Radiologic-Pathologic Correlation of Unusual Lingual Masses: Part I: Congenital Lesions

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Because the tongue is superficially located and the intial manifestation of most diseases occurring there is mucosal change, lingual these lesions can be easily accessed and diagnosed without imaging analysis. Most congenital lesions of the tongue, however, can manifest as a submucosal bulge and be located in a deep portion of that organ such as its base; their true characteristics and extent may be recognized only on cross-sectional images such as those obtained by CT or MRI. In addition, because it is usually difficult to differentiate congenital lesions from other submucosal neoplasms on the basis of imaging findings alone, clinical history and physical examination should always be taken into consideration when interpretating CT and MR images of the tongue.

Although the radiologic findings for congenital lesions are nonspecific, CT and MR imaging can play an important role in the diagnostic work-up of these unusual lesions. Delineation of the extent of the tumor, and recognition and understanding of the spectrum of imaging and the pathologic features of these lesions, often help narrow the differential diagnosis.

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Although the vast majority of lingual masses are squamous cell carcinomas, a variety of congenital lesions may affect the tongue. These usually manifest as a submucosal bulge and are located in a deep portion such as its base. Thus, the characteristics and extent of these lesions may be recognized only on cross-sectional CT or MR images.

In this article, we describe the imaging findings of congenital lingual masses, provide clinical and pathologic-radiologic correlation, and discuss the role of CT and MR imaging in the diagnostic work-up of these lesions.

Lingual thyroid

Embryologically, failure of the thyroid gland to separate from the base of the tongue results in a lingual thyroid, which accounts for 90% of ectopic thyroids. Remnant thyroid tissue can be demonstrated at the base of the tongue in up to 10% of autopsy specimens (1). Most cases are asymptomatic but symptomatic patients have dysphagia (50%), dysphonia (44%), or dyspnea (28%). Malignancy in lingual thyroid is rare.

Lingual thyroid has an absolutely characteristic CT appearance: its iodine content causes it to have very high attenuation values in relation to surrounding soft tissue. The mass of ectopic thyroid tissue usually lies in the center of the tongue base within intrinsic muscle; intravenous contrast enhancement will increase the differential density between thyroid tissue and surrounding musculature (Fig. 1). Rarely, a lingual thyroid has been found to show inhomogeneous contrast enhancement on CT, resulting from

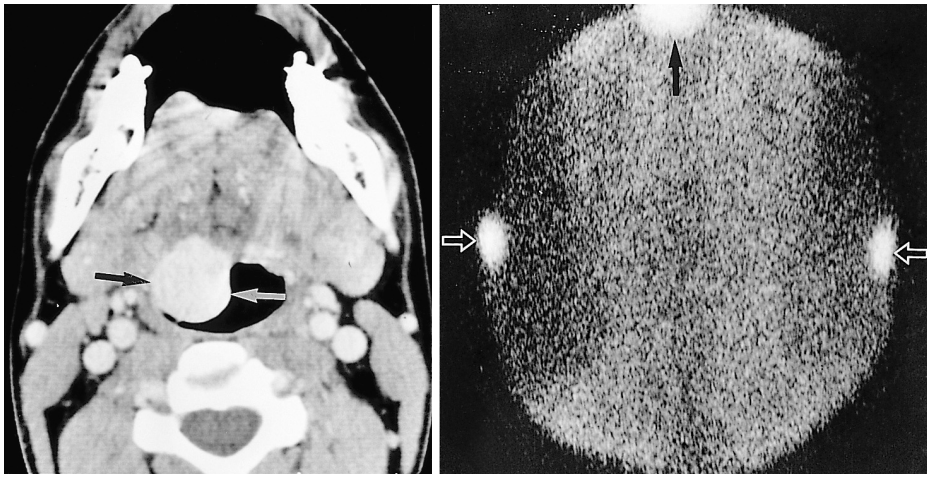


Fig. 1. Lingual thyroid in a 31-year-old man with mild dyspnea.
A. Contrast-enhanced axial CT scan shows a well-marginated, homogeneously enhancing mass without evidence of cystic change or calcification at the dorsal aspect of the off-midline tongue base (arrows). CT scan at the level of the lower neck failed to disclose normal thyroid tissue in the thyroid bed at the anterior aspect of the thyroid cartilage (not shown).
B. Anteroposterior ^{131}I scan shows round hot uptake at the center of the oropharynx, and this matches the location of the lesion revealed by CT (arrow). No uptake is seen in the normal thyroid bed indicated by isotope markers (open arrows). CT and scintigraphy can thus be used to diagnose lingual thyroid. No treatment was undertaken.

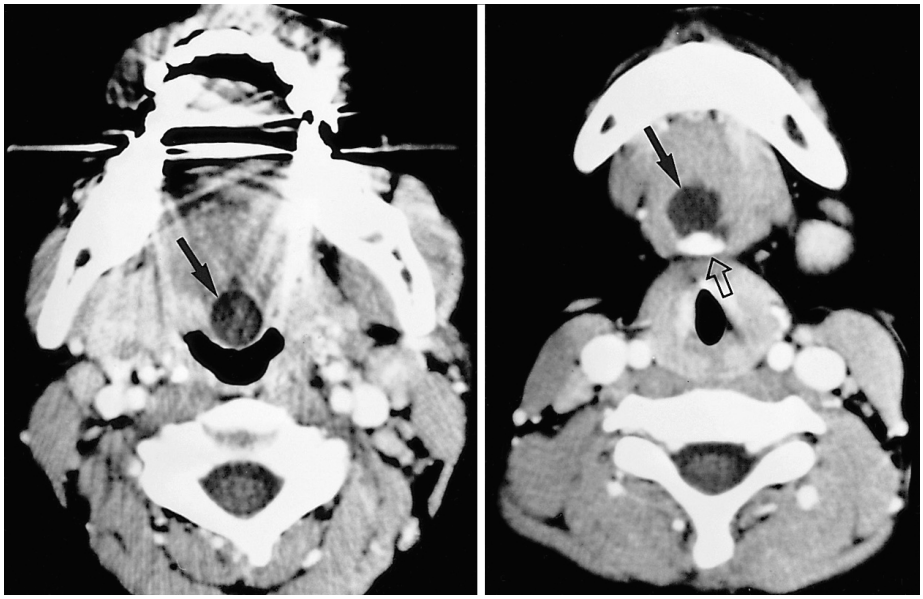
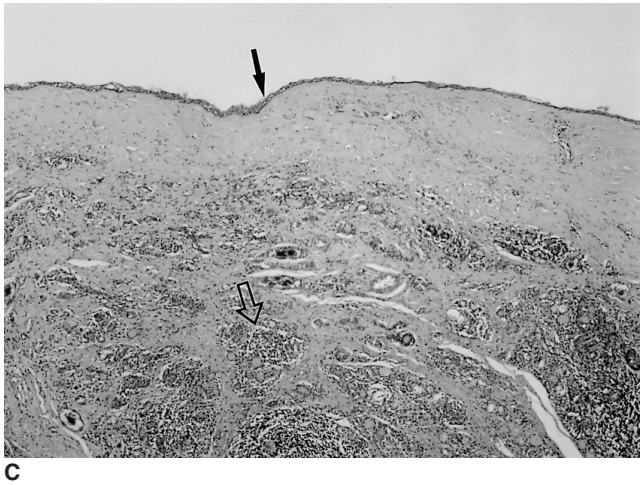


Fig. 2. Lingual thyroglossal duct cyst in a 2-year-old boy with stridor and dysphagia.
A. Contrast-enhanced axial CT scan shows a well-demarcated cyst in the region of the foramen cecum at the base of the tongue (arrow).
B. Axial CT scan 1.5cm caudal to A shows downward extension of the cyst to the level of the hyoid bone (open arrow) along the tract of the thyroglossal duct.
C. Photomicrograph (original magnification ×40; H & E staining) demonstrates remnant thyroid tissue (open arrow) and inflammatory cells, suggesting complication, lined by cuboidal to columnar epithelium (arrow).



marked thyroiditis and goitrous changes (2). The findings of radionuclide scanning are pathognomonic in the diagnosis of lingual thyroid. If the thyroid gland cannot be found in its usual location, radionuclide scanning is necessary to determine the size, location and activity of thyroid tissue. Because the radiation dose required is lower, the use of technetium-99m pertechnetate is preferred to iodine 131.

Lingual thyroglossal duct cyst

Thyroglossal duct cyst (TGDC) commonly presents as a midline anterior neck cyst at any point along the path from the foramen cecum of the tongue to the pyramidal lobe of the thyroid gland. It results from the persistence and dilata-

tion of remnants of an epithelial tract formed during migration of the thyroid during the period of embryogenesis (3). In 85% of cases, a TGDC is located below the hyoid bone. Lingual TGDC is a rare form, and only 2% of TGDCs are located at the base of the tongue (3). A TGDC usually contains thick, gelatinous mucoid fluid, and microscopically, may be lined with transitional, cuboidal, columnar or stratified squamous epithelium, and this may be ciliated or nonciliated. Depending on its oral pharyngeal location, lingual TGDC may cause dysphagia or dyspnea.

CT characteristically reveals a lingual TGDC as a well-circumscribed, homogeneous low-density lesion, the values of which correspond to those of fluid (10–18 HU) (Fig. 2). Elevated attenuation of the cystic fluid reflects increased protein content. Depending on this content, T1-weighted imaging of an uncomplicated TGDC demonstrates low to intermediate signal intensity, while T2-weighted images are hyperintense. Unless inflammation is present, the cyst wall is usually thin and non-enhancing (4).

Bronchogenic cyst

Bronchogenic cyst is the most common type of intrathoracic foregut cyst, and results from anomalous budding or branching of the tracheobronchial tree during development (5). For this cyst to be classified as bronchogenic in origin, it must be lined by columnar respiratory epithelium and contain seromucous glands. A congenital bronchogenic cyst of the tongue is extremely rare. Microscopically, it is lined by ciliated pseudostratified epithelium of the respiratory type, but in parts there may be patches of squamous metaplasia (Fig. 3). The cyst wall frequently contains bundles of smooth muscle, mucous and mixed glands, as well as cartilaginous nodules and plates.

CT usually demonstrates a cystic mass molding the surrounding structures, and the cyst may be uni- or multilocular (5). Unless complicated by infection or hemorrhage, the size of a bronchogenic cyst is usually stable, and the density of its contents is near that of water, ranging from −10 to +10 HU (Fig. 3).

Epidermoid cyst

True dermoid, epidermoid, and teratoid cyst comprise the spectrum of teratoma. The essential difference between dermoid and epidermoid cyst lies in the presence of skin appendages within the wall of the former and the absence of these features in the latter (6). Because of its squamous epithelial lining, an epidermoid cyst may have cheesy keratinaceous material within its lumen. When an epidermoid cyst occurs in the oral cavity, the floor of the mouth is most commonly involved, though sites which include the lips, tongue, and buccal mucosa have also been reported.

Epidermoid cyst is typically revealed by CT as a well-defined, lobulated mass with hypoattenuation similar to that of fluid. Occasionally, it may be hyperattenuating on unenhanced CT scans. Peripheral enhancement may be revealed by contrast-enhanced CT, but central enhancement

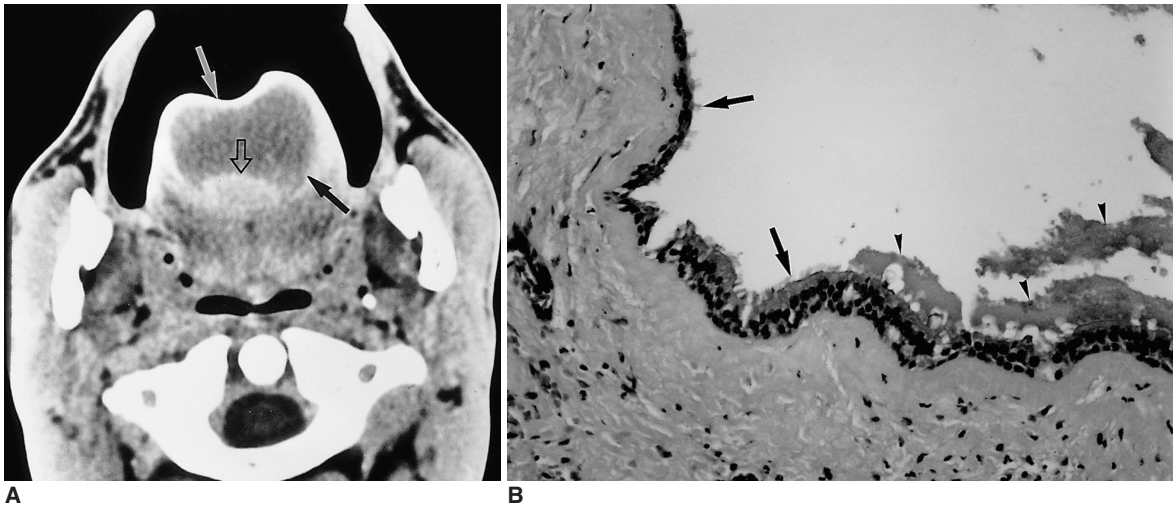


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Fig. 3. Bronchogenic cyst in a 19-year-old man with macroglossia since infancy.
A. Contrast-enhanced CT scan shows a large, lobulated, thin-walled cystic mass at the central portion of the tongue (black arrow). Note the submucosal location of this mass and the intact overlying mucosa (white arrow). The mass demonstrates homogeneous low attenuation but a high-attenuated solid component at the posterior aspect suggests mucin content (open arrow). The differential diagnosis included dermoid cyst and lymphangioma.
B. Photomicrograph (original magnification ×200; H & E staining) shows that the cyst is lined by ciliated pseudostratified epithelium of the respiratory type (arrows) and contains a small amount of mucin secretion (arrowheads).

within the tumor is unusual.

MR imaging of an epidermoid cyst typically depicts a cystic mass, hypointense on T1-weighted images and hyperintense on T2-weighted images (Fig. 4). The use of contrast material shows that the cyst wall is 2−6 mm thick (Fig. 4).

Venous malformation

Venous malformations (VMs) are part of a spectrum of benign vascular lesions commonly found in the pediatric population, and may occasionally be difficult to distinguish solely on the basis of their clinical appearance. On the basis of their histologic features, Mulliken and Glowacki classified congenital vascular anomalies as true hemangiomas or one of a variety of vascular malformations (7). The latter are true congenital lesions, which by definition are always present at birth, though are not always detected (8). They are classified according to the main channel type present: capillary, lymphatic, arterial, venous, or mixed (7). Vascular lesions have been further categorized on the basis of their hemodynamic characteristics as low- or high-flow types, a distinction that is important when choosing the management strategy. Whereas true hemangiomas often involute with age, vascular malformations remain stable or slowly grow with the patient and typically require some form of therapy when cosmetic disfigurement, bleeding, or

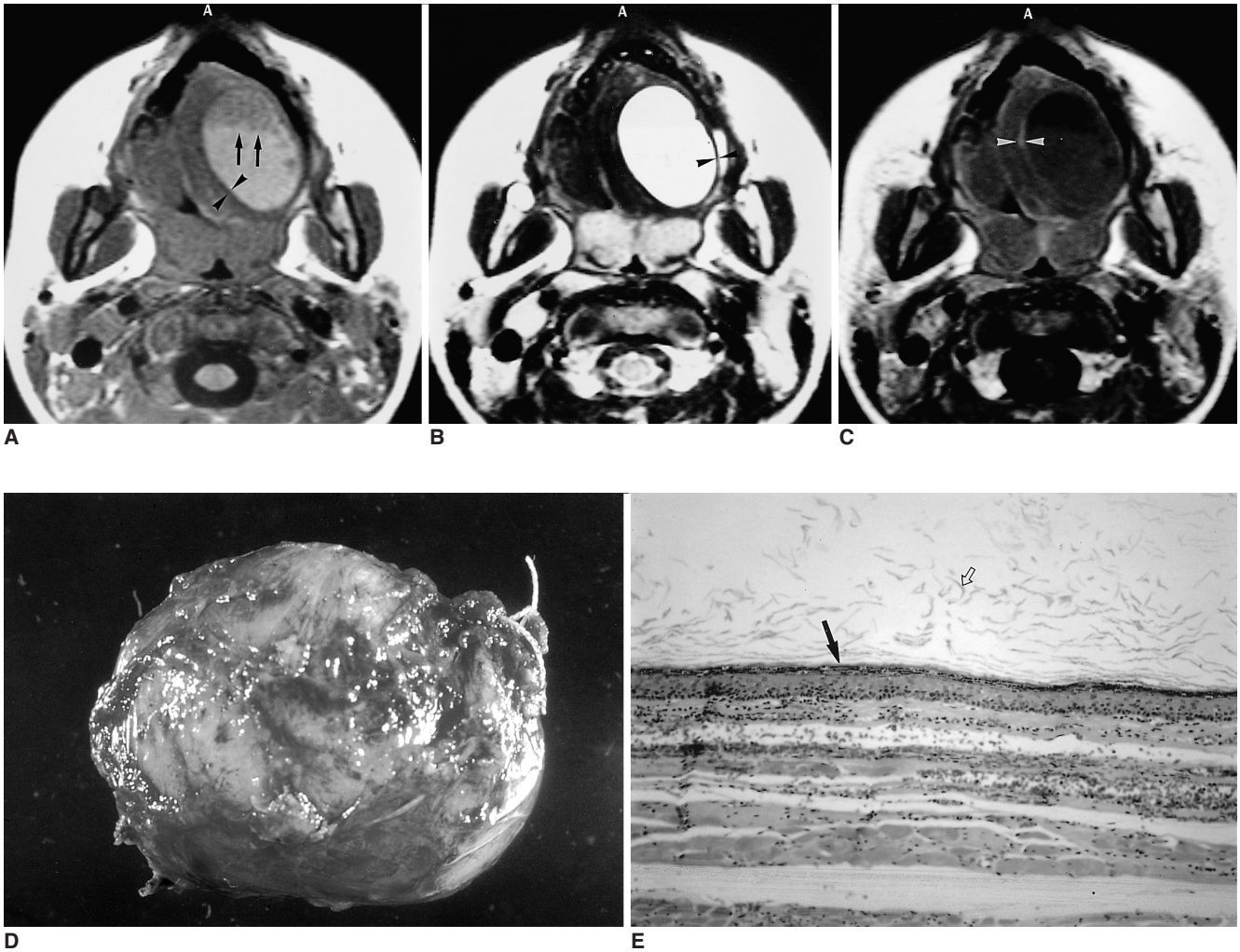
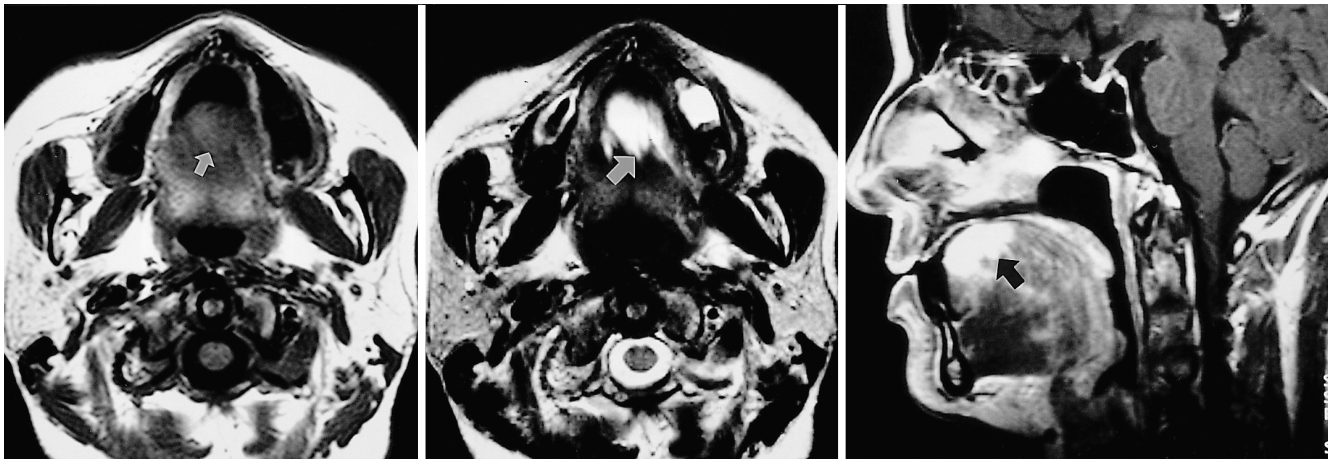


Fig. 4. Epidermoid cyst in a 5-year-old boy with bulging mass of the tongue since infancy.
A. T1-weighted axial image shows a large, unilocular cystic lesion with a fluid-fluid level (arrows) surrounded by a thin low signal rim (arrowheads) at the left lateral aspect of the tongue.
B. T2-weighted axial image demonstrates the high signal intensity of the lesion and a surrounding, thin, low signal rim that may represent either adjacent compressed tissue or a capsule (arrowheads).
C. Enhanced T1-weighted axial image reveals enhancement of the peripheral rim only (arrowheads).
D. Photograph of a gross specimen shows a well-defined, thin-walled cystic mass.
E. Photomicrograph (original magnification (100; H & E staining) indicates that the cyst is covered with simple squamous cell epithelium (arrow), has a fibrous wall and atrophic muscle, and contains keratinaceous materials (open arrow).



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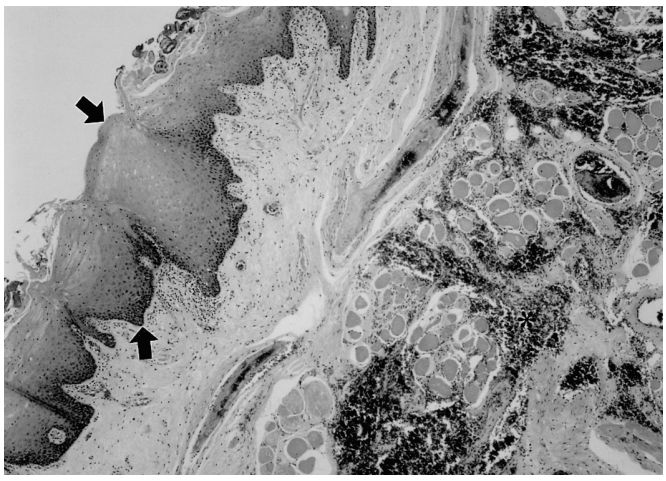

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Fig. 5. Venous malformation in a 35-year-old female with a bluish tongue mass.
A. T1-weighted axial image shows a mass with slightly high signal intensity (arrow) at the anterior portion of the tongue.
B. T2-weighted axial image shows a lesion with high signal intensity (arrow).
C. Enhanced T1-weighted sagittal image demonstrates strong enhancement of the lesion (arrow).
D. Photomicrograph (original magnification ×40; H & E staining) shows a poorly defined venous malformation covered with stratified squamous epithelium (arrows). Note the presence of large, markedly dilated vascular spaces (*) lined by endothelium and communicating with each other.

functional impairment occurs.

Vascular malformations can be investigated with various imaging modalities. Conventional radiographs, though generally of little use, are invaluable if they demonstrate the presence of phleboliths, as these are pathognomonic of VM. CT and MRI provide good anatomic detail, with precise delineation of the extent of lesions and their relation to surrounding structures (Fig. 5). In addition, MR imaging is helpful in that when flow void is identified, it can further characterize the type of flow present. Angiography can be invaluable for delineating feeding and draining vessels and in defining the hemodynamics of vascular lesions. Since its findings are generally normal, it does not, however, contribute to the diagnosis of VM (9). The characteristic features of VM are noted after direct intralesional puncture and lesion opacification, with the demonstration of tortuous, dilated venous channels and the absence of arteriovenous communication. Thus, angiography can help rule out high-flow vascular malformation, with its arterial components.

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