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Granular Cell Myoblastoma of the Cervical Esophagus: MR Findings

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Summary: Granular cell myoblastomas are uncommon, typically benign tumors. Involvement of the esophagus is rare, and frequently they are asymptomatic. We report a granular cell myoblastoma involving the cervical esophagus in a patient who had paralysis of the true vocal cord and progressive dysphagia. At MR imaging, the circumscribed mass was hypointense on T1-weighted images and mildly hyperintense on T2-weighted images, with homogeneous contrast enhancement.

Index terms: Granuloma; Neck, neoplasms

Granular cell myoblastomas were described by Abrikossoff in 1926, at which time these tumors were thought to be muscular in origin (1). However, the derivation of these rare neoplasms is controversial, and presently they are believed to be of neurogenic origin (2). Hence, the term *myoblastoma* is a misnomer and these lesions in general are now referred to as *granular cell tumors*. They predominantly involve the skin and subcutaneous tissues, tongue, breasts, and respiratory tract (3). The gastrointestinal tract accounts for only 5% to 6% of all cases (3), with one third of these localized to the esophagus (4). Of those involving the esophagus, approximately two thirds are located in the distal half. We describe the magnetic resonance (MR) imaging appearance of a granular cell tumor of the cervical esophagus. MR imaging is useful in defining the extent of the primary tumor, which not uncommonly invades the adjacent soft tissues, and is invaluable in identifying lymphadenopathy suggestive of malignancy (5).

Case Report

A 44-year-old black man was referred to our institution with a 2-year history of dysphagia and a sensation of "a lump in his throat." These symptoms had progressed over the prior year with a consequent 5-kg weight loss over the 3 months before admission. The patient also had a 1-year

history of progressive hoarseness. His medical history was remarkable for resection of a benign tongue mass in 1975, which reportedly was a granular cell tumor. Physical examination revealed no palpable neck mass, but was notable for paralysis of the right true vocal cord.

MR studies showed a circumscribed mass inseparable from the cervical esophagus that extended into the tracheoesophageal groove on the right. In addition, the mass extended to and invaded the right lobe of the thyroid gland (Fig 1). There were also imaging findings consistent with right true vocal cord paralysis, including medial orientation of the right false cord and the aryepiglottic fold as well as dilatation of the piriform sinus on the right. No cervical or mediastinal lymphadenopathy was noted.

Direct intraoperative laryngoesophagoscopy was performed; however, the surgeons were unable to advance the rigid cervical esophagoscope beyond the postcricoid region because of obstruction of the lumen of the cervical esophagus. Paralysis of the right true vocal cord was confirmed.

Subsequently, after gaining surgical exposure by dissecting away the overlying soft tissues and muscles, a firm mass approximately 2 × 4 cm in size was noted arising eccentrically from the cervical esophagus on the right, just below the cricoid. The mass extended posteriorly across the midline to the left. The tumor was completely extraluminal and spared the mucosa. There was a small amount of interdigitation of the tumor with the extrinsic muscle fibers of the esophagus. After a tracheostomy was placed for airway protection, the mass was completely resected by blunt dissection. A right thyroid lobectomy was also performed as a result of the MR findings that suggested thyroid involvement. Pathologic examination confirmed infiltration of the adjacent soft tissues and skeletal muscle as well as focal infiltration of the right lobe of the thyroid gland. Findings at histologic examination were consistent with a benign granular cell tumor, including sheets of uniform cells with bland nuclei and cytoplasm with abundant eosinophilic granules without nuclear pleomorphism or mitoses (Fig 1E and F).

During surgery, it was necessary to stretch the left recurrent laryngeal nerve in order to dissect it off the tumor. This resulted in paresis of the left true vocal cord after

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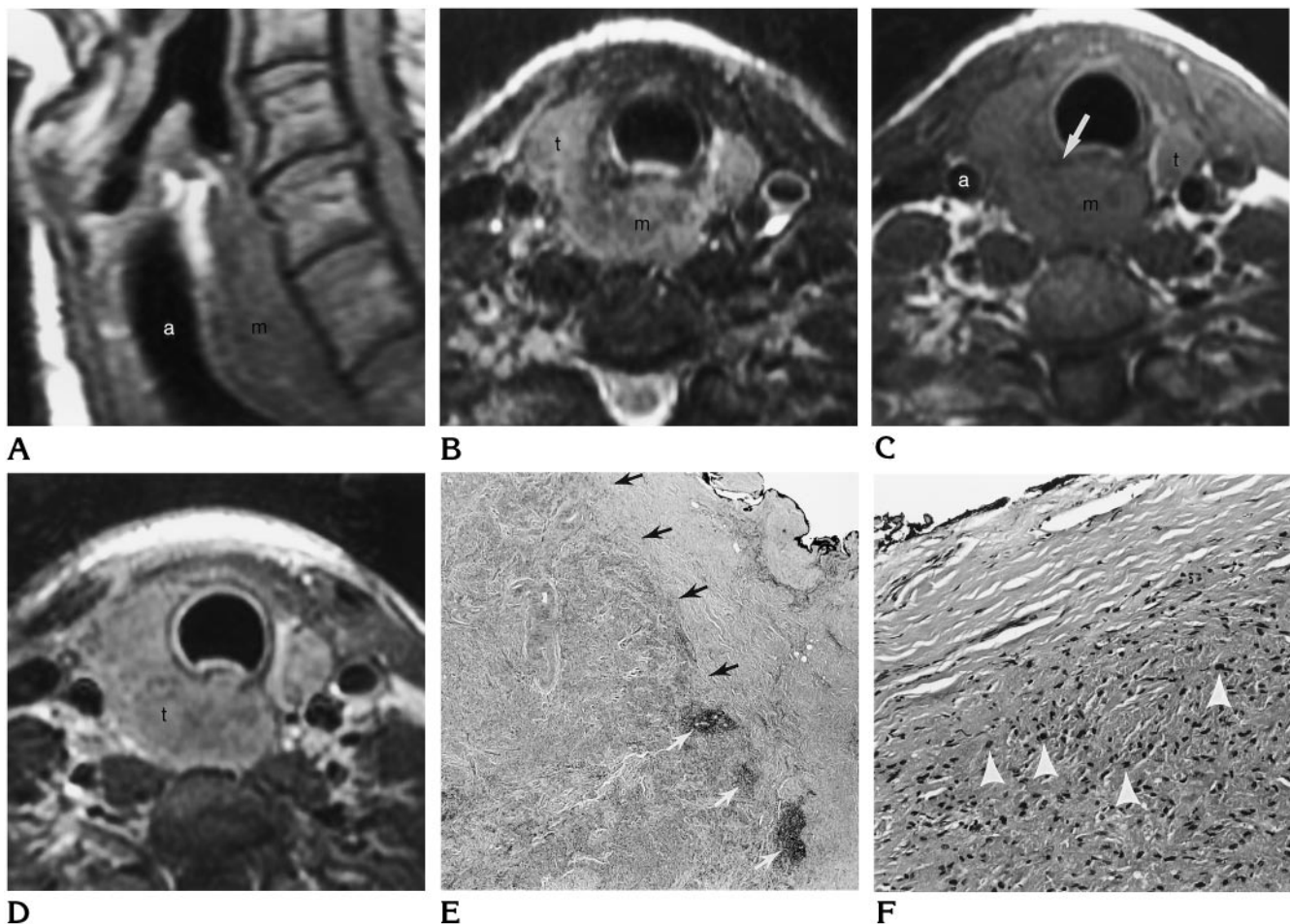


Fig 1. MR images in a 44-year-old man with a granular cell tumor of the cervical esophagus.

A, Sagittal short-repetition-time, short-echo-time image reveals a submucosal hypopharyngeal and upper esophageal mass (*m*) that displaces the airway (*a*) anteriorly.

B, On T2-weighted image, the mass (*m*) is virtually isointense with the thyroid gland (*t*). The esophageal lumen cannot be identified.

C, Note preservation of the fat plane between the left side of the thyroid gland (*t*) and the mass (*m*). This is obliterated on the right side and infiltration of the thyroid gland is inferred. Note that the region of the tracheoesophageal groove (*arrow*) on the right side has been infiltrated by the neoplasm. The lesion remains medial to the internal carotid artery (*a*).

D, The mass enhances uniformly and its exophytic nature is suggested by the tongue of tissue (*t*) extending to the right side.

E, Histologic section shows tumor (*left*) invading and replacing collagen in adjacent connective tissue (*right*). A small cluster of lymph nodes (*white arrows*) is noted at the border of the tumor and native stroma (*black arrows*) (hematoxylin-eosin, original magnification $\times 4$).

F, At higher magnification, the tumor cells are uniform, with abundant cytoplasm and small dark nuclei (*arrowheads*). There is no high-grade nuclear atypia, necrosis, or mitoses (hematoxylin-eosin, original magnification $\times 86$).

surgery, which improved during the following 3 weeks. The patient's postoperative course was otherwise uncomplicated.

Discussion

In comparison with malignant squamous cell carcinoma and adenocarcinomas, benign tumors of the esophagus are uncommon, accounting for approximately 20% of all esoph-

geal neoplasms (6). Many are asymptomatic and discovered incidentally on endoscopic or radiologic examinations performed for unrelated reasons. When symptomatic, patients may experience dysphagia, odynophagia, abdominal pain, weight loss, and occasionally hematemesis. The clinical presentation will depend not only on the size of the underlying tumor but also on its location (distal, middle, or upper esophagus). Benign tumors of the esoph-

agus are usually categorized as mucosal or submucosal in origin. Mucosal lesions include papillomas, adenomas, and inflammatory polyps, whereas submucosal masses include leiomyomas, lipomas, hemangiomas, and cysts. Leiomyomas are by far the most common of the submucosal lesions (7). Granular cell tumors of the esophagus are rare, with less than 100 reported cases, many of these representing incidental findings at autopsy (8).

Granular cell tumors may occur at any age, but frequently present in the third or fourth decades of life. They are more commonly seen in women (3) and black patients (4). They predominantly involve the tongue (40%), the skin and subcutaneous tissues (30%), the breasts (15%), and the respiratory tract (10%). They have been reported in other sites, including the biliary tree (9), urinary bladder (10), female reproductive tract, middle ear, and pituitary stalk (11). Granular cell tumors are usually isolated lesions, but multiple tumors may occur in up to 15% of patients (4, 12, 13), as was the case with our patient, who had had a granular cell tumor of the tongue. In the gastrointestinal tract, they most often appear as small (2 cm or less in size) round or oval submucosal masses (14). Rarely, they may appear as a benign focal stricture (15). The diagnosis of a benign mass is usually suggested by demonstration of a demarcated, smooth submucosal or extrinsic mass lesion on double-contrast esophagography or endoscopy (16). The appearance of these tumors is indistinguishable from and often mistaken for leiomyomas (8).

Few cases of granular cell tumors have been assessed with cross-sectional imaging. Computed tomography in a reported case of a granular cell tumor involving the proximal esophagus showed a well-defined mass circumferentially involving the esophagus (14). MR imaging of a granular cell tumor involving the subglottic larynx showed the mass to be hypointense on T1-weighted images and mildly hyperintense on T2-weighted images with homogeneous contrast enhancement (17). In our case, MR imaging showed a circumscribed mass of the cervical esophagus that had similar imaging characteristics. However, in this patient, MR images also showed infiltration of the adjacent soft tissues and invasion of the right lobe of the thyroid gland. Pathologically, infiltration of nearby soft tissues, including muscle, has been well described in association with

these benign tumors (14, 16). One malignant granular cell tumor has been reported in the literature. It arose in the chest wall and had heterogeneous signal intensity on T2-weighted images with inhomogeneous enhancement after administration of contrast material (5).

The histogenesis of granular cell tumors is controversial. These tumors were initially thought to be of muscular origin, and hence were referred to as *granular cell myoblastomas* (1). However, pathologic evidence suggests that they are of primitive neuroectodermal origin (2, 18). Histologically, granular cell tumors have a characteristic appearance, demonstrating small nests of round or polygonal cells with centrally located small dense nuclei and abundant cytoplasmic eosinophilic granules. S-100 protein has been noted within the granular cells and is a marker of neural crest derivatives, supporting the neurogenic origin of these tumors (2, 18). Acanthosis or pseudoepitheliomatosis of the squamous epithelium overlying these tumors is not uncommon, and may be mistaken for squamous cell carcinoma (4, 12, 13, 19).

Malignant granular cell tumors are uncommon but must be suspected when the primary mass is greater than 4 cm in diameter, when there is extensive infiltration of adjacent tissues, when there is heterogeneous signal intensity or enhancement, or when pathologically enlarged lymph nodes are encountered. However, even benign granular cell tumors may have an infiltrating growth pattern, as was the case in our patient. This finding alone does not indicate malignancy. Histopathologic features suggesting malignant behavior include the presence of necrosis, vesicular nuclei without nucleoli, spindle-cell formation, and mitotic activity (5). Therefore, preoperative radiologic examination to determine the extent of the tumor and the presence of lymphadenopathy must be considered with the pathologic interpretation.

The treatment of choice for large, symptomatic granular cell tumors, like other benign esophageal neoplasms, is wide local excision, which is usually curative (4, 13, 14). Local recurrence is seen in less than 10% of patients, usually in cases in which tumor involves the surgical margins (4). Treatment of small incidental, asymptomatic granular cell tumors is more controversial. Options include endoscopic (16) or open excision (14).

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