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Rhabdomyoma of the Pharyngeal Musculature Extending into the Prestyloid Parapharyngeal Space

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Summary: A 66-year-old woman reported fullness in the right ear for 6 months before admission. A submucosal mass shown by MR imaging and CT to arise in the prestyloid parapharyngeal space was the cause of her symptoms. Histologically, the mass proved to be a rhabdomyoma, a rare benign neoplasm, that arose from the muscular wall of the pharynx.

Index terms: Pharynx, neoplasms; Pharynx, parapharyngeal space

Rhabdomyomas are benign soft-tissue neoplasms originating most often from striated muscle of the myocardium. The most common site of extracardiac rhabdomyomas is the head and neck region. The tumor may be solitary; however, in 20% of cases they are multifocal. Rhabdomyomas account for less than 2% of all striated muscle tumors (1).

Case Report

A 66-year-old woman was admitted to the ear, nose, and throat department with a 6-month history of an intermittent sensation of fullness in the right ear. Six weeks before admission a feeling of pulsatile tinnitus developed in the right ear. There was no history of epistaxis, nasal congestion, otalgia, or weight change. Physical examination revealed a retracted tympanic membrane on the right side with evidence of myringosclerosis. Examination of the oral cavity revealed an anterior right-sided bulge of the soft palate and nasopharyngoscopy showed a medial right-sided bulge of the lateral nasopharyngeal wall. No mucosal lesion was noted. Audiometry showed a slight, bilateral conductive hearing loss.

A computed tomographic (CT) examination done at another institution revealed a mass in the prestyloid parapharyngeal space from the level of the nasopharynx to the submandibular space, displacing the submandibular gland inferiorly. The carotid artery was displaced posteriorly and laterally. The lesion was believed to originate within the prestyloid parapharyngeal space and possibly to involve the wall of the pharynx medially and the deep lobe of the parotid gland laterally. The working diagnosis, made on the basis of the CT findings, was a pleomorphic adenoma (benign mixed tumor) arising in a salivary gland rest within the prestyloid parapharyngeal space.

A magnetic resonance (MR) imaging study performed at our institution showed the lesion to be only slightly hyperintense relative to muscle on T2-weighted images (Fig 1A and B). The mass was inseparable from the superior pharyngeal constrictor (Fig 1C and D). It was also inseparable from the deep lobe of the parotid gland on two to three sections (Fig 1E and F); however, it did not appear to arise from the parotid gland (Fig 1G and H). Because of the possible involvement of the pharyngeal wall and the fact that the mass had a lower signal intensity on T2-weighted images than that of the far more common pleomorphic adenoma of the parapharyngeal space, the possibility of this being a malignant tumor of the minor salivary gland was discussed with the head and neck surgeon. An imaging-guided biopsy was decided against, since this would not influence the surgical approach.

Surgery, via a transcervical approach, involved gross resection of the lesion with clear margins. The excision required a local resection of the pharyngeal wall (the site of tumor origin). Both a frozen section and the final pathologic specimen revealed a rhabdomyoma. The tumor was composed of large, pink polygonal cells with peripheral nuclei and extensively focally vacuolated cytoplasm. Spider cells as well as cross-striations in numerous cells were present. The nuclei were small and uniform, and no significant mitotic activity was noted. Flow cytometry showed diploid DNA with an S-fraction of 3.6%. The postoperative course was uneventful.

Discussion

The parapharyngeal space is divided into prestyloid and post(retro)styloid compartments by most authors (2, 3). Some refer to the poststyloid parapharyngeal space as the carotid space (4, 5). The mass in this case had its center in the prestyloid parapharyngeal space.

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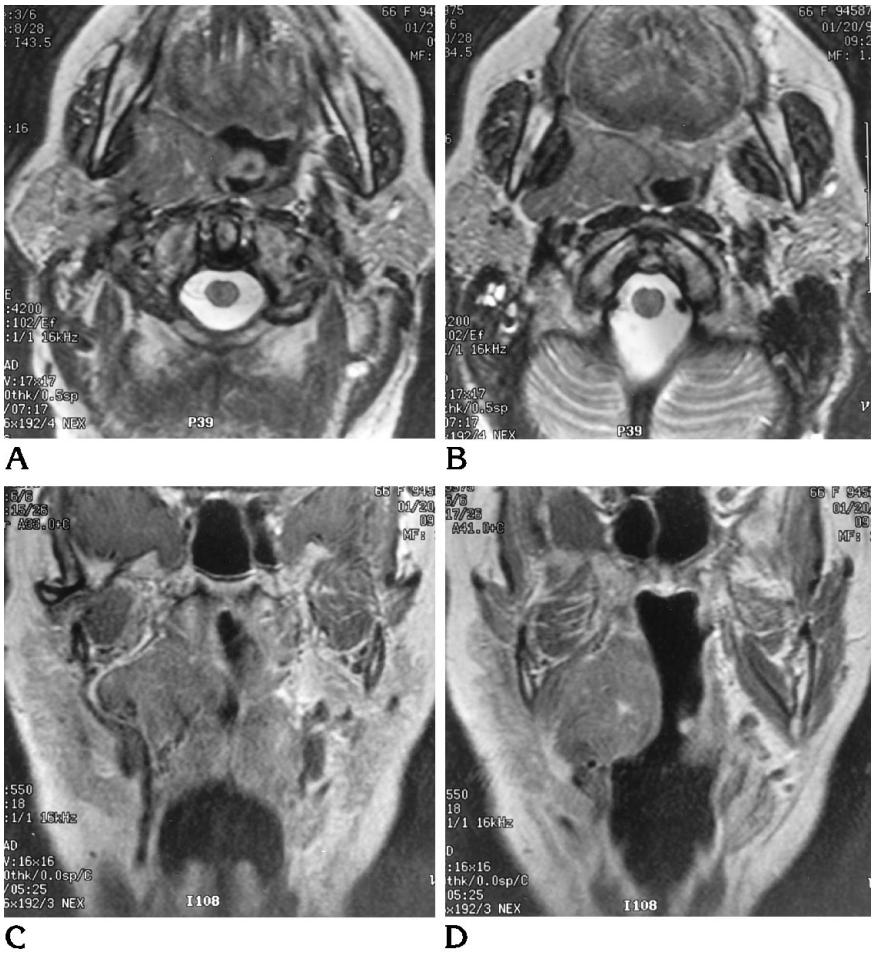

Fig 1. Sixty-six-year-old woman with 6-month history of fullness in the right ear.

A and *B,* Fast spin-echo heavily T2-weighted (4200/102/4 [repetition time/echo time/excitations]) axial MR images show a mass in the right prestyloid parapharyngeal space slightly hyperintense relative to muscle.

C and *D,* Spin-echo T1-weighted (550/18/3) coronal MR images show the mass to be inseparable from the superior constrictor muscle, bulging the lateral wall of the nasopharynx medially.

Figure continues.

The contents of the prestyloid parapharyngeal space are fibroadipose tissue, neurovascular structures, such as the ascending pharyngeal artery and the pharyngeal venous and nervous plexus, and rests of salivary gland tissue (4). Tumors arising in the prestyloid parapharyngeal space are almost exclusively of minor salivary gland origin. Malignant tumors arising in these salivary rests are uncommon. Between 80% and 90% of the tumors are pleomorphic adenomas (benign mixed tumor) (3). Benign and malignant tumors arising in the deep portion of the parotid gland may extend into the prestyloid parapharyngeal space and be indistinguishable from those arising in the space itself. Surgical approaches to the parapharyngeal space vary with the site of origin (6). If the lesion is parotid in origin, a transparotid/transcervical approach will be used to find and protect the facial nerve during tumor resection. If the lesion is separate from the parotid gland, a transcervical approach, usually via the submandibular space, is used. As a practical matter, at our institution, if the mass is not separable from the deep lobe of the parotid gland on high-quality CT scans or MR images over an area of about 1 to 2 cm, a combined transparotid/transcervical approach is always used. If a lesser area of contact is present, a transcervical approach may be used. The rhabdomyoma in this case was easily removed via a transcervical approach.

On MR images or CT scans, benign mixed tumors enhance slightly to moderately after administration of contrast material owing to a variably vascular stroma (7–9). T1-weighted MR images typically show low to intermediate signal intensity relative to muscle; however, areas of highly proteinaceous fluid or hemorrhage may have a high signal intensity. On T2-weighted sequences zones of high signal intensity equivalent to cerebrospinal fluid usually correspond to zones of microcystic morphology caused by many small glandular spaces filled

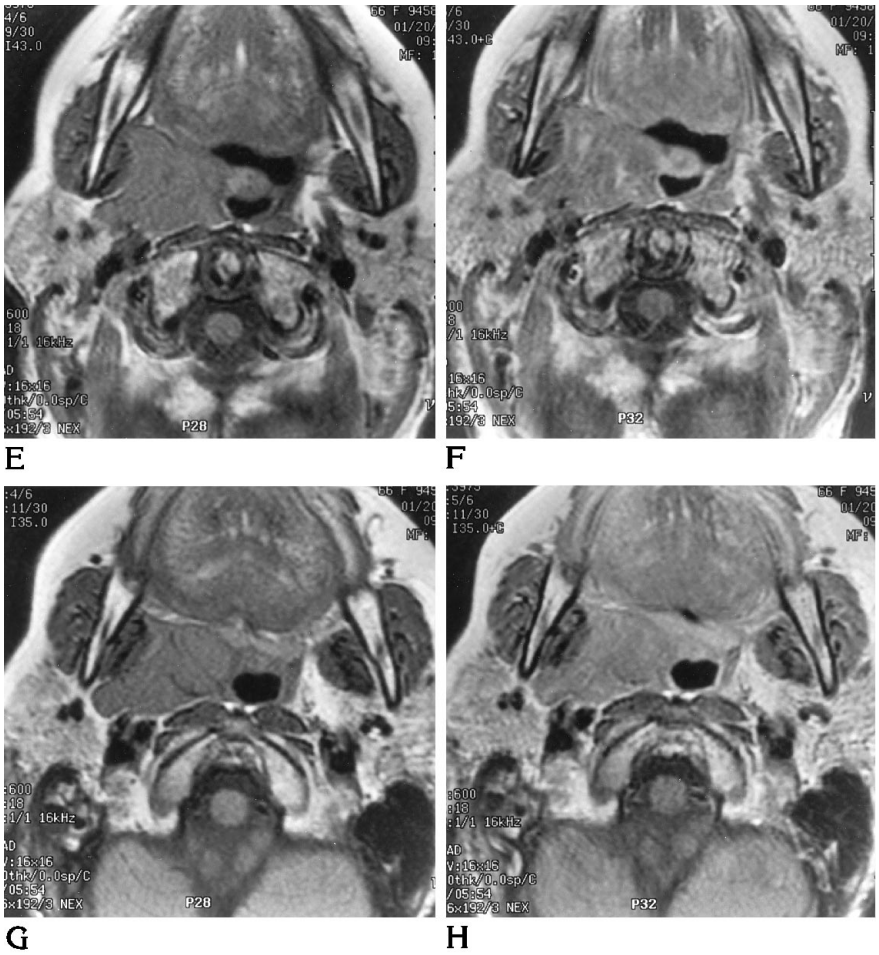

Fig 1, *continued*.

Precontrast (*E*) and postcontrast (*F*) spin-echo T1-weighted (600/18/3) axial MR images show that the mass is not clearly separate from the parotid gland; the tumor enhances slightly.

Precontrast (*G*) and postcontrast (*H*) T1-weighted (600/18/3) axial MR images 8 mm below those in *E* and *F* show the mass involves the pharyngeal wall and the prestyloid parapharyngeal space.

with proteinaceous secretions. If calcification or fibrosis is present, a low signal intensity may be seen on all sequences (2, 3). The mass in this case had little to suggest that it was a pleomorphic adenoma on the basis of its internal morphology compared with the more typical appearance of these lesions just described; only its position suggested an origin in the minor salivary gland.

Carcinomas of a salivary gland rest are often more homogeneously cellular than are the benign mixed tumors just described. As a result, these malignant tumors usually appear as a homogeneously enhancing, possibly infiltrative mass on contrast-enhanced MR images and CT scans. On MR images, the signal intensity tends to be equivalent to muscle on T1-weighted sequences and slightly hyperintense relative to muscle on T2-weighted sequences (3). These trends are seen in other malignant tumors of the parapharyngeal space, such as sarcomas or tumors of vascular or fibrous origin, or in nontumorous conditions, such as pseudotumors (3). In our case, the lack of high signal intensity of the tumor on T2-weighted MR images and possible pharyngeal wall invasion suggested that a malignant tumor of a salivary gland rest was more likely than usual. In retrospect, an origin from the pharyngeal wall could have been considered an alternative explanation because no fat plane was visible between the mass and the superior constrictor muscle. This observation coupled with the internal morphology of the mass as seen on T1-weighted and T2-weighted MR images might have suggested the rare possibility of a pharyngeal wall tumor. Because of the morphology of this lesion, other possible origins, such as branchial cleft cyst or lymphangioma, were not even considered.

Rhabdomyomas are benign tumors of mesenchymal origin. The common cardiac types are regarded as hamartomas and associated with tuberous sclerosis in 50% of the cases. Extracardiac rhabdomyomas are less frequent

and have a predilection for the head and neck region.

Ninety cases of solitary or multifocal extracardiac rhabdomyomas have been described (1), with nine cases arising from the lateral pharyngeal wall and extending into the prestyloid parapharyngeal space (6). The mean age of onset of the disease is 60 years and the male: female ratio is 3:1 (10). In the head and neck region, the larynx, pharynx, floor of the mouth, and base of the tongue are the most common sites of origin (10). There is no tendency for malignant degeneration (6).

The tumor may be quite large at the time of diagnosis, with the duration of symptoms before diagnosis ranging from 2 weeks to 3 years (10). The slow-growing mass will displace the lateral pharyngeal wall and present as an asymptomatic submucosal mass. Pressure on the eustachian tube probably accounted for the intermittent right ear fullness and middle ear disease that were the presenting disorders in our patient. Other clinical manifestations include dysphagia and airway obstruction independent of or related to cranial nerve impairment (6). The treatment of choice is complete excision. Local recurrence rates of up to 42% result from incomplete resections (10).

Histologically, rhabdomyomas can be distinguished from other tumors—such as granular cell myoblastoma (a neurogenic tumor of Schwann cell origin), fetal rhabdomyoma, rhabdomyosarcoma (6), and, less frequently, hibernoma, oncocytoma, and paraganglioma—by their large, round or ovoid cells with eosinophilic, glycogen-rich cytoplasm and focal cross-striations (1, 10). Immunohistochemical stains for myoglobin, muscle-specific actin, and desmin confirm skeletal muscle differentiation. Mitoses have not been seen (1, 10).

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