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J K CurÖ©, E P Tagge, M S Richardson and D M Mulvihill

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MR of Cystic Aberrant Cervical Thymus

Joel K. Cure, Edward P. Tagge, Mary S. Richardson, and Denise M. Mulvihill

Summary: The clinical, MR imaging, surgical, and histologic findings in two cases of cystic aberrant cervical thymus were reviewed. Aberrant cervical thymic tissue was sharply circumscribed and extended deep to the posterior pharyngeal wall at the level of the piriform sinus in both cases. Both lesions had cyst contents that were of increased signal intensity on T1-weighted images. Solid components displayed MR signal characteristics identical to normal thymus.

Index terms: Neck, abnormalities and anomalies; Pediatric neuroradiology

Most pediatric neck masses are inflammatory and resolve with conservative therapy. In clinically atypical lesions, imaging studies can provide useful clues regarding the nature and prognosis of the mass. Magnetic resonance (MR) imaging is very useful in the evaluation of pediatric neck masses (1). This case report reviews the MR imaging findings in two children who had surgically proved aberrant cervical thymic tissue.

Case Reports

Case 1

A 6-year-old girl had a nontender mass on the right side of the neck below the angle of the mandible. Ultrasound examination showed an echogenic complex cystic and solid lesion extending deep into the neck. No enlarged vessels were identified supplying the mass, and no vascular flow was evident within the lesion on color flow or spectral Doppler examination. MR showed a mixed cystic and solid lesion extending from behind the right pharyngeal wall at the level of the piriform sinus laterally, anterior to the common carotid artery, and deep to the right sternocleidomastoid muscle (Fig 1). The mass extended inferiorly to the level of the thyroid gland. Cysts within the lesion were bright on both T1-(Fig 1A) and T2-(Fig 1B) weighted images. Solid portions of the lesion were slightly hyperintense relative to muscle on T1-, proton density- and T2-weighted images. Postcontrast images (Fig 1C) showed mild enhancement of the cyst walls and solid components of the mass. At operation, the superficial portion of the mass was encountered just deep to the platysma. The abnormality did not pass through the pharyngeal musculature or the thyrohyoid membrane. The posterior aspect was wrapped around the vagus nerve. Once freed from the vagus nerve, the mixed cystic and solid butterfly-shaped mass was removed; it measured 5.5 × 2.5 × 1.3 cm. Cut cystic components contained serosanguineous fluid. Microscopic examination revealed epithelialoid histiocytes surrounded by lymphocytes, nonkeratinizing squamous cell-lined cysts, and scattered Hassall corpuscles (onion skin–like aggregates of epithelial cells) within solid portions of the lesion. Numerous cholesterol granulomas were noted (Fig 1D), many lying along the margins of the cysts. The pathologic diagnosis was benign thymic tissue.

Case 2

An otherwise healthy 7-year-old boy had a nontender, asymptomatic mass on the left side of the neck that was visible only when he extended his neck. MR showed a cystic lesion of bright signal intensity on both T1- and T2-weighted images (Fig 2A and B). The lesion extended from behind the left side of the pharynx at the level of the left piriform sinus laterally, anterior to the neurovascular bundle, and inferiorly to the level of the thyroid gland. Most of its bulk lay anteromedial to the left sternocleidomastoid muscle; it did not penetrate the pharyngeal muscles or thyrohyoid membrane. At surgery, a sharply circumscribed 5.5 × 5.4 × 1.5-cm cystic mass was removed. The mass contained a 2.5-cm cyst filled with yellow fluid. Microscopic examination of the lesion revealed multiple large cysts lined by a thin layer of flat epithelium. Cholesterol granulomas and Hassall corpuscles (Fig 2C) were noted in the walls of the cysts. The pathologic diagnosis was thymic cyst.
Discussion

Cervical masses of thymic origin (aberrant cervical thymus) are uncommon. Two thirds of the cases are seen in the first decade of life, whereas the remainder manifest in the second and third decades (2). Lesions can be located anywhere from the angle of the mandible to the thoracic inlet, though most are found at the level of the thyroid gland, anterior to the carotid artery (3). Patients with aberrant cervical thymic tissue are usually asymptomatic. However, lesions can rapidly enlarge after an upper respiratory tract infection or after intralesional hemorrhage or infection (4). Rapidly enlarging lesions occasionally produce dyspnea and stridor because of airway compression (5–7).

Aberrant cervical thymic tissue arises along the pathway of thymic descent from the embryonic pharyngeal wall to the anterior mediastinum. The thymus is derived from the ventral cell mass of the third pharyngeal pouch. Its primordia appear and migrate into the chest during the sixth through eighth weeks of gestation. The cell mass initially hollows and elongates caudally and medially, forming the thy-mopharyngeal “tracts” or “ducts.” Epithelial proliferation soon obliterates the hollow tubes, and the bilateral solid thymic primordia meet in the midline below the thyroid gland and descend into the anterior mediastinum, coming to lie anterior to the parietal pericardium. This pathway can be traced on cross-sectional im-

Fig 1. A, Axial T1-weighted MR image (466/12/2 [repetition time/echo time/exci-
tations]) shows lesion deep to larynx, extending laterally, anterior to the right common
carotid artery. Note hyperintense cystic components (white arrowheads) and solid
portion (black arrowhead), which was slightly hyperintense relative to muscle.
B, Axial T2-weighted, fat-suppressed fast spin-echo image (4700/98/1, echo train
length, 8), same location as A. Cystic components (arrowheads) are hyperintense. Solid
component (arrow) is slightly more intense than muscle.
C, T1-weighted, fat-suppressed image (716/12/2) after administration of gado-
pentetate dimeglumine shows mild enhancement of the solid component (white arrow-
head) and cyst walls (black arrowheads).
D, Photomicrograph of thymic cyst specimen (hematoxylin-eosin, ×40) shows a
multiloculated cyst with two mural cholesterol granulomas (arrowheads), seen within
thymic tissue. The darker staining tissue at the bottom of the image represents lymphoid
and epithelial components of the thymic tissue. More superiorly lies paler staining
connective tissue septa surrounding cysts within the thymic tissue.
ages from the piriform sinus through the thyrohyoid membrane, between the common carotid artery and vagus nerve, behind the glossopharyngeal nerve, lateral to the thyroid gland, and inferomedially into the anterior mediastinum (8). Aberrant cervical thymic tissue is the result of partial or complete failure of descent of the thymic primordia into the mediastinum. As many as 50% of cases of cervical thymic tissue are attached to mediastinal thymus, either via contiguous thymic tissue or a fibrous cord (9). Aberrant cervical thymic tissue can be cystic, solid, or mixed. Most lesions are cystic, are more frequently lateral than in the midline, and occur with equal frequency on the left and right sides (8). Cystic aberrant thymus can result from remnants of the thymopharyngeal ducts, from degeneration of Hassell corpuscles (which are thymopharyngeal duct derivatives), or from cystic degeneration of solid thymic tissue caused by infection or tumor (8, 10). It can be impossible to differentiate remnants of the thymopharyngeal duct lacking solid thymic tissue from third branchial cleft cysts (11). Histologically, aberrant cervical thymic tissue contains thymic lobules, lymphoid follicles, Hassell corpuscles, and cysts. The cysts can be small or quite large and are lined by a variety of cell types, including nonkeratinizing squamous, cuboidal, or columnar cells. Cyst contents range from straw-colored proteinaceous fluid to necrotic debris or old hemorrhage. Cholesterol crystals may be found in the luminal fluid or embedded in the cyst wall. Cholesterol granulomas are a frequent finding (9, 10, 12). Occasionally parathyroid tissue (also derived from the third pharyngeal pouch) is found in the lesions (4, 11).

Neoplasms have been reported in solid aberrant cervical thymic tissue, though not in cervical thymic cysts; solid cervical thymic tissue should be removed. Although there is a theoretical risk of immune deficiency if all functioning thymic tissue is removed with an aberrant cervical thymus (13), this complication has never been documented. Nonetheless, given the relatively small numbers of cases reported, some authors recommend only partial resection of cervical thymus when mediastinal thymus cannot be confirmed (4, 14). In our two cases, the upper part of the chest was not imaged; thus, mediastinal thymus was not confirmed. Fortunately, both children remain immunologically healthy.

In children, normal thymic tissue is homogeneous and slightly more intense than muscle on T1-weighted MR images and slightly less intense or isointense relative to fat on T2-weighted images (15). The MR appearance of solid cervical thymic tissue parallels that of mediastinal thymus (case 1). Cystic components of aberrant cervical thymus in our two cases were bright on T1-weighted images. Mediastinal thymic cysts are usually hypointense on T1-
weighted images and bright on T2-weighted images. If hemorrhagic, the cyst contents can be hyperintense on T1-weighted images because of the T1 shortening effect of methemoglobin (16). Lesional hyperintensity on T1-weighted images can also reflect high cyst protein content or cholesterol granulomas. Bright signal intensity has been noted in middle ear (17) and petrous apex (18) cholesterol granulomas on T1-weighted MR images.

The differential diagnosis of cystic aberrant cervical thymus on MR includes simple or complicated (hemorrhagic or infected) third branchial cleft cysts, lymphangiomas, venous malformations, necrotic or suppurative lymphadenopathy, thyroglossal duct cysts, and epidermoid and dermoid cysts. The possibility of aberrant cervical thymus should be strongly considered when children have cystic or mixed cystic and solid lesions lying behind the posterior wall of the pharynx, or elsewhere along the pathway of thymic descent into the chest. This location and extent is not typical of most other diagnostic possibilities except third branchial cleft cysts or lymphangiomas, which may be impossible to exclude. Blood-fluid levels, if present, would be more suggestive of a lymphangioma. Solid-appearing components would not be typical of uncomplicated third branchial cleft cysts. Once the diagnosis of aberrant cervical thymus is considered, the presence of mediastinal thymic tissue can be confirmed preoperatively, avoiding the potential risk of iatrogenically compromised immunity.

References

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