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American Journal of Sonography

Case Report

Sonography of Primary Cervical Neuroblastoma in the Infant

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Received : 07 January 2021
Accepted : 05 April 2021
Published : 24 April 2021

DOI
10.25259/AJS_1_2021

Quick Response Code:

ABSTRACT

A palpable neck mass in a newborn or young child is overwhelmingly benign but can require additional evaluation to exclude the rare malignant etiology. We present a 10-month-old female with a non-tender, non-erythematous firm nodule in the left neck initially suspected to be enlarged lymph nodes. Sonographic imaging was concerning for a diagnosis of neuroblastoma confirmed by surgical resection and pathologic examination. The sonographic characteristics associated with neuroblastoma are important to recognize to accurately diagnose the neoplasm and improve patient management.

Keywords: Color Doppler, Neoplasm, Neuroblastoma, Pediatric, Sonography

INTRODUCTION

Soft-tissue neck lesions in the infant and young child are common and usually not of long-term clinical concern. The most likely etiology of a pediatric neck mass is congenital (such as thyroglossal duct cyst or branchial remnant) or infectious origin such as reactive lymph nodes which can progress to phlegmonous transformation or abscess. Although rare, malignant etiologies may need to be excluded. These include the small blue cell tumors such as neuroblastoma, rhabdomyosarcoma, and lymphoma. We describe an enlarging neck mass in an infant. Sonographic imaging of the lesion suggested neoplasm, likely neuroblastoma given the young age of the infant. The other benign and malignant conditions that can occur are summarized in [Table 1].

Table 1: Benign and malignant conditions

Benign	Malignant
Thyroglossal duct cyst	Neuroblastoma
Branchial remnant cyst	Rhabdomyosarcoma
Hemangioma	Lymphoma
Lymphatic malformation	
Fibromatosis colli	
Lymph node	
Infectious phlegmon/abscess	

CASE HISTORY

A 10-month-old previously healthy, normally developed female presented with an incidentally discovered non-tender, non-erythematous firm left neck nodule. Suspected to be enlarged lymph nodes, antibiotics were empirically administered by the primary physician. The lesion continued to enlarge despite antibiotic therapy. The pediatrician ordered neck sonography for further evaluation of the neck mass.

Imaging findings

Sonography was performed utilizing 18–9 mHz linear transducer. The sonographic images revealed a well-circumscribed ovoid, mixed echogenic solid lesion in the neck measuring nearly 5 cm in greatest dimension with increased internal color Doppler flow [Figures 1a and b]. This appearance was not consistent with benign reactive lymph nodes and a malignant lesion was considered. Considering the child's young age, primary suspicion was a possible neuroblastoma. Other differential diagnostic possibilities included lymphoma, extraskeletal Ewing's sarcoma, and rhabdomyosarcoma; these are highly unusual in an infant. Abdominal sonographic imaging was performed to evaluate the adrenal glands. These images revealed no suprarenal mass to suggest metastatic spread to the neck from the much more common adrenal neuroblastoma [Figures 2a and b].

Pathologic findings

The child was referred to pediatric surgery for excision. Pathologic evaluation revealed a small round, poorly differentiated blue cell tumor [Figure 3]. The mitosis-karyorrhexis index was high in the sample. Ipsilateral lymph nodes were also resected and were positive for the same neoplasm. The neoplasm was immunonegative for myogen and MYOD, CD99, and CD45, eliminating from the diagnostic differential rhabdomyosarcoma, extraosseous Ewing's sarcoma, and lymphoma. Strongly positive CD56 and neuron-specific Enolase immunohistochemistry supported a diagnosis of neuroblastoma.

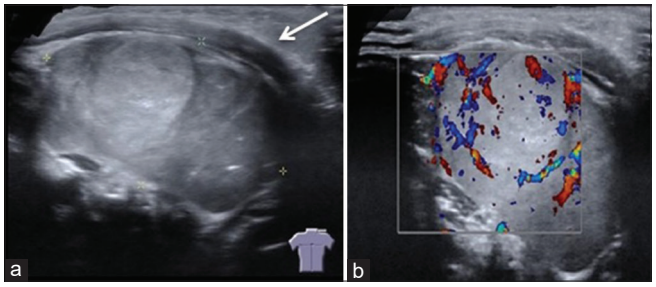

Figure 1: A 10-month-old female infant with primary cervical neuroblastoma who presented with palpable neck mass. (a) Longitudinal sonographic image of the neck demonstrates solid mass with mixed echogenicity (marked by calipers), displacing the adjacent sternocleidomastoid muscle (arrow). (b) Longitudinal sonographic image of the neck mass with color Doppler that confirms increased internal vascularity concerning for neoplasm.

Patient follow-up

Post-resection, the child was referred to pediatric oncology. Further cross-sectional imaging confirmed no distant metastatic spread. The infant underwent successful chemotherapeutic treatment and is disease-free 2-year post-diagnosis.

DISCUSSION

Neuroblastoma is the most common malignant neoplasm found in the extracranial soft tissues in children under the

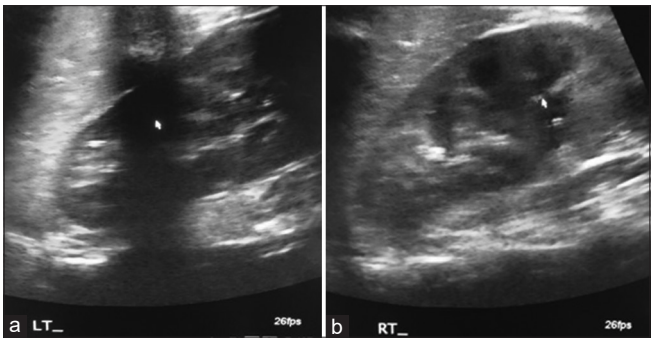

Figure 2: A 10-month-old female infant with primary cervical neuroblastoma who presented with palpable neck mass. (a) Longitudinal sonographic images of left suprarenal region confirm absence of abdominal adrenal mass. (b) Longitudinal sonographic images of right suprarenal region confirm absence of abdominal adrenal mass.

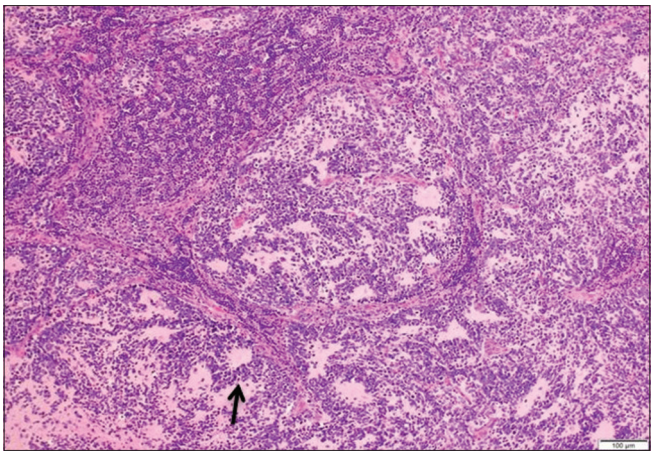

Figure 3: Low-power micrograph of the resected tumor cells showing sheet-like growth of neuroblastoma/small blue cells with several structures suggestive of rosette formation (arrow) that consists of a halo or spoke-wheel arrangement of cells surrounding a central core or hub. Rosettes are a classic histologic architectural pattern seen with tumors such as neuroblastoma.

age of 2 and is the third most common in overall pediatric cancer incidence.[1] Neuroblastoma is a tumor of neuroblasts (immature, undifferentiated nerve cells) derived from the embryologic neural crest. Neuroblasts may be identified in sympathetic nervous tissue at any point along the posterior spinal sympathetic chain.[2]

Primary neuroblastoma is usually found in the adrenal glands or abdomen (71%), sonographically presenting as a mixed echogenic mass in the suprarenal location [Figure 4]. The tumor can also appear as a posterior mediastinal mass in the chest (15%). Cervical primary neuroblastoma is unusual, only 3–5% of neuroblastomas arise in the neck.[2,3] More often, cervical neuroblastoma is observed when neuroblastoma becomes metastatic, spreading from common origin sites such as the adrenal glands.

Palpable neck masses in children are typically enlarged lymph nodes due to an infectious etiology. The commonality of cervical lymphadenopathy may for a time mask cervical neuroblastoma when initially mischaracterized. Treatment failure with antibiotics should be followed by reevaluation and additional diagnostic studies.

Ultrasound is the preferred initial imaging modality for a pediatric neck mass. Neuroblastoma will sonographically present as a heterogeneous mass with increased internal vascularity.[4,5] Because of the high likelihood that neuroblastoma is metastatic, subsequent sonography of the adrenal glands should follow. In this patient, ultrasound of the abdomen bilaterally revealed no mass lesion to suggest metastatic spread to the neck from the much more common adrenal neuroblastoma.

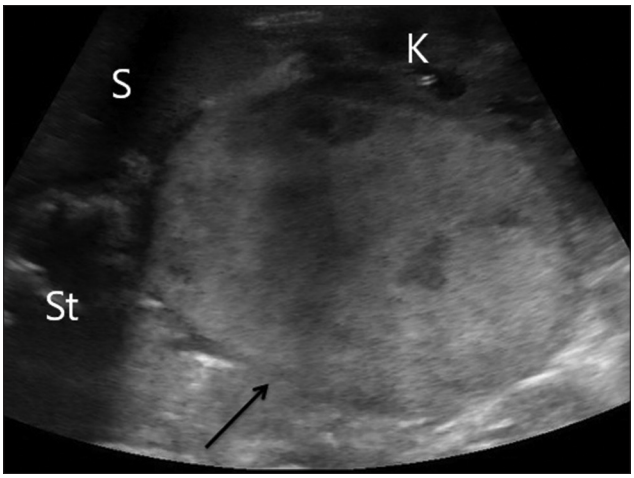

Figure 4: A 1-month-old female infant with neuroblastoma who presented with palpable left upper quadrant abdominal mass. Longitudinal prone sonographic image of the left upper quadrant identifies a round, mixed echogenic suprarenal/adrenal mass (arrow) that abuts the spleen (S) and stomach (St) and displaces the kidney (K).

Primary cervical neuroblastoma is rare but this location tends to have a more favorable outcome.[4] Neuroblastoma makes up 7% of childhood cancers yet is responsible for 15% of childhood cancer deaths. Prognostic indicators include tumor stage when identified and the age of the child. Children older than 18 months have a lower survival rate than those younger.[1,3] Although it is rare to find neuroblastoma in children over the age of 5, these children have significantly decreased survival rates.

Staging of this tumor using International Neuroblastoma Risk Group guidelines was Stage 2b (confined to the left neck but with ipsilateral lymph node involvement).[6] The current survival rate for primary cervical neuroblastoma in infants is 74–82%; 30 years prior 5-year survival was only 52%.[7] The improved infant mortality can be attributed to improved identification and treatment methods.[8]

Disease stratification can also be dependent on genetic factors such as the proto-oncogene MYCN. This transcription factor gene is responsible for production of a protein involved in tissue and organogenesis, cell proliferation, and apoptosis. Amplification of the MYCN gene in neuroblastoma is a prognostic marker suggesting a poor prognosis for the individual harboring this mutation.[2,9] The MYCN oncogene is found in 5% of localized neuroblastoma; however, when the mass disseminates, the gene is observed in 30–40% of cases.[9] The infant in this case did not have an aggressive tumor and lacked MYCN gene amplification. Almost all cases of neuroblastoma occur sporadically and do not relate to genetic inheritance.[2]

When a child presents with neuroblastoma specifically in the head-and-neck area, other concerning conditions may exist. In older children, rhabdomyosarcoma and lymphoma should be considered as well as the rare extraosseous Ewing's sarcoma. A related complication commonly observed with pediatric neck masses is Horner's syndrome which occurs when the oculosympathetic system is disrupted. Unilateral ptosis, miosis, and anhidrosis are symptoms seen in afflicted individuals.[1] Neuroblastoma when arising in the sympathetic chain can damage the superior cervical sympathetic ganglion, resulting in Horner's syndrome.[2]

Treatment options are dependent on the age of the child and the stage of the neuroblastoma, requiring an initial surgical biopsy/resection to characterize the tumor. High-risk neuroblastoma (occurring in children >18 months of age with distant metastases as well as in children with unresectable tumors) has a 40–50% 5-year survival rate. Localized and resectable neuroblastoma, as in this patient, has a 5-year survival >95%.

CONCLUSION

We describe a non-tender, palpable left neck mass in a 10-month-old infant. Sonographic examination displayed

a well-circumscribed ovoid, mixed echogenic solid lesion with increased internal color Doppler flow. The distinctive and unique sonographic and clinical features in this infant allowed for a primary diagnosis of neuroblastoma. Sonography of the bilateral adrenal glands in the abdomen revealed no mass lesion to suggest metastatic spread to the neck from the much more common adrenal origin of neuroblastoma. Surgical resection, pathologic examination, and subsequent appropriate chemotherapy resulted in a 2-year disease-free outcome.

Declaration of patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Irwin MS, Park JR. Neuroblastoma: paradigm for precision medicine. Pediatr Clin North Am 2015;62:225-56.
2. Alvi S, Karadaghy O, Manalang M, Weatherly R. Clinical manifestations of neuroblastoma with head and neck involvement in children. Int J Pediatr Otorhinolaryngol 2017;97:157-62.
3. Singh H, Mohan C, Mohindroo NK, Sharma DR. Cervical neuroblastoma. Indian J Otolaryngol Head Neck Surg 2007;59:288-90.
4. Kadom N, Lee EY. Neck masses in children: Current imaging guidelines and imaging findings. Semin Roentgenol 2012;47:7-20.
5. Chu CM, Rasalkar DD, Hu YJ, Cheng FW, Li CK, Chu WC. Clinical presentations and imaging findings of neuroblastoma beyond abdominal mass and a review of imaging algorithm. Br J Radiol 2011;84:81-91.
6. Monclair T, Brodeur GM, Ambros PF, Brisse HJ, Cecchetto G, Holmes K, *et al*. The international neuroblastoma risk group (INRG) staging system: An INRG task force report. J Clin Oncol 2009;27:298-303.
7. Rekha MI, Suresh M. Imaging of the head and neck in the pediatric patient. J Pediatr Neuroradiol 2016;5:185-95.
8. Colon NC, Chung DH. Neuroblastoma. Adv Pediatr 2011;58:297-311.
9. Yoshimoto M, De Toledo SR, Caran EM, de Seixas MT, de Martino Lee ML, de Campos Vieira Abib S, *et al*. MYCN gene amplification. Identification of cell populations containing double minutes and homogeneously staining regions in neuroblastoma tumors. Am J Pathol 1999;155:1439-43.

How to cite this article: Justice KC, Kriss SM, Davis DG. Sonography of Primary Cervical Neuroblastoma in the Infant. Am J Sonogr 2021;4:2.