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Fibromyxoma of the Retropharyngeal Space: MR Appearance with Pathologic Correlation

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Summary: A 19-year-old woman reported difficulties in swallowing and breathing. A submucosal mass, shown by MR imaging in the retropharyngeal space, was the cause of her symptoms. Histologically, the mass proved to be a fibromyxoma. The oval well-delineated lesion appeared hypointense relative to muscle and strongly enhanced after injection of contrast material on T1-weighted images; on T2-weighted images, it appeared hyperintense, and was seen to contain fibrous septa, which were hypointense on all sequences. This case delineates the characteristic MR features of a fibrous component within a rare benign tumor of the retropharyngeal space.

Fibromyxomas are rare benign tumors of uncertain histogenesis that can occur in a number of locations, including the heart, the jawbone, and the skeletal muscles. Fibromyxomas are uncommonly encountered in the head and neck region. Fu and Perzin (1) reported six cases of myxomas in a study of 256 nonepithelial lesions of the nasopharynx, paranasal sinuses, and nasal cavity, only two of which contained a fibrous component. To our knowledge, only one case of retropharyngeal myxoma has been reported in the literature (2), and this study did not include an MR examination. We report the MR and pathologic findings in a patient with a retropharyngeal fibromyxoma.

Case Report

A 19-year-old woman with difficulties in swallowing and breathing for 6 months was referred to our hospital for evaluation. Physical examination revealed a submucosal bulging of the posterior pharyngeal wall. A lateral radiograph of the neck showed a well-delineated retropharyngeal mass measuring 40 mm, extending from the second to the fourth vertebrae. MR images were acquired with a 1.5-T MR system using a conventional SE technique and a neck coil.

MR images showed a clearly limited oval mass in the retropharyngeal space extending from the second to the third vertebrae. The lesion was hyperintense relative to muscle but was not as intense as CSF on T2-weighted images (2000/150 [TR/TE]) and was moderately hypointense on T1-weighted images (450/20). After contrast administration, the lesion enhanced strongly; and hypointense, regular, centrally located septa were seen on all sequences (Fig 1A–E). The mass abutted the cer-

vical spine, but the signal of the vertebrae was not invaded. On the basis of these findings, the possibility of a neurofibroma was considered.

The patient underwent a left cervicotomy, in which the lesion was enucleated. During surgery, the tumor appeared firm, whitish, encapsulated, and adherent to the anterior common vertebral ligament, but it was easily separated from the pharyngeal wall. The gross specimen, measuring $4 \times 2 \times 2.5$ cm, was encapsulated. On sectioning, the lesion consisted of gelatinous areas with intervening fibrous septa. Histologically, the tumor was lobulated and delimited by a pseudocapsule. It was composed of a loosely cellular proliferation of stellate or spindle-shaped cells within an abundant myxoid matrix and some collagenous fibrillary material, but was devoid of inflammatory cells (Fig 1F and G). Immunohistochemical analysis showed strong positivity of tumor cells for antivimentin antibody, but the cells remained negative for PS 100, anti-GFAP (glial fibrillary acid protein), and anti-NF (neurofilament) antibodies. The histologic diagnosis was fibromyxoma.

Discussion

The retropharyngeal space is demarcated anteriorly by the buccopharyngeal fascia, posteriorly by the prevertebral fascia, and laterally by the carotid sheath. It extends from the base of the skull to the posterior mediastinum. The principal components of this space are lymph nodes, nerves, and fat. Lesions of the retropharyngeal space are rare (3), and may include abscess, cellulitis, lymphadenopathy, and malignant tumors.

Myxoma of the retropharyngeal space is very uncommon. In their review of the literature, Tse and Vander (4) described 42 cases of soft-tissue myxoma in the head and neck region but found none in the retropharyngeal space. In 1994, Pahor et al (2) reported a case of myxoma located in the retropharyngeal space and reviewed 59 other cases. In this series, the palate was the most common location (10 of 60 tumors) followed by the parotid (six of 60 tumors). The locations of the 63 cases of head and neck myxomas we identified in the literature are listed in the Table 1.

The gross appearance of myxoma is that of a well-delineated but unencapsulated, round to ovoid mass, tan-yellow to gray-white in color, with a gelatinous

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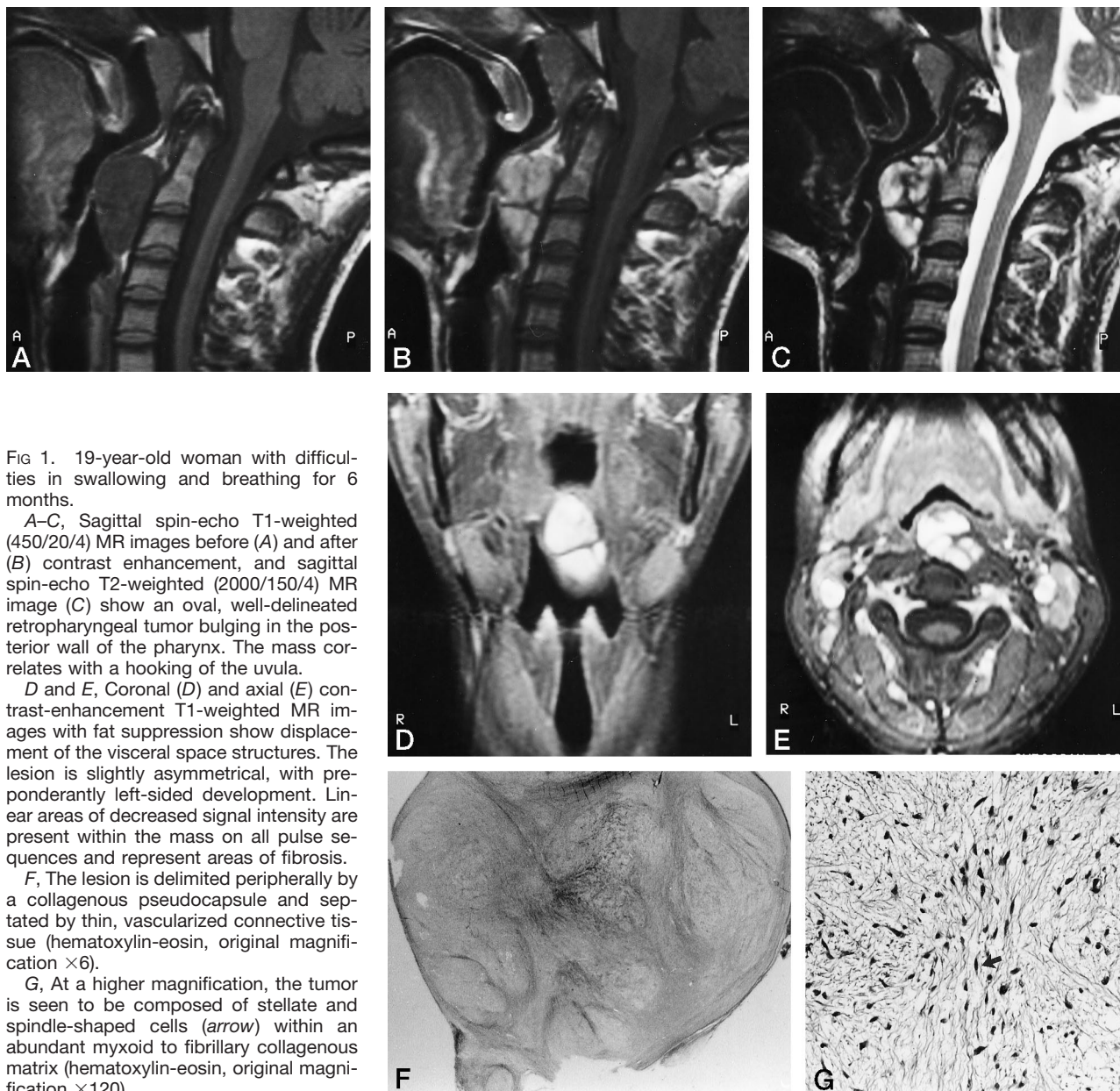


FIG 1. 19-year-old woman with difficulties in swallowing and breathing for 6 months.

A–C, Sagittal spin-echo T1-weighted (450/20/4) MR images before (A) and after (B) contrast enhancement, and sagittal spin-echo T2-weighted (2000/150/4) MR image (C) show an oval, well-delineated retropharyngeal tumor bulging in the posterior wall of the pharynx. The mass correlates with a hooking of the uvula.

D and E, Coronal (D) and axial (E) contrast-enhancement T1-weighted MR images with fat suppression show displacement of the visceral space structures. The lesion is slightly asymmetrical, with preponderantly left-sided development. Linear areas of decreased signal intensity are present within the mass on all pulse sequences and represent areas of fibrosis.

F, The lesion is delimited peripherally by a collagenous pseudocapsule and septated by thin, vascularized connective tissue (hematoxylin-eosin, original magnification $\times 6$).

G, At a higher magnification, the tumor is seen to be composed of stellate and spindle-shaped cells (arrow) within an abundant myxoid to fibrillary collagenous matrix (hematoxylin-eosin, original magnification $\times 120$).

appearance. The consistency varies from soft to firm depending on the fibrous tissue content. These tumors infiltrate adjacent tissue. Microscopically, the tumor is typically composed of an abundant myxoid stroma in which there are a few stellate or spindle-shaped cells and variable amounts of collagen. Depending on the extent of its presence, collagenous fibrillary material may confer the term fibromyxoma to the tumor. Fibromyxomas must be differentiated from myxoid degeneration in soft-tissue sarcomas (liposarcoma, myxoid chondrosarcoma), malignant fibrous histiocytomas, and neurofibromas (5–8).

Myxomas have been described as neoplasms of the primitive mesenchyma. The cause of myxoma remains unknown. Wirth et al (9) suggested a common histogenesis of myxoma and fibrous dysplasia, proposing that “a basic error in tissue metabolism persists far beyond the initial growth period.” Tse and Vander

(4), whose patient presented with a cleft lip and palate, agreed with this hypothesis. Enzinger (10) proposed that stellate or spindle cells in a myxoid matrix are altered fibroblasts secreting acid mucopolysaccharides instead of mature collagen. Glazunov and Puckhov (11) postulated a viral origin on the basis of nuclear and cytoplasmic inclusion bodies in certain intramuscular myxomas.

These tumors may occur at any age, but are prevalent in the third and fourth decades (4). Clinical presentation is nonspecific but is related to the size and location of the tumors and may include difficulties in breathing, dysphagia, and nasal or tubal obstruction if the lesion is adjacent to the airway. On plain films, fibromyxomas of the retropharyngeal space appear as dense, soft-tissue masses located anteriorly in relation to the cervical spine, as described by Pahor and Samant (2).

Location of head and neck myxomas

Anatomic Location	No. of Cases
Palate	10
Parotid	6
Scalp	3
Cheek	3
Lateral neck	5
Lip	4
Alveolar ridge	2
Oral	5
Oropharynx	3
Larynx	3
Face	2
Masseter	2
Retropharynx	2
Other locations (one case of each)	13
Total	63

Conclusion

MR imaging was useful in demonstrating the location of fibromyxoma of the retropharyngeal space, its well-limited appearance, and the absence of extension or connection to the cervical spine. As with other tumors containing myxoid tissue, and myxomas in other locations, the myxoid component of the tumor exhibited low signal intensity relative to muscle on T1-weighted images and markedly hyperintense signal intensity on T2-weighted images. In this case, we noted centrally located bands of low signal intensity on both T2-weighted and contrast-enhanced T1-weighted images that are related to fibrous septa (7, 13–15). Both neurofibroma and fibromyxoma exhibit a central area of low intensity that corresponds to fibrocollagenous tissue. However, the target pattern is usually observed in cases of neurofibroma (16). Although superior to CT in showing anatomic definition, MR imaging does not provide tissue characterization; it is, however, valuable for identifying the fibrous component. Myxomas are considered benign

tumors with a tendency to recur locally. Enucleation or radical excision involving the adjacent tissue is usually the proposed treatment.

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