

Castleman's Disease in the Retropharyngeal Space: CT and MR Imaging Findings

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Summary: We report CT and MR imaging findings in a case of Castleman's disease involving the retropharyngeal space in a middle-aged woman. On CT scans, a well-margined, homogeneous, and densely enhancing mass was detected in the right retropharyngeal space. The mass was isointense to the muscle on T1-weighted MR images, hyperintense to the muscle on T2-weighted MR images, and showed homogeneous, strong enhancement on contrast-enhanced T1-weighted MR images. The linear hypointense signal in an arborizing pattern was observed within the mass on all pulse sequences.

Castleman's disease is a rare lymphoproliferative disorder of uncertain cause characterized by a distinctive pattern of hypervascular lymphoid hyperplasia (1). Although it is usually reported as a solitary mediastinal mass, involvement of other anatomic sites has been reported, with the head and neck being the second most common area (2). We describe a case of hyaline-vascular-type Castleman's disease located in the retropharyngeal space.

Case Report

A 34-year-old woman initially presented with a 5-year history of painless swelling on the right side of her oropharynx and a 2-year history of a slightly enlarging mass on the right side of her neck. Her medical history was unremarkable.

Unenhanced CT scans showed a large, well-circumscribed, solid mass in the right retropharyngeal space, which was isodense to the muscle. The mass showed homogeneous, strong enhancement on the contrast-enhanced CT scans (Fig 1A).

One month later, MR imaging was performed. On the MR images, the mass was located within the right retropharyngeal space, displacing the parapharyngeal fat anteriorly, the pharyngeal mucosal space medially, the styloid process anterolaterally, and the carotid space laterally. It was isointense to the muscle on the T1-weighted images (Fig 1B). There was a non-enhancing linear hypointense signal in an arborizing pattern within the mass on T1- and T2-weighted images (Fig 1C and D). The mass occupied the right retropharyngeal and para-

pharyngeal space from below the skull base to the hyoid bone level on the contrast-enhanced T1-weighted coronal image (Fig 1E). There was a small lymph node with a 1-cm diameter in the right internal jugular chain just below the main mass.

Excision of the neck mass was performed via a transcervical approach. A 5 × 8-cm, well-encapsulated, ovoid mass was removed without much difficulty. Histopathologic evaluation revealed Castleman's disease (giant lymph node hyperplasia) of the hyaline-vascular type (Fig 1F). The mass was thought to arise from the lateral retropharyngeal lymph node. The small lymph node just below the main mass was proved reactive hyperplasia without tumor cells.

Discussion

In 1956, Castleman et al (3) described a group of patients with large thymoma-like masses in the anterior mediastinum, which they called *mediastinal lymph node hyperplasia*. The cause of Castleman's disease is uncertain; it is thought to be inflammatory or hamartomatous in nature (4, 5). Two distinct histologic variants are recognized (4). The most common is the hyaline-vascular type (more than 90% of the cases), which consists of small lymphoreticular follicles distributed within a hypervascular hyalinized stroma. The plasma cell type is less common (fewer than 10% of the cases) and consists of larger lymphoreticular nodules that are separated by sheets of plasma cells and a somewhat less vascular stroma. Patients with the hyaline-vascular-type disease are usually asymptomatic, but they may complain of symptoms caused by the compression of adjacent structures or may present with a palpable mass. Systemic manifestations are commonly seen in the plasma cell type and include fever, anemia, and hyperglobulinemia.

Castleman's disease is usually limited to one site, although an aggressive multicentric form has been described. The most common site is the mediastinum (approximately 70%). Additional sites of occurrence include the axilla, retroperitoneum, mesentery, vulva, pancreas, pelvis, and neck (1, 4). Fewer than 10% of cases arise in the head and neck (6-11). In our case, the tumor was located within the right retropharyngeal space. It was thought to arise from a retropharyngeal node, probably the lateral group, which is located at the anterolateral aspect of the prevertebral muscle, although a carotid space origin could not be completely excluded. Several cases of Castleman's disease involving the retropharyngeal or parapharyngeal space, medial to

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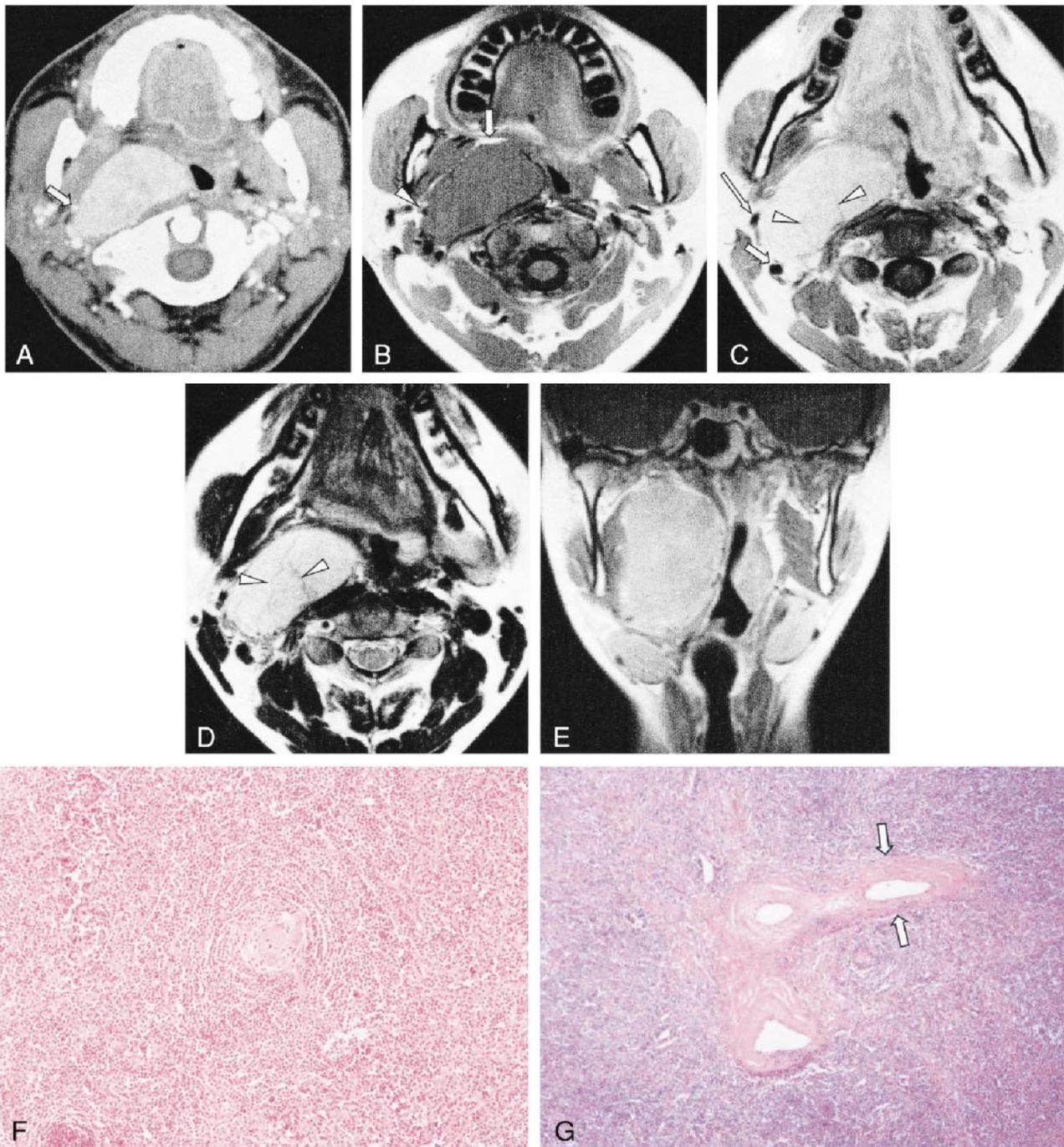


FIG 1. Images from the case of a 34-year-old woman with an enlarging mass on the right side of her neck.

A, Contrast-enhanced axial CT scan shows a well-marginated, intensely enhancing mass in the right retropharyngeal space. Note the styloid process, which is anterolaterally displaced by the mass (*arrow*).

B, T1-weighted (450/12) axial MR image shows a well-marginated mass isointense to the muscle in the right retropharyngeal space. The mass displaces the parapharyngeal fat anteriorly (*arrowhead*), and internal carotid artery laterally.

C, Contrast-enhanced T1-weighted (450/12) axial MR image (obtained at a slightly lower level than that shown in panel B) reveals strong enhancement of the lesion. The displaced right internal carotid artery (*short arrow*) and external carotid artery (*long arrow*) are separated. Note the linear hypointense signal within the mass (*arrowheads*).

D, T2-weighted (3700/99) axial MR image (obtained at the same level as that shown in panel C) shows the lesion hyperintense to the muscle. The linear hypointense signal seen in panel C is also seen as a hypointense signal (*arrowheads*).

E, Contrast-enhanced T1-weighted (513/12) coronal MR image shows the mass occupying the entire right retropharyngeal and parapharyngeal space from below the skull base to the hyoid bone level.

F, Photomicrograph (original magnification, $\times 100$; hematoxylin and eosin stain) of the right retropharyngeal lymph node shows a follicle with an onion skin–appearing, prominent mantle and a penetrating hyalinizing vessel into the germinal center, consistent with hyaline-vascular-type Castleman's disease.

G, Low-power photomicrograph (original magnification, $\times 40$; hematoxylin and eosin stain) of the right retropharyngeal lymph node shows perivascular lamellar fibrosis (*arrows*), which corresponds to the linear hypointense signals in an arborizing pattern on the MR images.

the carotid sheath, as in our case, have been reported (7–9).

The typical CT findings of Castleman's disease are a well-marginated, homogeneous, and densely enhancing mass (6, 7), although ring enhancement can also be seen (1, 10). The hyaline-vascular variant tends to show more prominent enhancement on CT scans, most likely because of its greater vascularity (1, 10).

Typically, Castleman's disease appears on MR images as low-to-intermediate signals compared with muscle on T1-weighted images and high signals on T2-weighted images (7, 9). In a few previous reports (12, 13), linear, arborizing, hypointense signals within the mass have been attributed to calcification, fibrous septations, vessels, or sinus histiocytosis. The linear hypointense signals seen on MR images in our case were proved perivascular lamellar fibrosis on pathologic correlation (Fig 1G). These linear hypointense signals could be an important clue to the diagnosis of Castleman's disease, although they occur infrequently.

The MR imaging differential diagnoses for Castleman's disease include paraganglioma and schwannoma. Paragangliomas in this area usually displace the internal carotid artery anteriorly because the tumor arises around the vagus nerve. Furthermore, paragangliomas have multiple focal and serpentine areas of low signal intensity, representing vascular flow voids within the mass, especially in large lesions. Schwannomas that arise from the vagus nerve and the sympathetic chain usually displace the internal carotid artery anteriorly because these nerves are posterior to this vessel. Frequently, schwannomas have inhomogeneous signal intensity due to cystic change. These features could be the clue to the differential diagnosis, with Castleman's disease in the retropharyngeal or parapharyngeal space.

Complete surgical excision is the treatment of choice for Castleman's disease in the head and neck, with a 100% control rate for the hyaline-vascular type. The plasma cell type, however, requires excision with close follow-up and possible systemic chemotherapy (14). Malignant transformation or association with other malignancies is rare and includes plasmacytoma, malignant lymphoma, and Kaposi's sarcoma (15). Its malignant potential, although rare, raises suggestions of a relationship with an immune dysfunction or even primary lymphoproliferative disorder (16).

Conclusion

Castleman's disease is a rare mass, which may involve the retropharyngeal space. On CT scans, it appears as a well-marginated, homogeneously enhancing mass. It has an isointense signal compared with muscle on T1-weighted MR images and a hyperintense signal compared with muscle on T2-weighted MR images, with homogeneous, strong enhancement. Linear hypointense signals in an arborizing pattern may be present within the mass. Castleman's disease should be considered in the differential diagnosis of masses involving the retropharyngeal space.

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