Laryngeal Fibrosarcoma: An Over-Diagnosed Tumor

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Sarcomas originating in the larynx are extremely rare and form less than 1 % of all malignant laryngeal tumors. In the past, fibrosarcoma was considered as the most common malignant mesenchymal neoplasm in the larynx and it was generally held that at least half laryngeal sarcomas were fibrosarcomas. Such imprecise statements have been included in the majority of books published on otolaryngology and surgical pathology.

Many spindle-cell lesions such as fibromatosis, nodular fasciitis, malignant fibrous histiocytoma, leiomyosarcoma, monophasic synovial sarcoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumor, spindle-cell squamous carcinoma, spindle-cell melanoma, reparative lesions and inflammatory lesions were formerly indiscriminately classified as fibrosarcomas.

Any reports dating from before 1980 on series of laryngeal fibrosarcoma are therefore unreliable and in more recent literature it is difficult to find a case of laryngeal fibrosarcoma supported by immunocytochemical and/or ultrastructural investigations. Many of the approximately 35 cases of laryngeal fibrosarcoma collected by Gorenstein et al. [1] are more likely examples of spindle-cell squamous carcinoma. Some of the cases of congenital or infantile fibrosarcoma previously reported ought to be reclassified as fibromatosis [2]. The author found no example of this tumor in a series of about 4,000 malignant tumors of the larynx, nor did he encounter any cases of pure fibroma of the larynx. On this point it is important to add that the term laryngeal 'fibroma' has been eliminated in the revised edition of the WHO International Classification of Tumors of the Upper Respiratory Tract [3], which naturally includes the larynx. It is now doubtful whether this tumor really exists in the larynx. Examples of pure fibrosarcoma of the larynx do exist but they are exceedingly rare and there is room for doubt as to the 8 cases reported by Hacıhanefioglu and Oztürk [4] with no immunocytochemical or ultrastructural evidence. Nowadays, the incidence of fibrosarcoma is also low elsewhere in the body and only 1 case was found in a series of 200 soft-tissue sarcomas studied at the Royal Marsden Hospital of London [5].

Immunocytochemically, a true fibrosarcoma is a marker-negative tumor (except for vimentin). Myofibroblasts are often a significant component in fibrosarcomas so the tumor may also coexpress desmin and actin,

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but these tumors should be classified as myo-fibrosarcoma.

Fibrosarcoma has the following histological features: it has an overall, highly cellular, spindle-cell pattern in a 'herringbone' arrangement; it stains positive for collagen and negative for muscle; it is intracytoplasmically periodic-acid-Schiff-negative; reticulin preparation usually reveals a dense mesh-work of collagen fibers between individual cell elements; immunohistochemical investigation demonstrates vimentin reactivity but lysozyme, S-100 protein, Leu-7, neuron-specific enolase, cytokeratin, epithelial membrane antigen and myoglobin are lacking; the ultrastructural cell composition is fibroblastic [6]. Considering the uncertainty as to the true nature of cases reported as fibrosarcomas, it is currently difficult to assess the behavior of this neoplasm in the larynx.

Fibrosarcoma has to be differentiated from laryngeal fibromatosis which may recur but does not metastasize [7]. Accurate histological diagnosis is essential in fibrosarcoma because of the differences it involves in treatment and prognosis by comparison with other tumors. Primary and recurrent tumors call for complete wide local excision, with a generous border of normal tissue. Neck dissection is not necessary for fibrosarcoma unless there is clinical evidence of metastases. In conclusion, we must learn to distinguish a malignant tumor accurately before considering treatment.

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