ADULT MULTIFOCAL EXTRACARDIAC RHABDOMYOMA

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The occurrence of multifocal rhabdomyoma is exceedingly rare with less than half a dozen cases collected in the literature. We present a case of multifocal rhabdomyoma of the head and neck that involves extension of the tumor to the vallecula, the glossoepiglottic fold, and the aeriepiglottic fold. (*J Natl Med Assoc.* 1993;85:147-150.)

Key words • rhabdomyoma • tumor

Extracardiac rhabdomyomas are extremely rare tumors of skeletal muscle.[1,2] These tumors were first described in the late 1800s.[2] Less than 75 cases have been reported in the literature with about 75% occurring in the head and neck area.[3] These tumors are biologically and histologically benign.[1] They are usually slow-growing and may remain asymptomatic for long periods of time.[4]

Adult extracardiac rhabdomyomas have been reported in various sites in the head and neck.[5] These sites include the nasopharynx, oropharynx, hypopharynx, tonsil, submandibular area, and larynx. These tumors are usually unifocal; however, several authors have reported single cases of adult multifocal extracardiac rhabdomyomas.[6-8] Characteristically, these tumors have a well-defined margin and are composed of sheets of closely packed large round cells with granular eosinophilic cytoplasm, some containing cross striations. Here we present the details of a multifocal rhabdomyoma found in the right submandibular area of the neck.

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CASE REPORT

A 71-year-old white male presented to the outpatient clinic at Kern Medical Center with a 1-year history of a slow-growing mass in the right neck. He complained of occasional dysphagia, but denied odynophagia, hoarseness, and respiratory distress. He had a history of alcohol abuse and he had a smoking history of two to three packs of cigarettes per day for approximately 60 years.

His past medical history revealed two prior myocardial infarctions, the first one having occurred 23 years prior to this hospitalization and the second one occurring 10 years before this admission. The patient also had a history of pulmonary tuberculosis, angina, and hypertension. His pulmonary tuberculosis, which was diagnosed 21 years prior to the current hospitalization, resolved after 16 months of treatment with isoniazid and rifampin. His hypertension was controlled with dyazide, and he required only occasional nitroglycerin for control of his angina.

On physical examination, the oropharynx revealed a nonulcerated, smooth, exophytic soft tissue mass involving the right tonsil, extending inferiorly into the lateral glossoepiglottic fold to the apex of the aeriepiglottic fold. The larynx and vocal cords were normal. The neck exam revealed a 3 × 4 cm right submandibular mass.

Laboratory studies were within normal limits except for an elevated blood urea nitrogen of 30 mg/dL. Chest radiographs showed evidence of old granulomatous disease with no acute pulmonary process. Computerized axial tomography of the head and neck revealed a mass in the right neck and tonsillar area extending inferiorly to involve the lateral pharyngeal wall into the glossoepiglottic fold area of the pharynx (Figure 1). In addition, there was a right 2 × 3 cm submandibular mass adjacent to the larger mass noted above (Figure 2). A left subclavicular mass approximately 2 × 3 cm with diffuse swelling also was noted.

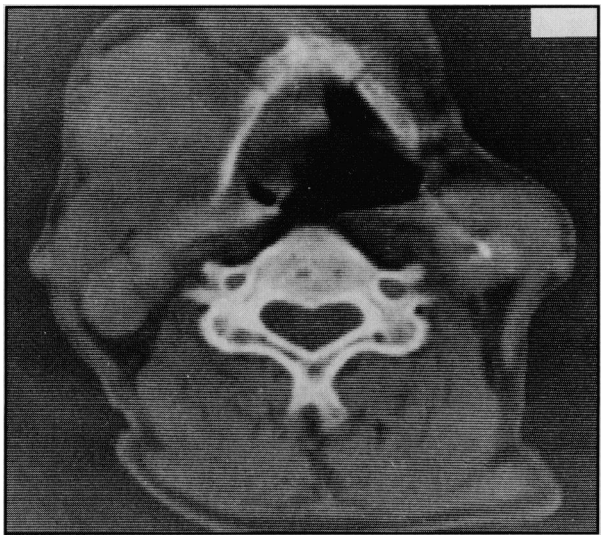

Figure 1. Computed tomography scan of head and neck showing tonsillar mass.

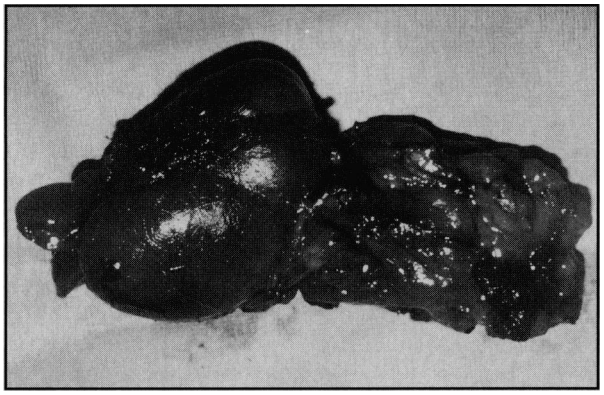

Figure 3. Gross specimen from right neck.

This mass involved the left lobe of the thyroid with a cystic area. The differential diagnosis included squamous cell carcinoma of the oropharynx with cervical metastasis versus lymphoma.

The patient underwent triple endoscopy under general anesthesia. Direct laryngoscopy confirmed the above described mass. Esophagoscopy and bronchoscopy were normal. An initial biopsy and frozen section were nondiagnostic; therefore, an excisional biopsy of the right tonsil and pharyngeal mass was performed. Sections of the tonsil showed multiple fragments of hyperplastic reactive tonsillar tissue, including minor salivary gland tissue and skeletal muscle. Sections of tumor revealed variably sized deeply eosinophilic polygonal cells with small, peripherally located nuclei and extensive areas of intracellular vacuolization. These cells were separated by fine vascular stroma with cross striations.

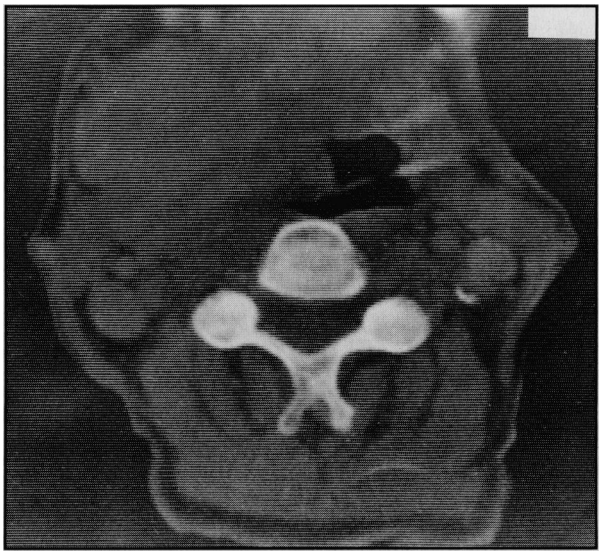

Figure 2. Computed tomography scan of neck showing submandibular mass.

The histopathology was consistent with adult extracardiac rhabdomyoma. The subclavicular mass proved to be a benign follicular adenoma of the thyroid.

After permanent tissue sections were taken, a right neck exploration and excision of the right submandibular and neck mass was performed. A sausage-shaped 4×3 cm mass was noted to be deep and medial to the sternocleidomastoid muscle. The submandibular mass was resected with preservation of the adjacent nerves. The patient did well after the surgery. He was seen in follow-up for the rhabdomyoma and had no complications. Approximately 8 months after the tumor was removed, the patient was seen in the emergency room at Kern Medical Center and died from a myocardial infarction.

PATHOLOGIC FINDINGS

Gross Findings

The tumor tissue from both the tonsil and the neck masses was homogeneous, spongy, and red-brown in appearance (Figure 3). The masses ranged in size up to 5.3 cm in greatest dimension (tonsil). Multiple lymph nodes accompanied the neck masses, but no tumor cells were observed within these. In addition, the tumor masses contained no lymphoid tissue.

Microsopic Findings

The tumor tissues from the tonsil and neck masses were identical in appearance, consisting of homogeneous sheets of round to polygonal, eosinophilic cells

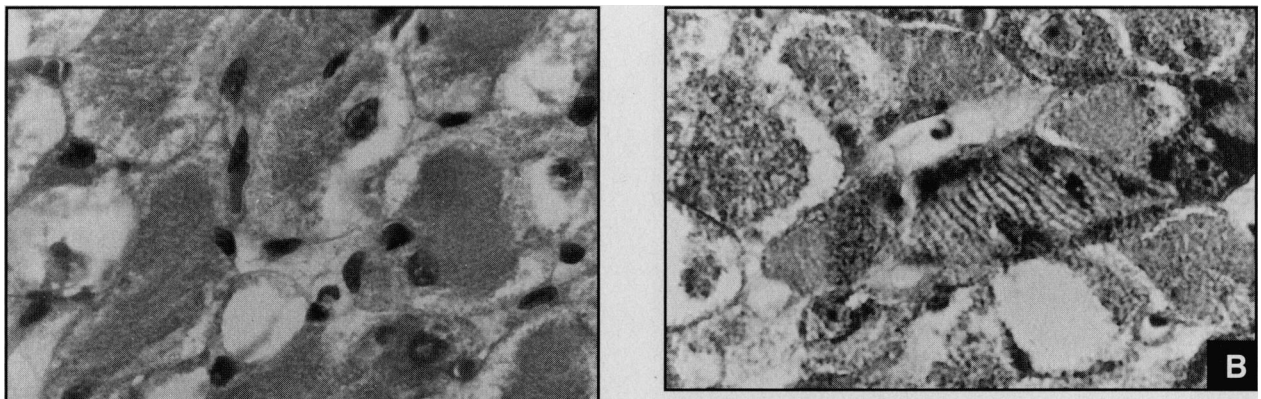

Figure 4. High power photomicrograph with phosphotungstic acid-hematoxylin staining. A) This section shows round, oval, and polyhedral cells with eccentric nuclei. B) The cell in the center of the field displays prominent cross striations.

of varying size (Figure 4A). A fine vascular network and thin fibrous septa separated these tumor masses. The cells contained granular eosinophilic cytoplasm and were frequently vacuolated (Figure 4B). The nuclei were vesicular, round to oval, frequently peripherally located, and exhibited prominent nucleoli. Mitoses were absent. Both cross striations and intracytoplasmic rod-like crystalline structures were easily found in sections stained with the phosphotungstic acid-hematoxylin stain, characteristic of adult rhabdomyoma (Figures 5A & B).

DISCUSSION

Benign extracardiac rhabdomyomas in adults are extremely rare. These benign tumors of skeletal muscle usually occur in the head and neck area.[1,9,10] Rhabdomyomas were first described in the literature in the late 1800s and description of the first case is generally credited to Pendl.[2] Adult rhabdomyomas are mesenchymal in origin,[6] and have been postulated to be the most benign neoplasm in the human body.[11] The incidence of these lesions in unknown.[11] It appears from reviewing the literature that these tumors are most frequently found in muscles derived from pharyngeal arches.[8] These lesions have been classified into fetal and adult types, which have been described extensively in the literature.[12,13]

Rhabdomyomas have been reported in all age groups, but Golz indicates that the mean age of occurrence for adult extracardiac rhabdomyoma is 52.2 years.[3] Extracardiac rhabdomyomas also seem to have a preponderance in males as reported in a series by Jones.[2] At the time of the publication of his report, only three cases of multifocal rhabdomyomas had been described in the literature. The patient we describe here represents another case of multifocal disease, but the most interesting detail of this case is the location of the lesions, which involved the vallecula, the glossoepiglottic fold, and the aeriepiglottic fold.

The symptoms related to rhabdomyomas depend on the site of the lesion. Occasionally, when lesions involve the oral cavity and oral pharynx, a mass in the throat may be observed. In other cases, symptoms of dysphagia, hoarseness, or respiratory distress may be reported when the lesions involve the aerodigestive tract. Lesions involving the neck region and submandibular region may present as neck masses. This patient presented with a mass in the tonsil, which extended into the hypopharynx with mild dysphagia, and also with a mass in the submandibular region, extending into the anterior neck region. This tumor most likely evolved from the palatoglossus and palatopharyngeus muscle as well as from the sternocleidomastoid and the mylohyoid muscle. The portion of the tumor that was involved in the tonsillar region was smooth, exophytic, and easily dissected from the tonsillar fossa. It was easily removed from the inferior pharyngeal constrictor, vallecula, glossal epiglottic fold, and aeriepiglottic fold.

Adult multifocal rhabdomyomas constitute a small but homogeneous group of benign neoplasms of mesenchymal origin.[6] Goldman and Ash have each

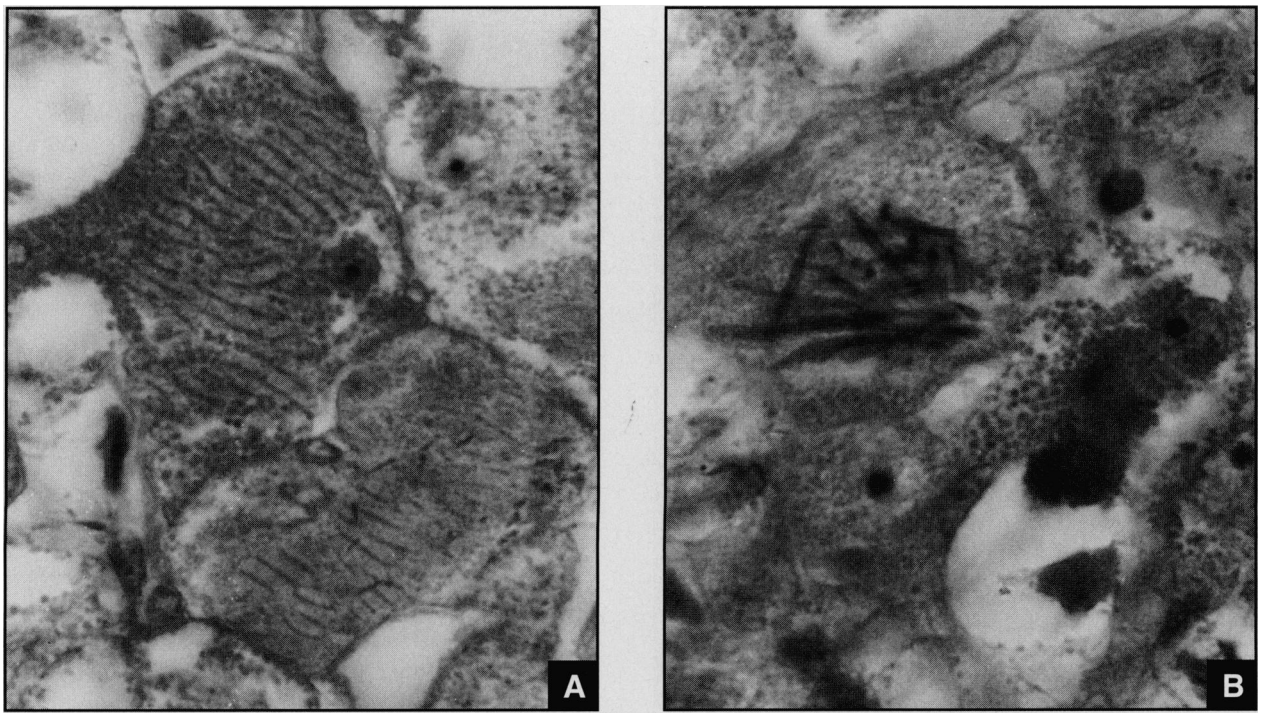

Figure 5. Oil immersion photomicrograph with phosphotungstic acid-hematoxylin staining. A) Cytoplasmic cross striations and granules are seen in the tumor cells. B) The disorganized striations of the myoma cells appear as needle-like structures in the center giant cell.

reported one case of multicentric rhabdomyoma.[7,14] Assor and Thomas reported the first case of multifocal rhabdomyoma on both sides of the neck.[7] These tumors have been reported to have a predilection to occur above the clavicle.

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