Retropharyngeal Aberrant Thymus

Samir S. Shah, MD*; Stephen Y. Lai, MD, PhD‡; Eduardo Ruchelli, MD§; Ken Kazahaya, MD‡; and Soroosh Mahboubi, MD‖

ABSTRACT. *Introduction.* **Upper airway obstruction from a retropharyngeal mass requires urgent evaluation. In children, the differential diagnosis includes infection, trauma, neoplasm, and congenital abnormalities. Aberrant cervical thymic tissue, although occasionally observed on autopsy examination, is rarely clinically significant. We present the case of an infant with respiratory distress attributed to aberrant thymic tissue located in the retropharyngeal space.**

Case. **A 6-week-old infant was brought to the emergency department for evaluation of stridor associated with periodic episodes of cyanosis. Lateral neck radiograph revealed widening of the retropharyngeal soft tissues. The patient's symptoms did not improve with intravenous ampicillin-sulbactam. Magnetic resonance imaging (MRI) performed on the seventh day of hospitalization revealed a retropharyngeal mass that extended to the carotid space. The mass was easily resected using an intraoral approach. Microscopic examination demonstrated thymic tissue. A normal thymus was also observed in the anterior mediastinum on MRI. The patient recovered uneventfully and had no further episodes of stridor or cyanosis.**

Discussion. **Aberrant cervical thymic tissue may be cystic or solid. Cystic cervical thymus is more common, and 6% of these patients present with symptoms of dyspnea or dysphagia. Aberrant solid cervical thymus usually presents as an asymptomatic anterior neck mass. This case is unusual in that solid thymic tissue was located in the retropharynx, a finding not previously reported in the English literature. Additionally, the patient presented in acute respiratory distress, and the diagnosis was confounded by the presence of mild laryngomalacia. In retrospect, our patient likely had symptoms of intermittent upper airway obstruction since birth. The acute respiratory distress at presentation was likely the result of laryngomalacia exacerbated by the presence of aberrant thymic tissue and a superimposed viral infection.**

Aberrantly located thymic tissue arises as a consequence of migrational defects during thymic embryogenesis. The thymus is a paired organ derived from the third and, to a lesser extent, fourth pharyngeal pouches. After its appearance during the sixth week of fetal life, it descends to a final position in the anterior mediastinum, adjacent to the parietal pericardium. Aberrant thymic tissue results when this tissue breaks free from the thymus as it migrates caudally. Therefore, aberrant thymic tissue may be found in any position along a line from the angle of the mandible to the sternal notch, and in the anterior mediastinum to the level of the diaphragm. In an autopsy study of 3236 children, abnormally positioned thymic tissue was found in 34 cases (1%). The aberrant thymus was most often located near the thyroid gland (n = 19 cases) but was also detected lower in the anterior neck (n = 6 cases), higher in the anterior neck (n = 8 cases), and at the left base of the skull (n = 1 case). The presence of thymic tissue in the retropharyngeal space in our patient is more unusual given the typical embryologic origin and descent of the thymus in the anterior neck to the mediastinum.

Children with aberrant thymus may have associated anomalies. Twenty-four of 34 children (71%) with aberrant thymus detected at autopsy had features consistent with DiGeorge syndrome, and only 5 of the remaining 10 patients had a normal mediastinal thymus present. Our patient had normal serum calcium levels after excision and a mediastinal thymus was visualized on MRI.

Biopsy is required for diagnosis of cervical thymus and should also be considered to exclude other causes. MRI is helpful in delineating the presence, position, and extent of thymic tissue. Immunologic sequelae or recurrence after resection of an aberrant cervical thymus has not been reported. *Pediatrics* **2001;108(5). URL: http://www.pediatrics.org/cgi/content/full/108/5/e94; *retropharynx, thymus, child.***

ABBREVIATION. MRI, magnetic resonance imaging.

The differential diagnosis of a retropharyngeal mass in the infant is broad. Infections and neoplasms, such as neuroblastoma or teratoma, are the most common causes of retropharyngeal mass in this population. Less common causes include ectopic thyroid, angioneurotic edema, congenital myxedema, and lymphadenopathy associated with Langerhans' cell histiocytosis or Kawasaki disease.[1] We report the case of an infant with solid retropharyngeal thymic tissue who presented with respiratory distress, an unusual presentation of an uncommon entity.

CASE REPORT

The patient was a 6-week-old black boy brought to the emergency department with a 2-day history of coughing associated with cyanosis. His history was remarkable for noisy breathing since the first week of life that worsened with supine positioning and agitation. He had not been evaluated previously and had not required prior hospitalization.

On examination, the patient was in mild respiratory distress. There was audible stridor associated with periodic episodes of cyanosis. His temperature was 38.3°C; heart rate, 120 beats per

From the Divisions of *General Pediatrics, ‡Otorhinolaryngology, §Pathology, and ‖Radiology, Children's Hospital of Philadelphia, Philadelphia, Pennsylvania.

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Reprint requests to (S.S.S.) Children's Hospital of Philadelphia, 34th St and Civic Center Blvd, Second Floor, Division of General Pediatrics, Philadelphia, PA 19104. E-mail: shahs@email.chop.edu

minute; respiratory rate, 60 breaths per minute; blood pressure, 110/50 mm Hg; and percutaneous oxygen saturation, 100% in room air. Mild subcostal retractions were present, but the lungs were clear to auscultation. The abdomen was soft without hepatosplenomegaly or palpable mass. The remainder of the physical examination was normal.

Laboratory analysis revealed a white blood cell count of 24 500/mm^3 (0% band forms, 30% segmented neutrophils, and 57% lymphocytes). On cerebrospinal fluid examination there were 3 leukocytes/mm^3 and 1360 erythrocytes/mm^3; no bacteria were seen on Gram stain. Urinalysis revealed no protein or leukocyte esterase. Blood, cerebrospinal fluid, and urine cultures were sterile. Antigens of adenovirus; influenza A and B viruses; parainfluenza virus types 1, 2, and 3; and respiratory syncytial virus were not detected by immunofluorescence of nasopharyngeal washings. Hemoglobin, platelet count, serum electrolytes, blood urea nitrogen, creatinine, calcium, and prothrombin and partial thromboplastin times were normal.

Mild laryngomalacia was diagnosed by flexible laryngoscopy. A lateral neck radiograph revealed widening of the retropharyngeal soft tissues (Fig 1). Chest radiograph was normal. Intravascular contrast-enhanced computerized axial tomography of the neck demonstrated a nonenhancing soft-tissue mass in the left parapharyngeal region that was thought to represent an inflammatory process with phlegmon. The patient was treated with ampicillin-sulbactam intravenously. Although the patient remained afebrile, a lateral neck radiograph repeated on the fifth day of hospitalization was unchanged and the patient continued to have frequent episodes of severe stridor associated with cyanosis. Therefore, magnetic resonance imaging (MRI) of the neck was performed under general anesthesia on the seventh day of hospitalization. MRI revealed a retropharyngeal mass (2.5 cm × 2.4 cm × 0.7 cm) extending from the first to the second cervical vertebrae, with extension to the left carotid space (Fig 2). A normal thymus was observed in the anterior mediastinum.

Additional laboratory evaluation was performed because of concern for neoplasm. Serum lactate dehydrogenase, uric acid, β-human chorionic gonadotropin, ferritin, and urine homovanillic acid and vanillylmandelic acid were normal. Ultrasound of the abdomen, including the kidneys, was normal. Biopsy of the retropharyngeal mass was performed on the 10th day of hospitalization. An intraoral approach was used and a well-circumscribed mass was easily resected. Microscopic examination demonstrated thymic tissue (Fig 3). The patient recovered uneventfully and was discharged from the hospital on the 13th day of hospitalization without stridor or further episodes of cyanosis. No imaging studies were performed postoperatively.

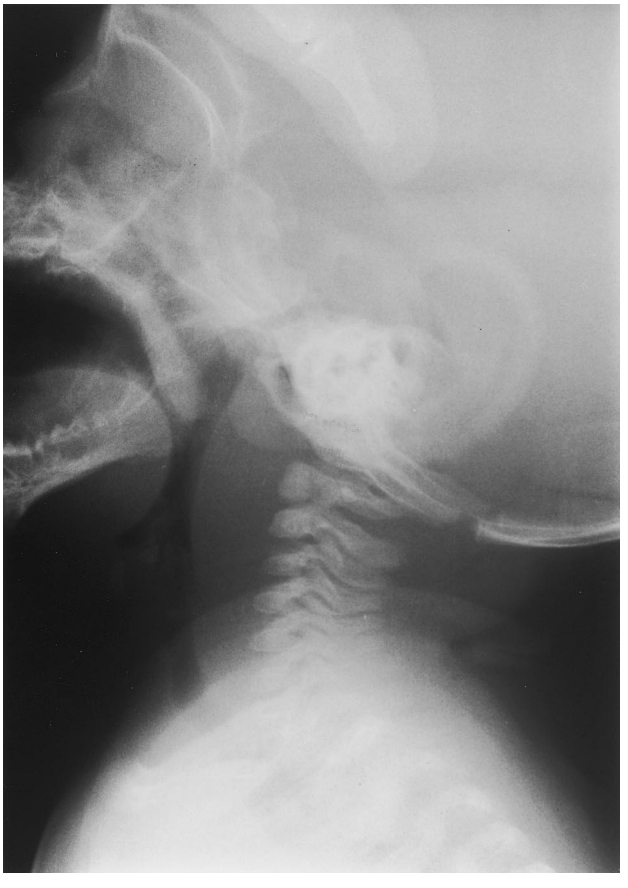

Fig 1. Lateral neck radiograph demonstrating widening of the retropharyngeal soft tissues.

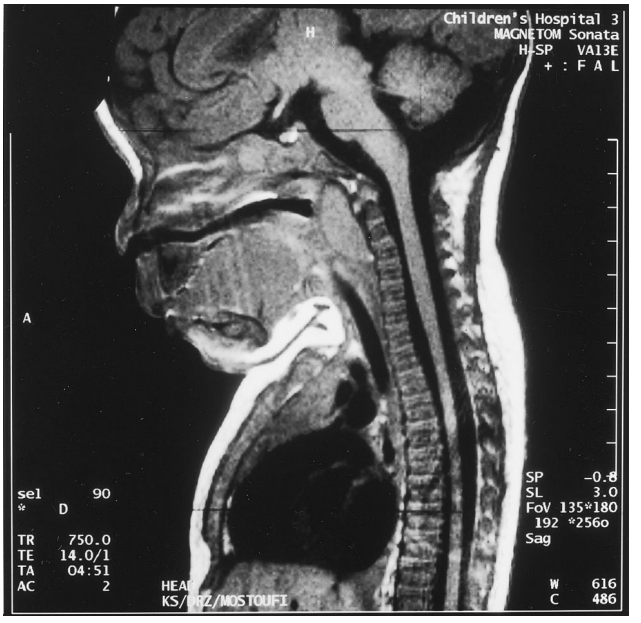

Fig 2. MRI scan (sagittal T_1-weighted image) of the neck demonstrating the retropharyngeal mass and the presence of normal mediastinal thymus.

DISCUSSION

Upper airway obstruction from a retropharyngeal mass requires urgent evaluation. In children, the differential diagnosis includes infection, trauma, neoplasm, and congenital abnormalities.[1] Aberrant cervical thymic tissue, although occasionally observed on autopsy examination, is rarely clinically significant.

Aberrant cervical thymic tissue may be cystic or solid. Cystic cervical thymus is more common, and 6% of these patients present with symptoms of dyspnea or dysphagia.[2–5] Aberrant solid cervical thymus usually presents as an asymptomatic anterior neck mass.[6–8] This case is unusual in that solid thymic tissue was located in the retropharynx, a finding not previously reported in the English literature. Additionally, the patient presented in acute respiratory distress, and the diagnosis was confounded by the presence of mild laryngomalacia. In retrospect, our patient likely had symptoms of intermittent upper airway obstruction since birth. The acute respiratory distress at presentation was likely the result of laryngomalacia exacerbated by the presence of aberrant thymic tissue and a superimposed viral infection.

Aberrantly located thymic tissue arises as a consequence of migrational defects during thymic embryogenesis. The thymus is a paired organ derived from the third and, to a lesser extent, fourth pharyngeal pouches. After its appearance during the sixth week

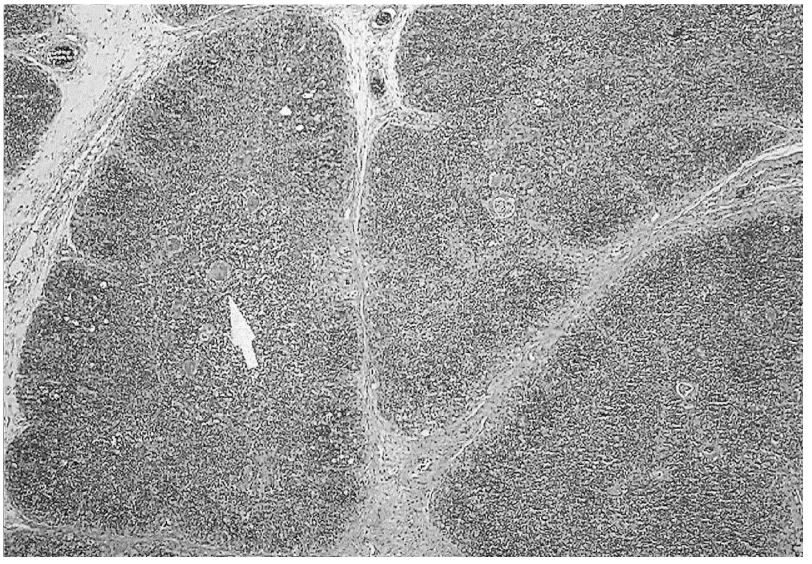

Fig 3. Microscopic section of resected mass demonstrating normal thymus. Small lymphocytes and Hassall's corpuscle (arrow) are seen (hematoxylin-eosin, ×25).

of fetal life, it descends to a final position in the anterior mediastinum, adjacent to the parietal pericardium. Aberrant thymic tissue results when this tissue breaks free from the thymus as it migrates caudally. Therefore, aberrant thymic tissue may be found in any position along a line from the angle of the mandible to the sternal notch, and in the anterior mediastinum to the level of the diaphragm.[9] In an autopsy study of 3236 children, abnormally positioned thymic tissue was found in 34 cases (1%). The aberrant thymus was most often located near the thyroid gland ($n = 19$ cases) but was also detected lower in the anterior neck ($n = 6$ cases), higher in the anterior neck ($n = 8$ cases), and at the left base of the skull ($n = 1$ case).[10] The presence of thymic tissue in the retropharyngeal space in our patient is more unusual given the typical embryologic origin and descent of the thymus in the anterior neck to the mediastinum.

Children with aberrant thymus may have associated anomalies. Twenty-four (71%) of 34 children with aberrant thymus detected at autopsy had features consistent with DiGeorge syndrome, and only 5 of the remaining 10 patients had a normal mediastinal thymus present.[10] Our patient had normal serum calcium levels postexcision and a mediastinal thymus was visualized on MRI.

Biospy is required for diagnosis of cervical thymus and should also be considered to exclude other causes.[3,11] MRI is helpful in delineating the presence, position, and extent of thymic tissue. A normal thymus in T_1-weighted images has intermediate signal intensity that becomes brighter in T_2-weighted images. Immunologic sequelae or recurrence after resection of an aberrant cervical thymus has not been reported.

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