delivery due to nonreassuring fetal heart rate. The patient remained hospitalized for the first 3 months of life, during which time serial chest radiographs were performed to monitor his respiratory status (Figures 1A–C). At 8 months of age, the patient was noted to have a palpable mass on his back and purple discoloration on his legs, which caused concern for a possible vascular malformation such as a hemangioma. Ultrasound of the back revealed a well-circumscribed, solid, hypoattenuating vascular mass (Figures 1E and 1F). The full extent of the mass was not well evaluated via ultrasound, and magnetic resonance imaging (MRI) was recommended for further evaluation.

Before MRI could be performed, the patient developed a fever, respiratory distress, and lower extremity weakness. He was admitted to the hospital, at which time MRI showed a T2-heterogeneous, diffusion-restricting posterior mediastinal mass in the left chest with compression of the lower thoracic thecal sac and spinal cord (Figure 2). Histopathologic analysis of a biopsy specimen confirmed the mass to be a neuroblastoma that was negative for *MYCN* and anaplastic lymphoma kinase (ALK). A nuclear medicine metaiodobenzylguanidine (MIBG) scan did not reveal any additional suspicious foci of uptake to suggest metastatic disease (Figure 3A). The neuroblastoma was staged as L2, as it was localized but had the imaging-defined risk factor of spinal cord compression. The patient underwent laminectomy and tumor debulking by a neurosurgery team before starting a chemotherapy regimen of carboplatin, etoposide, and pegfilgrastim to reduce the size of the mass prior to its resection.

Shortly after he recovered from his operation and was discharged from the hospital, the patient was readmitted for acute fever, increased work of breathing, and decreased oral intake. Chest radiographs showed increasing opacification in the left side of the chest, and a computed tomography angiogram (CTA) of the patient's chest confirmed a large left pleural effusion of intermediate density with rightward mediastinal shift, which caused concern for hemoperitoneum (Figure 4A). Chemotherapy was paused given the patient's acute decompensation. Interventional radiology emergently performed an aortic angiography, which showed that the neuroblastoma had parasitized arterial supply directly from the thoracic aorta and that there was active extravasation from a dominant supplying vessel at the superior margin of the tumor (Figure 4B). This vessel was embolized using Gelfoam and coils (Figure 4C). Chemotherapy was restarted after the patient's clinical condition improved, and repeat MRIs (Figures 4E and 4F) showed that the mass was responding to chemotherapy, decreasing in size and showing less diffusion restriction. The mass was subsequently resected by a

Figure 1. Prediagnostic Images of an Infant Boy with Neuroblastoma

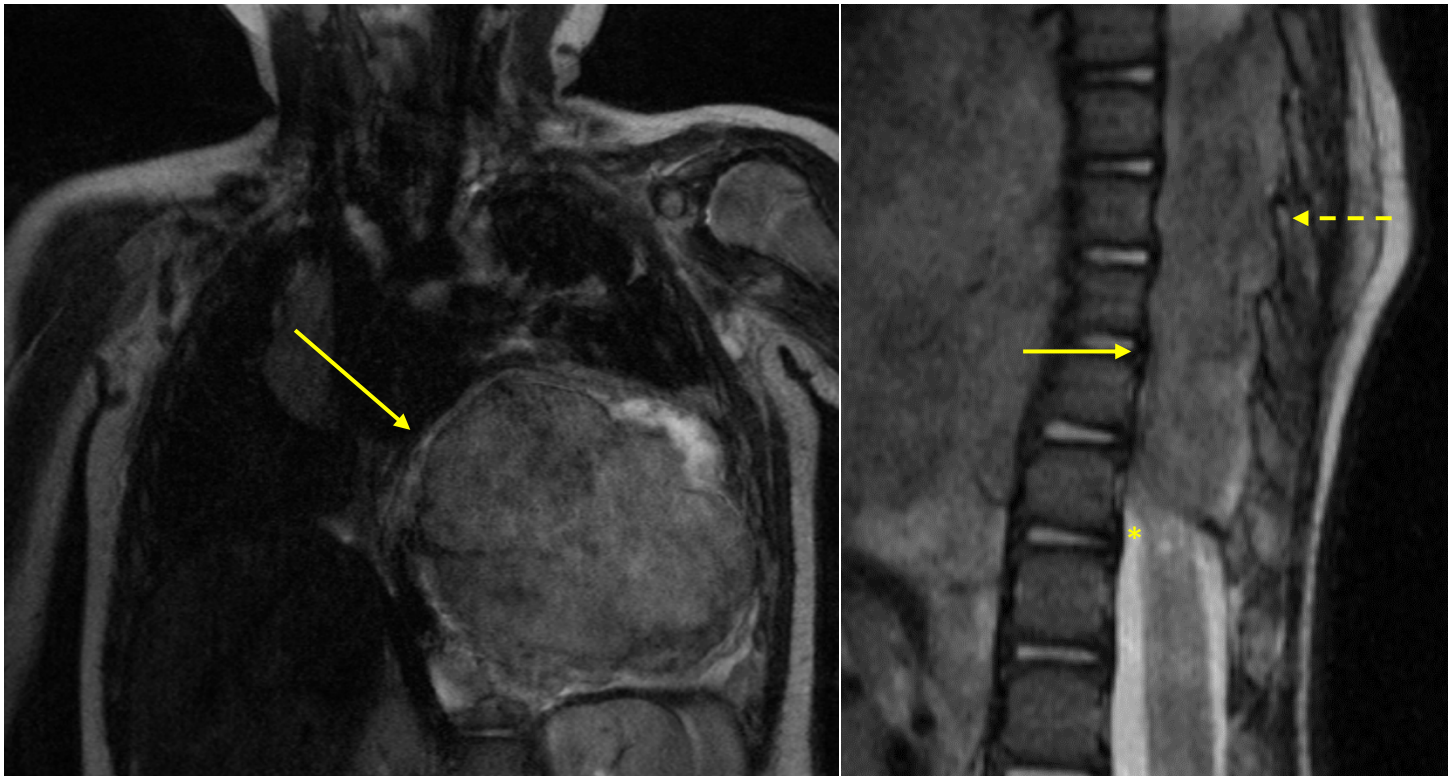


(A-D) During the first months of the patient's life, serial radiographs were obtained to monitor his respiratory status. (A) A chest radiograph obtained when the patient was 1 month old shows a normal cardiomediastinal silhouette for his age. (B) A chest radiograph taken when the patient was 2 months old shows an indistinct retrocardiac opacity (arrow), which is more prominent in (C) a radiograph obtained when the patient was 3 months old (arrow). (D) A radiograph obtained when the patient was 8 months old shows a large opacity in the left chest (arrow), which was evaluated with MRI shortly after this examination. (E, F) Ultrasound images obtained when the patient presented with a palpable mass on his back visualized the mass but were not diagnostic. (E) A grayscale ultrasound of the back in the area of the palpable mass showed a subcutaneous well-circumscribed solid hypoechoic mass (red circle) measuring up to 4.2 cm. (F) Doppler ultrasound showed significant vascularity within this mass. Deep extension was not well evaluated by ultrasound.

Figure 2. Magnetic Resonance Imaging of the Thoracic Spine and Chest of an Infant with Neuroblastoma

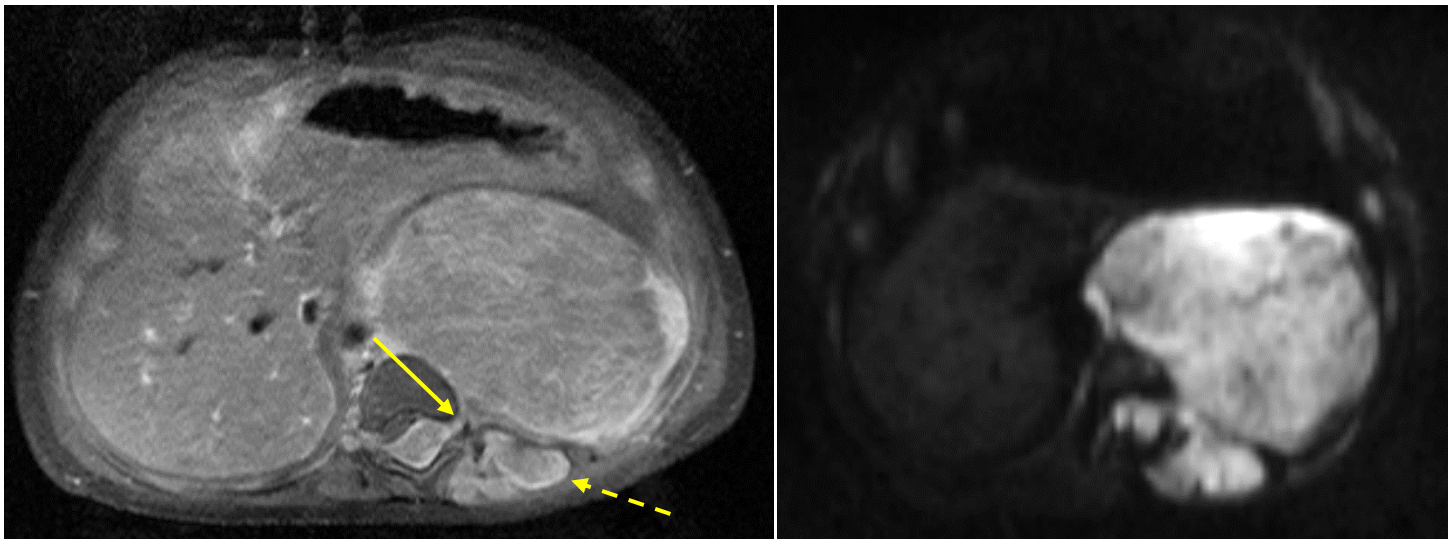
A T2-weighted MRI of the thoracic spine, coronal view

B T2-weighted image of the thoracic spine, sagittal view



C Postcontrast MRI of the chest, axial view

D Diffusion-weighted image (DWI) of the abdomen, axial view

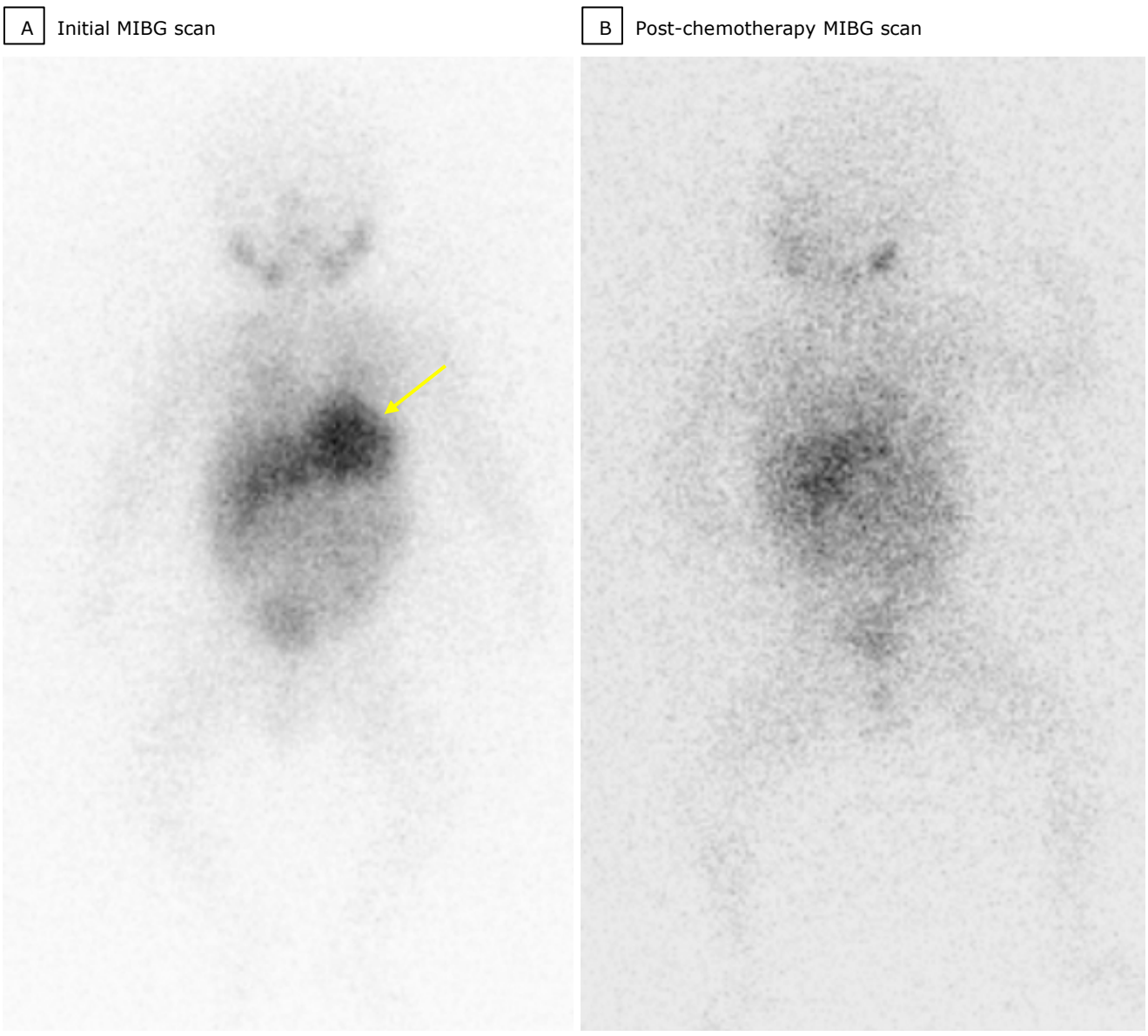


(A) Coronal T2-weighted MRI of the thoracic spine and chest demonstrates a large, T2-heterogeneous structure in the left chest and upper abdomen (arrow). (B) Sagittal T2-weighted MRI of the thoracic spine shows extension along the lower spinal canal with near-circumferential occupation of the epidural space at the levels of T7 through T12 (arrow), compressing and displacing the thecal sac and spinal cord. The spinal canal is enlarged, with scalloped cortical margins related to chronic mass effect (dashed arrow). At the level of compression, there is T2-hyperintensity of the spinal cord, likely related to chronic compressive myelopathy (asterisk). (C) Axial contrast-enhanced MRI of the chest shows a heterogeneously enhancing mass in the left chest with extension into the posterior epidural space through the neural foramina (arrow). There is also subcutaneous extension of the mass lesion, likely corresponding to the component seen on ultrasound (dashed arrow). (D) Axial diffusion-weighted MRI shows restricted diffusion of the mass.

vascular surgery team (Figure 5). Follow-up MIBG and MRI showed no evidence of recurrent or metastatic disease (Figure 3B).

Retrospective review of chest radiographs revealed a slowly growing mass in the left chest (Figures 1A–D). While the initial chest radiograph at one month was within normal limits, there was an indistinct retrocardiac opacity seen at two months, which had increased in size on radiographs made when the patient was three months old. Just prior to the patient's diagnosis at eight months, a

Figure 3. MIBG Scans of an Infant with Neuroblastoma



(A) MIBG scan showing a large mass (arrow) within the left hemithorax extending into the abdomen and demonstrating diffuse intense tracer activity consistent with biopsy-proven neuroblastoma. There were no abnormal foci of tracer activity elsewhere to suggest metastatic disease. (B) Repeat MIBG scan after chemotherapy administration shows near-complete resolution of the prior radiotracer uptake within the left paraspinous region and resolution of uptake in the remainder of the mass.

large left-sided opacity was seen correlating to the findings on subsequent MRI. While early findings were nonspecific and may have represented an infectious process, subsequent radiographs with a growing consolidation were more concerning for neoplasm.

Discussion

Neuroblastomas originate from the neural crest and can arise anywhere in the sympathetic nervous system.[1] The majority of tumors arise in the adrenal medulla, but there are many possible extra-adrenal sites, including the retroperitoneum, posterior mediastinum, pelvis, and paravertebral region.[4,5] Metastasis usually occurs via the hematopoietic system to locoregional lymph nodes and to bone marrow.[4]

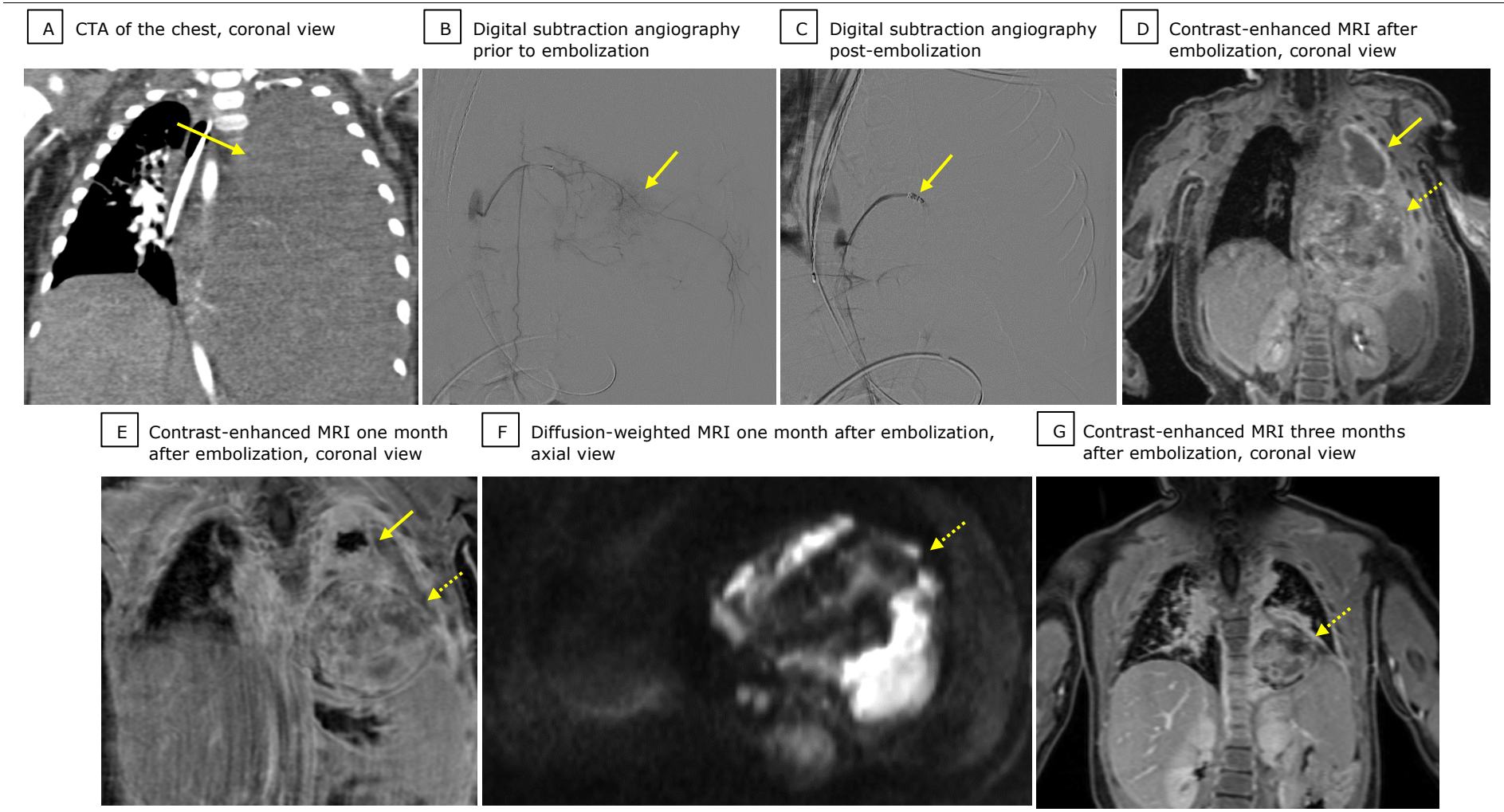
The staging of a neuroblastoma is based on the relationship between the tumor and adjacent structures on imaging.[1] In the chest, encasement of the aorta or its major branches, compression of the trachea or mainstem bronchi, or low mediastinal tumors with infiltration of the costovertebral junction in the lower thoracic spine are imaging-defined risk factors.[1] Involvement of the upper chest can cause Horner's syndrome, which includes ptosis, miosis, and anhidrosis.[4] Tumors of the neck and cervico-thoracic junction can encase vasculature including the carotid arteries, vertebral arteries, and internal jugular veins; they can also compress the trachea.[1,4] In the abdomen and pelvis, tumors infiltrating the porta hepatis, invading the renal pedicles, or encasing the aorta, celiac axis, superior mesenteric artery, vena cava, or iliac vessels are considered higher risk for surgical resection.[1]

Histologically, neuroblastomas are composed of groups of neuroblasts, which have round to oval nuclei and a stippled "salt and pepper" appearance with little cytoplasm, separated by thin septae.[5] The presence of certain neural crest markers on histologic studies is used to differentiate neuroblastoma from other pediatric small round blue cell tumors, including Wilms tumor, Ewing sarcoma, rhabdomyosarcoma, lymphoblastic lymphoma, and desmoplastic small round tumor.[5] The spectrum of neuroblastoma ranges from undifferentiated, in which there is uniformity of primitive small round cells, to differentiated neuroblastoma, in which cells demonstrate features of ganglion cells with large nuclei, vesicular chromatin, and conspicuous nucleoli.[5] The specimen in this case is most consistent with undifferentiated neuroblastoma.

The ultrasound in this case showed a heterogeneous vascular mass, which is a common but nonspecific finding for neuroblastomas.[8] MRI is generally the preferred method for determining the origin and extent of the tumor, including nervous system involvement and bone marrow disease.[6] Its advantages over CT include its lack of radiation and intrinsic high contrast.[9,10] Because the palpable mass in this case was located on the back and the patient subsequently experienced lower extremity weakness, assessing spinal involvement and possible compression was particularly important. Extension involving more than one-third of the spinal canal in the axial plane and abnormal spinal cord signal were image-defined features of high risk.[1]

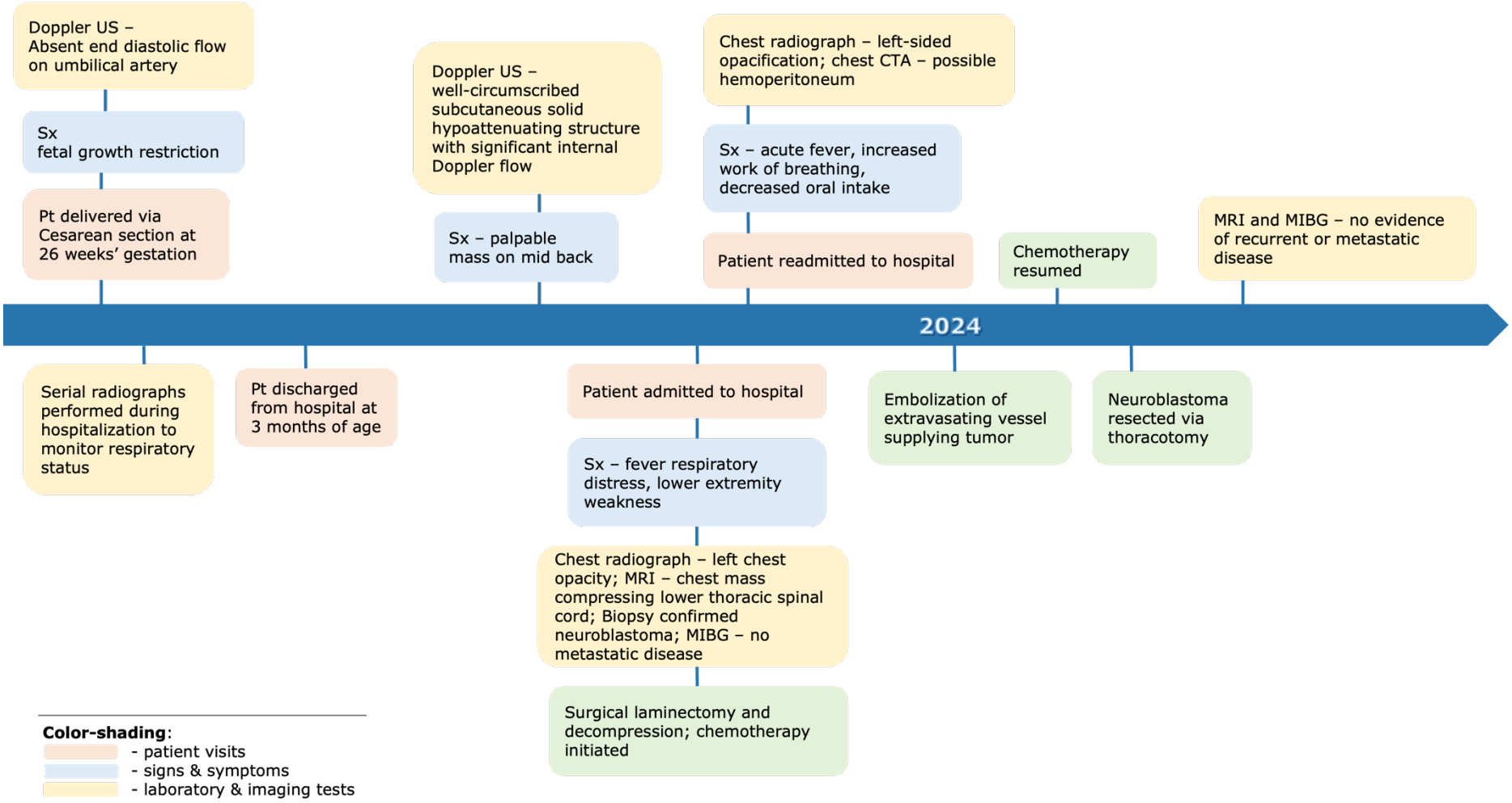
Typically, neuroblastomas are heterogeneous and isointense to hypointense on T1-weighted imaging and heterogeneous and hyperintense on T2-weighted imaging.[10] They usually have heterogeneous enhancement on postcontrast images.[10] Neuroblastomas also show strong diffusion restriction on diffusion-weighted imaging, which can provide valuable information about the heterogeneity of a tumor and guidance for biopsy.[7] The tumor in

Figure 4. Imaging Overview of Treatment Course in an Infant with Neuroblastoma



(A) Coronal CT angiogram of the chest performed after tumor debulking and readmission for fever shows a large left-sided hemothorax (arrow). (B) Digital subtraction angiography shows the vascular supply to the tumor prior to embolization by interventional radiology. The dominant supplying vessel arises from the thoracic aorta with a focus of contrast staining at the superior margin suspicious for active extravasation (arrow). (C) The bleed was successfully embolized with Gelfoam and coils (arrow). (D) Post-embolization coronal contrast-enhanced MRI shows presence of a hemothorax (arrow). (E) One month later, coronal contrast-enhanced MRI demonstrates that the size of the hemothorax has decreased (arrow) and the tumor size has decreased (dashed arrow) with chemotherapy. (F) Diffusion-weighted axial MRI shows heterogeneous restricted diffusion (dashed arrow), as opposed to near-homogeneous diffusion restriction of the initial tumor, likely reflecting chemotherapy response. (G) The hemothorax resolves and the tumor size continues to decrease three months after embolization on this postcontrast coronal MRI just prior to resection (dashed arrow).

Case report timeline

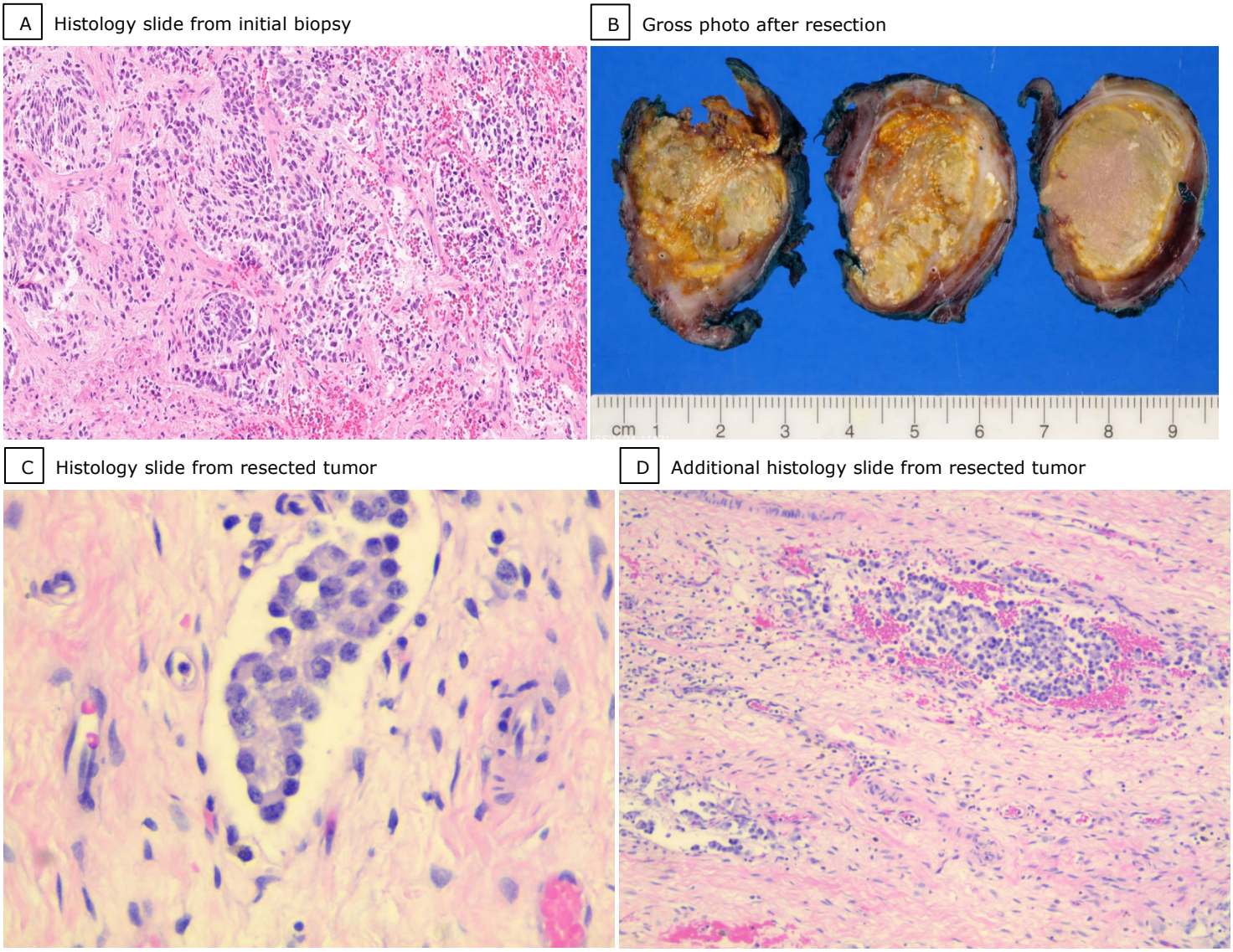


Abbreviations: CTA: computed tomography angiography; MIBG: meta-iodobenzylguanidine; MRI: magnetic resonance imaging; US: ultrasound; Sx: symptoms

this case initially demonstrated diffusion restriction throughout, but after chemotherapy, there was heterogeneous diffusion restriction, which may reflect the decreased concentration of neuroblastic cells. Nuclear medicine studies can be used for diagnosis and staging, with MIBG labeled to iodine-123 having high specificity for sympathetic tissue.[10] This patient had an initial staging MIBG scan, which showed no evidence of metastases, and a subsequent MIBG scan following resection to rule out recurrence.

The management of neuroblastoma varies based on the size and extent of the tumor and whether there is metastatic disease.[10] Once MIBG confirmed that there was no metastatic disease in this case, treatment focused on the challenging position of the primary tumor, which occupied the epidural space and

Figure 5. Pathologic Correlation with Histology Slides and Gross Specimen Images



(A) Histology slide from initial biopsy shows pre-treatment tumor with diffuse involvement by neuroblastic cells. (B) Gross specimen photo shows the resected tumor after chemotherapy. (C, D) Histology slides from the resected tumor show residual neuroblastic cells.

compressed the spinal cord. Tumor debulking alleviated the patient's symptoms of cord compression.

Surgical resection following chemotherapy is one of the mainstays of treatment for neuroblastoma.[1] The clinical course in this case was complicated by hemorrhage of the tumor, which required months of stabilization before the tumor could be resected. Outcomes are generally favorable and mortality risk is decreased when at least 90% of the tumor is resected; children under eighteen months of age at the time of diagnosis, such as the patient in this case, also generally have better outcomes.[1,9]

Conclusion

This case highlights the importance of a multidisciplinary approach to the diagnosis and management of neuroblastoma, especially when a tumor's location presents interventional challenges. After the diagnosis of the mass through MRI and histology and the staging of the mass with an MIBG scan, a neurosurgery team performed tumor debulking to reduce symptoms of spinal cord compression. The patient's oncology team recommended further reduction of the tumor size with chemotherapy prior to resection. Interventional radiologists performed emergency embolization when the tumor hemorrhaged. Finally, vascular surgery resected the tumor after its size was decreased enough to improve prognosis. Interdisciplinary discussion and a team-based approach were essential in management of this complex case.

An additional salient point from this case is the importance of early detection in cases of neuroblastoma, as demonstrated by the retrospective review of radiographs. While recognition of slowly growing masses on serial examinations presents a diagnostic challenge, radiologists can use more remote prior examinations as a baseline for detection. The patient had symptoms of spinal cord compression by the time MRI was performed, which could have potentially been prevented had the mass been identified from its appearance in earlier radiographs.

Author Contributions

Conceptualization, S.G.; Acquisition, analysis, and interpretation of data, S.B., S.G., J.B., and J.G.; Writing – original draft preparation, S.B.; Reviewing and editing, S.G., S.B., J.B., and J.G; Supervision, S.G. All authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. All authors had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

Disclosures

None to report.

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