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Carcinoma Showing Thymiclike Differentiation (CASTLE Tumor)

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Summary: A 67-year-old woman had had a neck mass for 10 years, which recently increased in size. Sonographic, CT, and MR examinations showed a mass in the carotid and posterior spaces (triangle) extending from below the submandibular gland to the supraclavicular fossa, displacing the common carotid artery and the sternomastoid anteriorly. The mass was solid, noncalcified with lobulated outlines, hypoechoic on sonograms, of soft-tissue density on CT scans, isointense on T1-weighted MR images, hyperintense on T2-weighted MR images, and enhanced mildly after injection of contrast material on CT and MR studies. Histologic examination revealed a carcinoma showing thymiclike differentiation, a rare tumor of the neck and thyroid gland.

Carcinoma showing thymiclike differentiation (CASTLE tumor) is a rare tumor occurring in the soft tissues of the neck and thyroid gland. We describe the sonographic, CT, and MR imaging appearance of this rare tumor and discuss the differential diagnosis.

Case Report

A 67-year-old woman had a painless mass in the left side of the neck that recently had increased rapidly in size. Ten years earlier, she had had surgery at the same site for a mass of unknown origin. Physical examination revealed a 6-cm hard mass in the left side of the neck attached to the underlying structures; the overlying skin was ulcerated (Fig 1A). Multiple lymph nodes were palpable adjacent to the mass. Flexible endoscopy revealed no lesion in the pharynx, larynx, or nasopharynx.

At sonography, the tumor was entirely separate from the thyroid, posterior to the neurovascular bundle, extending from just below the left submandibular gland to the left supraclavicular fossa. The carotid artery and the sternomastoid were displaced anteriorly, the internal jugular vein could not be identified. The tumor had a lobulated outline (Fig 1B), was solid, hypoechoic with a heterogeneous echo pattern (Fig 1C), and of moderate vascularity on color flow images. On CT scans, the tumor was of soft-tissue density, showed no evidence of calcification, and enhanced slightly after administration of contrast material (Fig 1D and E). The internal jugular vein was not opacified and lymph nodes were seen in the left supraclavicular fossa. There was a clear plane between the tumor and the left lobe of the thyroid (Fig 1F). MR imaging confirmed the extent

(Fig 1G) and the extrathyroid nature of the tumor, which measured $5 \times 7 \times 9$ cm in diameter. The left internal jugular vein was not identified but the carotid artery showed a normal flow void. The mass was isointense on T1-weighted sequences, enhanced only moderately (Fig 1H and I) with a central, irregular nonenhancing area after injection of contrast material, and was hyperintense on T2-weighted images (Fig 1J and K).

Fine-needle aspiration cytology (FNAC) suggested the diagnosis of a spindle cell tumor. The incisional biopsy showed a mitotically active, histologically malignant tumor with polygonal and spindle cells that were positive for vimentin and epithelial membrane antigen but negative for CAM5.2, AE1, AE3, S100, desmin, smooth muscle actin, synaptophysin, chromogranin, thyroglobulin, and calcitonin. Electron microscopy showed well-developed desmosomes. The incisional biopsy specimen was reported as undifferentiated sarcoma, possibly poorly differentiated synovial sarcoma. At surgery a 9-cm, solid, lobulated tumor was seen closely attached to the carotid sheath. The carotid artery was patent and displaced anteriorly, and the internal jugular vein was compressed; however, there was no thrombus. The mass was separate from the thyroid, adherent to the sternomastoid, and infiltrated the overlying subcutaneous tissues and skin. Multiple enlarged nodes were seen along the internal jugular vein, in the supraclavicular fossa, and behind the clavicle. The mass and the nodes along the internal jugular vein were removed but the nodes behind the clavicle were left behind.

Microscopically, most of the tumor resembled the initial biopsy specimen except that some areas of necrosis were visible and the tumor was partly subdivided by scattered, thick, fibrous trabeculae such as seen in thymomas. In one block, the tumor was differentiated and resembled a spindle cell thymoma. In two other blocks, there were numerous lymphocytes with very few spindle cells, suggesting a lymphocytic thymoma. The lymphocytes were positive for T-cell markers and negative for B-cell markers and they were positive for MIC 2 (CD99), a feature of thymic carcinomas. The final histologic analysis reported the mass to be a CASTLE tumor. The surgical specimen contained 22 nodes; only one showed tumor involvement, all the others were enlarged reactive nodes. In view of the nodal involvement, the patient received radiotherapy after surgery.

Discussion

A number of rare tumors occurring in the soft tissues of the neck and thyroid gland show complete to partial histologic resemblance to the fetal, mature, or involuted thymus and thymomas (1–6). These have

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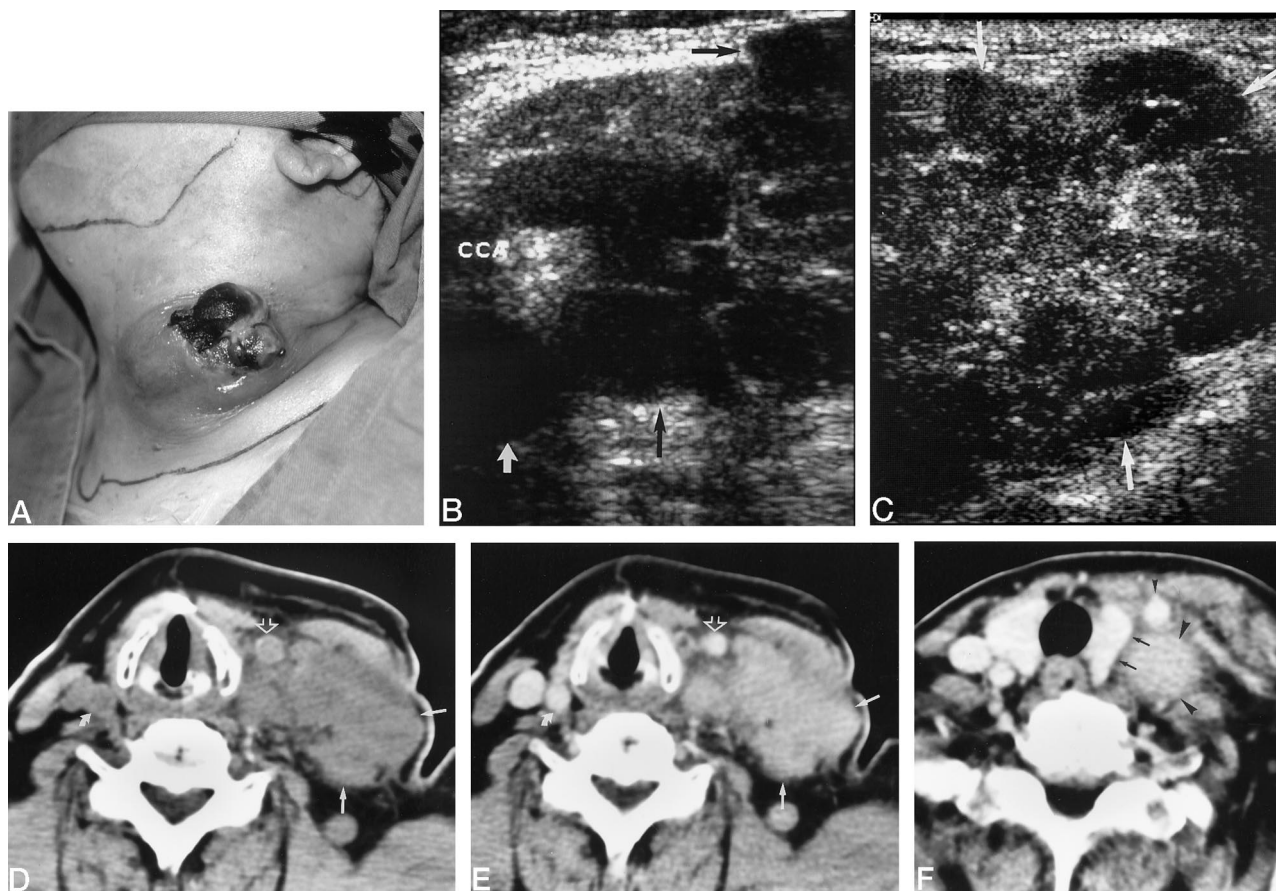


FIG 1. 67-year-old woman with a mass in the left side of the neck that recently had increased rapidly in size.

A, Photograph shows lobulated mass with skin ulceration.

B, Transverse sonogram shows a lobulated, hypoechoic mass (black arrow) in close association with the common carotid artery (CCA). Note the tumor extension behind the CCA (white arrow).

C, Longitudinal sonogram shows a lobulated, solid, noncalcified mass with heterogeneous internal echoes (arrow).

D and E, Plain (D) and contrast-enhanced (E) CT scans show a noncalcified mass (closed straight arrows) with moderate contrast enhancement. Note the anterior displacement of the left CCA (open arrow) as compared with the right CCA (curved arrow).

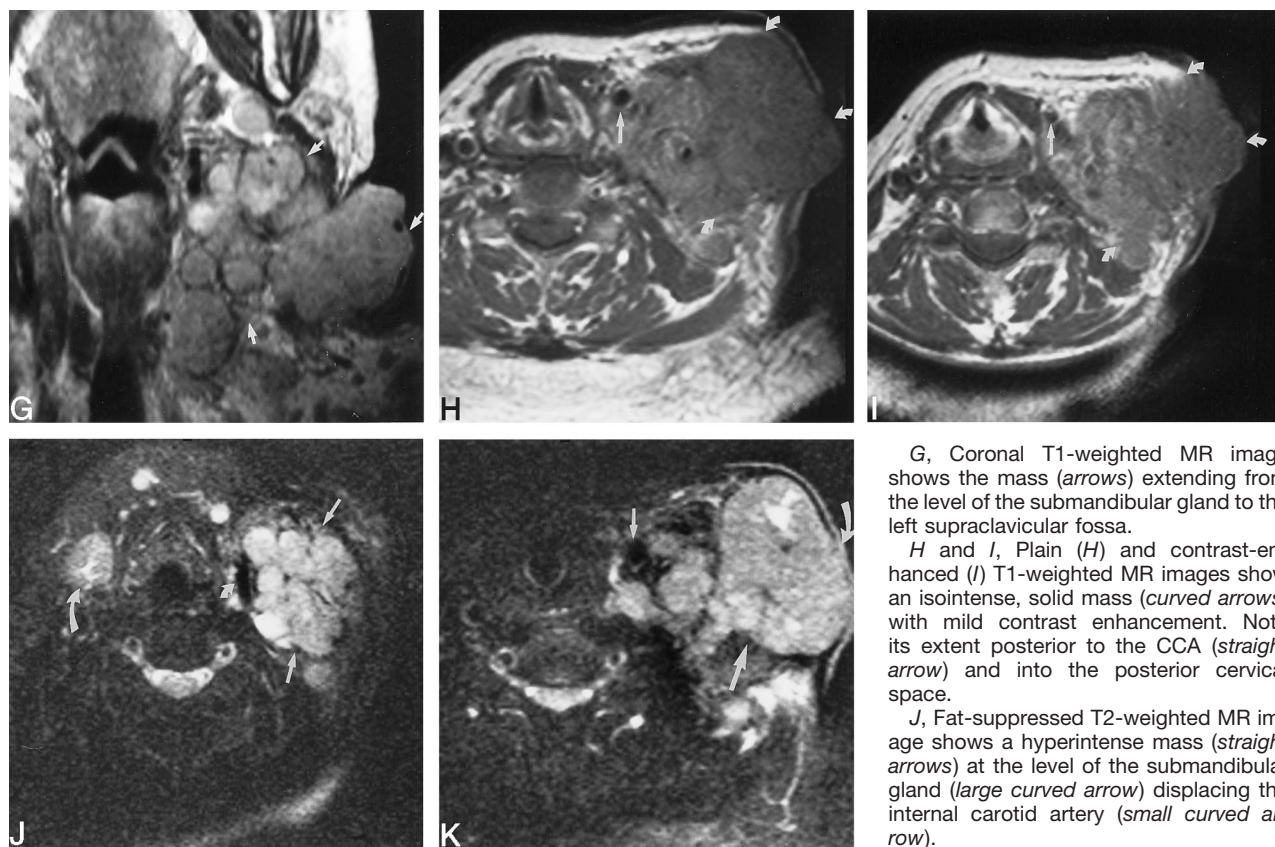
F, Contrast-enhanced CT scan shows the mass (large arrowheads) posterior to the CCA (small arrowhead) with a normal plane between the mass and the thyroid (arrows).

been reported in the literature under a variety of terms, and their histologic appearance has been described as ranging from completely benign to metastatic malignant. Chan et al (7) classified them into four groups on the basis of their morphologic features: ectopic hamartomatous thymomas, ectopic cervical thymomas, spindle epithelial tumors with thymiclike differentiation (SETTLE tumors), and CASTLE tumors.

Tumors of the CASTLE type are histologically similar to thymic carcinoma of the lymphoepithelioma or squamous cell variety. It has been postulated that these tumors arise either from ectopic thymus or remnants of branchial pouches, which retain the potential to differentiate along the thymic line (7). It is generally an indolent tumor that can recur after long intervals; regional nodal metastasis occurs in approximately half the cases, and, occasionally, tumors may pursue a more aggressive course (7). Chan et al (7) reviewed the clinical and pathologic features of 11 such tumors, nine from the thyroid and two arising from the soft tissues of the neck. We have since found

a few case reports describing this tumor, all of which arose from the thyroid gland (8–11).

In our case, because the mass was large, its exact origin in the neck was difficult to establish. It involved the carotid space and the posterior cervical space and had a long evolutionary period with a recent increase in size. The differential diagnosis therefore included lymphadenopathy, schwannoma, neurofibroma, and paraganglioma of the carotid bifurcation or carotid body tumor. Malignant nodes on sonograms are usually hypoechoic and round, often show a loss of normal echogenic hilus, may exhibit intranodal necrosis and lobulation, and may be accompanied by satellite lesions. On CT and MR studies, they may show heterogeneity of the internal architecture and rim enhancement after contrast administration. Abnormal nodes along the carotid sheath are usually anterior or anterolateral to the major vessels rather than posterior to them. Despite the presence of nodes in the supraclavicular fossa, in view of the long history and imaging features, we thought that the mass was unlikely to be matted adenopathy. Schwannomas and



K, Fat-suppressed T2-weighted MR image shows a hyperintense mass (large straight arrow) with skin and subcutaneous involvement (curved arrow). Small straight arrow identifies the CCA.

neurofibromas may have lobulated outlines and are hypoechoic on sonograms, but they usually show uniform enhancement on CT and MR studies. FNAC of these tumors may also produce spindle cells. However, neither of these tumors are normally associated with lymphadenopathy unless associated with a malignant change. The absence of enhancement on CT and MR examinations suggested the mass was unlikely to represent a malignant change in a schwannoma or neurofibroma. Tumors of the carotid body are centered on and splay the carotid bifurcation, are hypoechoic on sonograms, vascular on color flow images, and show intense enhancement on CT and MR studies (12). The location and the absence of enhancement ruled out a carotid body tumor. At this point, although we were uncertain about the nature of the mass, we knew we were dealing with an unusual tumor, which was identified as a CASTLE type lesion after surgery.

At histology, the differential diagnosis included primary undifferentiated or squamous cell thyroid carcinoma (4) and metastatic carcinoma from an unknown source; particularly, the upper aerodigestive tract, lung, and mediastinum (13). The distinction is important, since CASTLE tumors are generally indolent whereas the others are usually fatal. Treatment consists of surgical excision with or without radiotherapy (7).

Conclusion

Given their rarity and nonspecific imaging appearance, CASTLE tumors are generally diagnosed only after the common lesions discussed above have been ruled out. The radiologist must be aware of this entity and should consider it in the differential diagnosis of a long-standing mass that has lobulated outlines and is located along the course of the descent of the thymus (from the angle of the mandible to the sternum).

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