Unexpected Extracardiac Multifocal Adult Rhabdomyomas With 10 Lesions of the Head and Neck: Epidemiology, Diagnosis, and Therapy

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Adult rhabdomyomas (ARMs) are rare solitary benign neoplasms of the head and neck, and multifocal ARMs are even rarer. Owing to the low incidence and scanty knowledge of this peculiar entity, several patients have been reported to be misdiagnosed or overtreated. This report describes a patient with multifocal ARMs with as many as 10 synchronous lesions. In addition, all published cases of this entity in PubMed, Embase, and Web of Science databases were reviewed up to March 1, 2017. Overall, 10 of 29 reported cases had more than 2 lesions, with a maximum of 10 synchronous masses in the present report. Multifocal ARMs usually present as slow-growing lumps in the parapharyngeal region, with a predilection for older men. Treatments of multifocal ARMs should be tailored and close follow-up is recommended for tiny lesions located in vital structures. In addition, multifocal ARMs should be differentially diagnosed from other multifocal lesions in the head and neck region to avoid aggressive excision and produce a favorable outcome for patients.

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Rhabdomyomas are rare benign neoplasms with skeletal muscle differentiation; they compose fewer than 2% of muscle tumors.[1] Rhabdomyomas are divided into 2 types based on location, including the more common cardiac and extracardiac types. Cardiac rhabdomyomas are considered hamartomas and can regress spontaneously. They occur almost exclusively in the heart of infants or children and often are associated with tuberous sclerosis.[2] Extracardiac rhabdomyomas are true neoplasms, with a reciprocal translocation between chromosomes 15 and 17.[3] It can be further divided into fetal, adult, and genital types according to histologic appearance and clinical presentation.[4]

Adult rhabdomyomas (ARMs) typically present as a slowly growing mass in the head and neck region, with a predilection for elderly men. Although usually solitary, ARMs can occasionally be multinodular and rarely multifocal. According to the literature, in the past decade, only 28 cases of multifocal ARMs have been reported worldwide. Some of those were misdiagnosed or overtreated because of scanty knowledge of the condition. Although elaborate physical and imaging examinations are performed

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preoperatively, some multifocal ARMs are easily overlooked, resulting in reoperation. Therefore, a systematic analysis of multifocal ARMs is needed to summarize the features of this disease to develop individualized therapy.

This report presents a case of multifocal ARMs with 10 simultaneous lesions. To the authors' knowledge, this case has the most multiple lesions of multifocal ARMs reported thus far. A systematic literature review was carried out to summarize the clinicopathologic features and prognostic factors of such a rare entity for future reference.

Report of Case

A 72-year-old man presented to the Department of Oral Maxillofacial–Head and Neck Oncology at the Shanghai Ninth People's Hospital (Shanghai, China) with progressive swelling of asymptomatic masses in the base of tongue accompanied by snoring and slight dysphagia of 5 years' duration. His medical history included hypertension and a hemangioma in the liver. Physical examination found several soft mobile masses with some displacement of the base of tongue. Moreover, there was a soft nontender palpable mass in the left submandibular region. No enlarged lymph nodes were found in the surrounding cervical areas (Fig 1).

Contrast-enhanced computed tomography (CT) of the head and neck showed several solid, well-circumscribed, homogeneous tumors with a maximal diameter of 8 cm filling the base of the tongue. On coronal CT images, an irregular mass on the base of the tongue extended to the left submandibular space, leading to the lateral shift of the submandibular gland (Fig 2). Magnetic resonance imaging (MRI) displayed multifocal tumors isointense to surrounding muscle tissues. Contrast enhancement showed slight inhomogeneous hyperintensity inside the largest tumor (Fig 3). Aspiration of tumors from the left floor of the mouth contained diluted blood without agglutination. Therefore, schwannomas were included in the primary diagnosis. Owing to the aggravating symptoms,

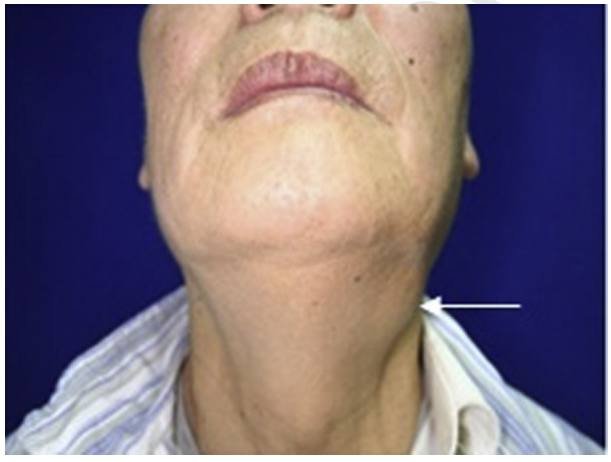

FIGURE 1. Physical examination showed a soft palpable mass (*arrow*) in the left submandibular region.

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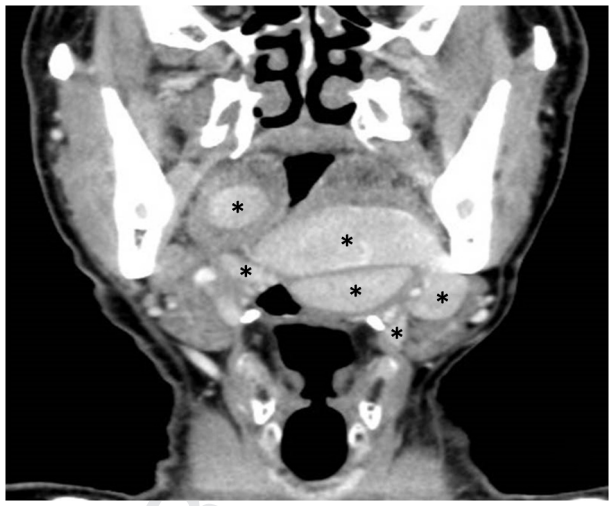

FIGURE 2. Contrast-enhanced computed tomogram showed several solid, well-circumscribed, homogeneous tumors with a maximal diameter of 8 cm in the floor of mouth and the base of tongue (*asterisks*).

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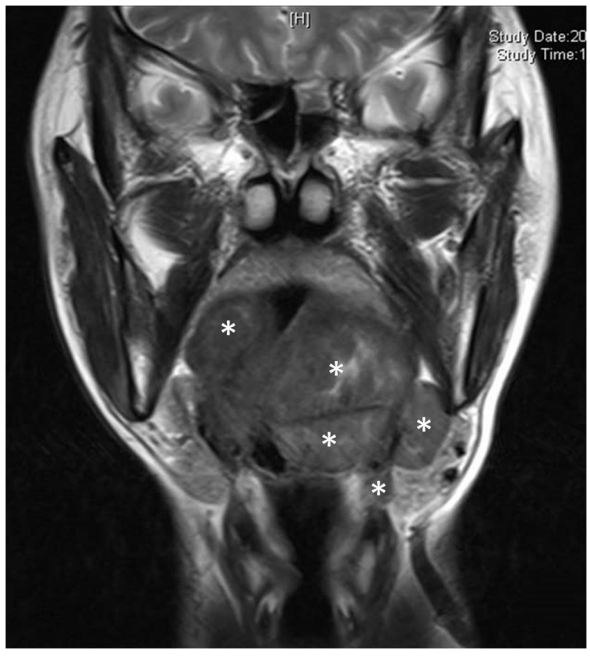

FIGURE 3. Magnetic resonance imaging displayed multifocal tumors isointense to surrounding muscle tissue (*asterisks*).

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