Thoracic neuroblastoma

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We describe the case of a 9-day-old female referred for a left posterior mediastinal mass, which we diagnosed as a thoracic neuroblastoma (TNBL) by biopsy. We present this case report to illustrate the classic radiographic, CT, MRI, and nuclear-medicine features of the disease. We believe it is an informative teaching resource for trainees and radiologists. TNBL is a solid childhood tumor of neuroblast origin, which presents in children most often in the first years of life. Treatment of TNBL is largely directed by staging and the criteria for staging rely heavily on multimodal imaging—especially MRI and I-123 metaiodobenzylguanidine (MIBG) bone scan.

Case report

A 9-day-old female was referred to our hospital for evaluation of a large left posterior mediastinal mass with thinning and bony destruction of the 4th and 5th ribs as visualized by CT. The patient was born to a 28-year-old G3P2-3 mother via spontaneous vaginal delivery. The patient’s oxygen saturations were in the 70s after delivery. In the days following her birth, the patient was noted to have labored breathing and sweating upon feeding. A chest radiograph was performed at an outside hospital, and it was mistakenly believed to show cardiomegaly and a large thymus (Figs. 1A and 1B). Echocardiogram was unremarkable except for septal right ventricular hypertrophy and increased pulmonary pressures. On the day the patient was referred to us, a chest CT revealed the mediastinal mass (Fig. 2).

After the child was transferred to our facility, an MRI study showed a 2.4 x 4.7 x 2.9-cm heterogeneously enhancing mass in the left posterior mediastinum that extended through the neural foramen at multiple levels and that demonstrated a posterior extradural component within the

Citation: Rudolf JW, Thapa M. Thoracic neuroblastoma. Radiology Case Reports. [Online] 2011;6:440.

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Competing Interests: The authors have declared that no competing interests exist.

DOI: 10.2484/rcr.v6i2.440
thoracic spine, deviating and flattening the spinal cord anteriorly from the T2 through T8 levels (Figs. 3A and 3B). The patient was immediately taken to the OR for thora
cotomy with biopsy of the chest mass as well as bilateral posterior iliac crest bone marrow aspirates and biopsies. Frozen sections demonstrated poorly differentiated histol-
gy (shwannian-poor) with a low mitotic-karyorrhectic in-
dex (1%), and no amplification of the N-MYC gene. Bone-
marrow biopsies were bilaterally negative for neuroblas-
toma metastasis. The infant was diagnosed with
intermediate-risk neuroblastoma, a Hickman catheter was
placed, and the patient was returned to the Infant Intensive
Care Unit. The infant tolerated the procedure well without
complication.

Radiation therapy was initiated four days after surgery,
following marked decline in the patient's neurologic func-
tion in the lower extremities. Her first cycle consisted of
1080 cGy delivered in 180-cGy fractions. An I-123 MIBG
scan, performed ten days after the surgery, showed intense
uptake of I-123 MIBG at the site of the mediastinal mass (Fig. 4). No abnormal soft tissue or osseous MIBG uptake
was observed. A second cycle of carpoplatin and etoposide
was started 16 days after initiation of the radiation therapy.

Followup appointments approximately three months after
the surgery showed a well-nourished, well-developed child,
in no apparent distress. The patient’s father noted that the
girl's respiratory status was improving despite periodic stri-
dor and tachypnea. The infant was found to be moving her
extremities more. A care plan was established for future followup. MRI performed at this time showed improvement.

Discussion

TNBL is a solid tumor of childhood composed of highly heterogeneous neuroblasts originating along paraspinal areas of the sympathetic nervous system (1, 2). Although thoracic neuroblastomas account for a minority of all neuroblastoma cases (11-26%), they remain the most common mediastinal mass in patients less than two years of age (3). TNBL has a relatively favorable prognosis and is generally associated with better outcomes than neuroblastomas arising in other areas (3, 4). Although the mechanism for the more favorable prognosis is unclear, it is known that thoracic neuroblastomas tend to be detected at an earlier age and present with a higher frequency of localization at the time of diagnosis when compared with nonthoracic neuroblastomas (3, 4).

Patients with TNBL may present with symptoms affecting a variety of organ systems: respiratory (infection, distress, pneumonia), neurologic (ataxia, Horner’s syndrome, myoclonic jerk), or urogenital (urinary tract infections) (5). However, the presentation is highly variable, and only half of thoracic neuroblastoma patients with intraspinal extension are symptomatic at the time of diagnosis (6). The average age at presentation is two years (5). Spontaneous regression is possible and can happen in up to 50% of cases (7). However, this phenomenon seems to occur only in patients under two years of age and is undocumented in older patients (5).

Preliminary imaging of neuroblastoma patients is targeted at addressing symptoms and may include chest and abdominal radiographs, skeletal films, abdominal ultrasound, or spinal MRI (1). Asymptomatic cases are often discovered by screening or as an incidental finding (2). In cases of TNBL, a chest radiograph is sufficient to suggest diagnosis (6). However, multimodal imaging is necessary to assess the extent of the disease. MRI is considered the best imaging modality for staging neuroblastomas due to its accuracy in determining nodal extension, intraspinal extension, and chest wall involvement (1, 6). On MRI, the tumor is typically heterogeneous in appearance, with variable enhancement and prolonged T1 and T2 relaxation times. In addition to MRI staging, an I-123 MIBG bone scan should be performed to look for distant disease (1, 2). Alternatively, I-131 may be used, but it is generally considered to be inferior to I-123 in this application (1). In cases of distant disease, dissemination occurs via the lymphatic circulation or hematogenously (2). Imaging of the brain may be indicated in the presence of neurologic symptoms (2). Differentiation between neuroblastoma, ganglioneuroblastoma, and ganglieneuroma may not be possible with imaging alone (1). TNBL cases are staged according to the International Neuroblastoma Staging System (INSS) (1, 2). Classification consists of four stages that weigh the localization and resectability of the tumor. Patients may also be histologically stratified with either the Shimada or the Paediatric Oncology Group (POG) classification systems (1, 2). The Shimada system uses morphologic features and patient age at diagnosis, while the POG also considers INSS stage, DNA index, and MYCN amplification status (2).

Treatment of TNBL is like that of other nonthoracic neuroblastomas (3). It is based on tumor stage and biological properties, and is largely risk-oriented (1, 3). Low-risk patients with localized tumors achieve excellent results with resection alone, whereas higher-risk patients with distant disease require chemotherapy or radiotherapy in addition to surgery (2). Radical surgery is rarely indicated, and the goal of surgery should be to achieve the preservation of anatomy as much as the resection of tumor (3, 4). Recent studies have shown there to be no difference in 5-year survival rate or event-free survival among patients undergoing complete or incomplete resection (3, 4). Accordingly, complete resection may not be a necessary treatment goal (4). While posterolateral thoracotomy has been the traditional surgical approach and has good prognosis, it is also associated with a long recovery period and high surgical morbidity (8). Recently, thoracoscopy has been employed with apparent success. It offers surgeons better visualization of the anatomy while providing patients with cosmetic advantages and shorter hospital stays (8).

TNBL is an unusual presentation of neuroblastoma. Although it is outnumbered by abdominal neuroblastomas at a level of three to one, it is still the most common mediasti-
nal mass in children under two years. TNBL should always be included in the differential diagnosis of young children presenting with movement disorders or nystagmus that is difficult to explain (3). Since tumor treatment depends highly on staging, multimodal imaging with an emphasis on MRI is critical to patient outcomes.

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