Case Report

Parapharyngeal space desmoplastic fibroblastoma: A report of a new location for this rare tumor

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Abstract

Desmoplastic fibroblastomas (DF) are rare, benign, soft tissue tumors affecting mainly the subcutaneous tissue or skeletal muscle at a variety of anatomical sites, but rarely occurred in parapharyngeal space. Herein, we describe an unusual case of 61-year-old man, who presented with a parapharyngeal space mass for 1-month. The clinical examination revealed the right lateral pharyngeal wall bulging into pharynx, and mucosa was smooth. Computed tomography showed a heterogeneous tumor with a well-defined margin, which involved the soft palate and extended to the pyriform, measuring approximately 3.5 cm × 2.5 cm × 1.0 cm, which was resected by transoral approach with tracheotomy. Histopathology and immunohistochemistry staining revealed a DF. To our knowledge, this is the first reported case of DF arising in parapharyngeal space.

Key words: Collagenous fibroma, desmoplastic fibroblastoma, parapharyngeal space, surgical approach

Received: 26th June, 2014; ***Revised:*** 9th August, 2014; ***Accepted:*** 24th August, 2014

INTRODUCTION

Desmoplastic fibroblastoma (DF) is a recently described tumor thought to arise predominantly from subcutaneous tissue or skeletal muscle. DF is rarely found in the head and neck region, and no case of DF in the parapharyngeal space has been reported to date. This article presents the first case of a DF identified in the parapharyngeal space; the DF was resected through the transoral approach.

CASE REPORT

A 61-year-old man was referred with a 1-month history of a parapharyngeal space mass. The clinical symptoms included abnormal sensation in the throat and occasional cough when eating. He had no respiratory disturbances, difficulty in swallowing, or hoarseness. His medical history was unremarkable except for a diagnosis of hypertension. Fiberoptic laryngoscopy showed that the right lateral pharyngeal wall bulged into the pharynx and the mucosa was smooth, but the right pyriform sinus could not be opened [Figure 1]. A computed tomography (CT) scan revealed that the tumor had a well-circumscribed border with heterogeneous enhancement bulged into the oropharynx and laryngopharynx between the soft palate and the pyriform sinus. The tumor measured 3.5 cm × 2.5 cm × 1.0 cm and caused narrowing of the pharynx [Figure 2a-c]. We considered that endotracheal intubation under anesthesia could cause bleeding or choking, so we performed a preventive tracheotomy. The patient underwent surgical excision of the tumor through the transoral approach. During the surgery, pathological examination was performed, and there was no evidence of malignant cells in the lesion. The tumor appeared not to have infiltrated into the surrounding tissues and was excised completely. Macroscopic examination disclosed a solid and gray-to-white lobulated mass measuring 3.5 cm × 2.5 cm × 1.0 cm. The diagnosis was DF [Figure 3]. The postoperative course was uneventful, and the endotracheal tube was removed on the 14th postoperative day. Repeat CT scan after 2-month [Figure 2d] and at the 10-month postoperative follow-up showed that he was free of disease.

Access this article online	
Quick Response Code:	Website: www.ijcep.org
	DOI: 10.4103/2348-8093.143515

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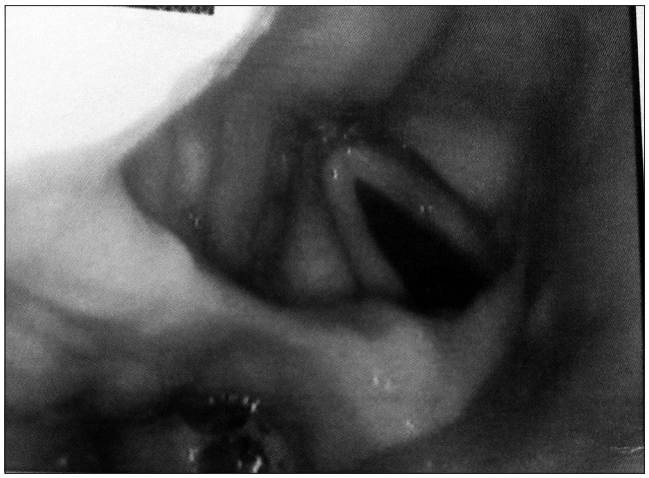

Figure 1: Electronic fiber laryngoscopy showed that the right pyriform sinus could not be opened and the mucosa was smooth

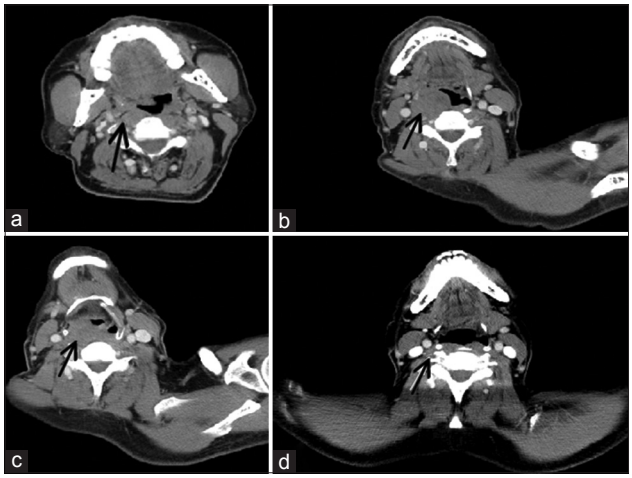

Figure 2: (a-c) A computed tomography (CT) scan showed that the tumor had a well-circumscribed border with heterogeneous enhancement. The tumor bulged into the oropharynx and laryngopharynx between the soft palate and the pyriform sinus, and caused narrowing of the pharynx (the arrow showed the tumor) (d) Repeat CT scan after 2-month of surgery (the arrow showed after resection of the tumor)

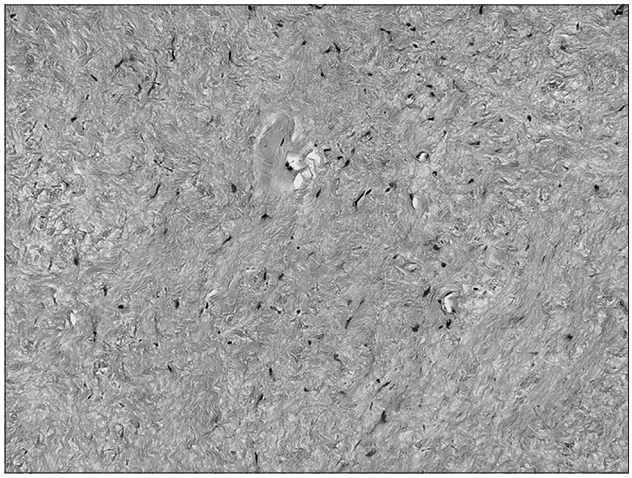

Figure 3: Histological appearance of the tumor (H and E, original magnification ×200) showed desmoplastic acellular fibrous tissue with dense collagen bundles forming the bulk of the lesion

DISCUSSION

The term 'Desmoplastic fibroblastoma' was first coined by Evans in 1995 and was renamed "collagenous fibroma" by Nielsen in 1996. The World Health Organization's classification of soft tissue and bone tumors adopted the name "DF" instead of collagenous fibroma in 2002.[1] DF is a rare, benign, relatively soft tissue tumor, and fewer than 100 cases have been reported in literature. DF is not familiar to otolaryngologists. To the best of our knowledge, this is the first case of a DF in the parapharyngeal space to be reported in the literature.

Desmoplastic fibroblastoma can occur at various anatomical locations, but it typically arises in the subcutaneous tissue or skeletal muscle. It occurs predominantly in males and has a median age of 50 years. The etiology of DF remains unknown, although trauma, endocrine and genetic factors have been suggested as possible etiological agents.[2,3] In our patient, there was no antecedent trauma or evidence of a preexisting lesion to suggest a reactive process. The tumor always presents as a slow growing mass over a period of > 6 months and does not cause symptoms until it reaches a large size,[4] and frequently it is found incidentally on an imaging study.[5]

Because of the limitations on examining this anatomical area, imaging studies are important for the evaluation of parapharyngeal space neoplasms. CT scans can demonstrate the exact size and extent of the tumor and its relationship with the surrounding structures. In general, magnetic resonance imaging (MRI) is superior to CT. MRI examination shows that DF is a well-defined mass and has low signal on both T1-weighted and T2-weighted pulse sequences.[6] Angiography is recommended for all enhancing lesions or vascularized masses. This feature was not observed in our patient, so there was no need for such a procedure. Ultrasound or CT-guided fine-needle aspiration cytology is usually performed to determine the nature of the parapharyngeal space mass.[5] However, needle aspiration cytology is deemed useless because of the low cellularity of DFs.[6] Thus, an excisional biopsy with immunostaining is required for diagnosis and treatment.

On gross examination, the size of DFs ranges from 1 to 20 cm, with a median size of 3 cm.[5] A DF is seen as a firm, well-circumscribed, round to oval or lobulated mass, which on the cut surface appears as a glistening gray-to-white mass.[7] Microscopically, DF tumor cells comprise fibroblasts and myofibroblasts in a collagenous background. Mitotic activity is either absent or minimal. Immunohistochemically, they are generally focally positive for vimentin and rarely for keratins but are not positive for desmin or CD34. It is important to emphasize that immunohistochemistry is not mandatory for the DF diagnosis, but it can aid in the exclusion of other

diagnoses. In our case, immunostaining for desmin, S-100, CD34, CD99, and epithelial membrane antigen were negative. Based on the clinical and histopathological findings, a DF of the parapharyngeal space was diagnosed.

For the surgeon, there are challenges involved in assessing the preoperative condition and in determining the appropriate surgical approach for parapharyngeal space tumors. The total surgical excision of parapharyngeal space tumors is the best treatment. The surgical approach for parapharyngeal space tumors includes transoral, transcervical, infratemporal, transoral endoscopic or robotic,[8] and the appropriate choice depends on the mass size and location.[3] DFs tend to behave in a nonaggressive manner, and no tumor recurrence during follow-up has been reported. Considering the characteristics of our patient observed on imaging, we used the transoral approach.

It is important to understand the characteristics of this rare condition to improve the differential diagnosis in relation to other parapharyngeal space masses such as schwannoma, salivary tumor, paraganglioma, carotid body tumor, adenoid cystic carcinoma, or metastatic carcinoma.[9] Other differential diagnoses of DF include neurofibroma, fibroma of a tendon sheath, malignant fibrous histiocytoma, fibromatosis, and calcifying fibrous pseudotumor.

In summary, we present the first reported case of DF occurring in the parapharyngeal space. When a soft tissue mass is discovered in the parapharyngeal space, the physician should consider DF as a possible diagnosis.

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How to cite this article: Lu D, Liu J, Yang H, Lin YY. Parapharyngeal space desmoplastic fibroblastoma: A report of a new location for this rare tumor. Int J Clin Exp Physiol 2014;1:235-7.

Source of Support: Nil, **Conflict of Interest:** Nil.