MALIGNANT OSSIFYING FIBROMYXOID TUMOR OF THE PARAPHARYNGEAL SPACE

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INTRODUCTION

Ossifying Fibromyxoid Tumor (OFMT) is a rare, recently described tumor. As such, there is a paucity of information in the literature regarding this neoplasm. Most reports cite that the tumor most commonly occurs subcutaneously in the soft tissues of the extremities.1,2 Additionally, OFMT has a predilection for men and exhibits a moderate recurrence rate (27%).3

Malignant forms of the tumor are even rarer than their benign counterparts. At the time of manuscript preparation, the authors found only 27 reported cases of the malignant OFMT (versus 82 reports of benign OFMT). Similar to benign OFMT, most of the reported malignant cases occurred subcutaneously in extremities. However, a few uncommon locations have been described such as the mandible,4 oral cavity,5 and scalp.6 While rare occurrences in the head and neck have been noted, the authors’ knowledge no such tumor has been reported in the parapharyngeal space. The authors present a case of a malignant ossifying fibromyxoid tumor in the parapharyngeal space.

CASE

A 33-year old Pakistani male presented for evaluation for a two-year history of progressively enlarging right-sided neck mass. The patient also complained of gradual dysphonia but denied any dysphagia, odynophagia, snoring, stridor, trismus, or otologic symptoms. His past medical and surgical history was otherwise intact and symmetric. With palpation of the neck, a firm right neck mass was detected, located anterior and deep to the right sternocleidomastoid and spanning from the angle of the jaw to the mid-level of the neck. Flexible nasopharyngolaryngoscopy revealed a slight bulge of the right pharyngeal wall and partial obliteration of the right piriform sinus (Figure 1). Vocal cord abduction and adduction was otherwise intact and symmetric.

Magnetic resonance imaging (MRI) of the lesion indicated a 5.2 x 5.0 x 7.0 right neck mass extending from the skull base to the level of the thyroid cartilage (Figure 2). It showed areas of cystic degeneration and enhancement with gadolinium. Magnetic resonance angiogram revealed lateral displacement of the right internal jugular vein anterior displacement of the external and internal carotid arteries. There was however, no splaying of the external and internal carotid arteries. Preoperative FNAB was not performed secondary to the potential risk of bleeding associated with a suspected paranglioma.

The patient underwent uncomplicated right transcervical excision of the parapharyngeal lesion. Histopathological analysis of the mass demonstrated features consistent with malignant OFMT. It demonstrated high nuclear grade and cellularity along with >2 MF/50 HPF. Immunohistochemical studies showed the presence of basal lamina, immunoreactivity of the neoplastic cells to S-100 protein and glial fibrillary acidic protein (GFAP), and collagen type IV.7 Uniquely, the tumor often demonstrates well-formed re-duplicated basal lamina on electron microscopy which correlates immunohistochemically with positivity for collagen type IV.8 The tumor’s histogenesis is uncertain, but when first described it was suspected to be of cartilaginous or neural origin.1 Since then studies have also suggested that it may be of Schwann cell, smooth cell, myoepithelial, skin adnexal, osseous, and chondroid origin, with Schwann cell origin being the most highly suspected type.9

Most cases of the tumor are benign. However, there has been controversy over the classification of this tumor as benign versus malignant. To address this issue, Folpe and Weiss conducted a study on 70 atypical and malignant variants of the OFMT and attempted to establish distinguishing criteria. The concluded that OFMTs could be considered malignant with a high metastatic potential if they were characterized by or high nuclear grade or high cellularity and >2 mitotic figures (MF) per 50 high-power fields (HPF).10

Ossifying fibromyxoid tumors are likely to recur with reports of up to 27% recurrence. However, it usually occurs ten or more years after resection of the primary lesion. Malignant lesions (those with >2 MF/50 HPF) and infiltrative growth patterns are associated with a higher potential for recurrence but surprisingly necrosis, tumor size, presence of satellite nodules, and positive margins were not associated with recurrence risk.8 Fortunately, metastases are much more rare but have been reported in lungs, mediastinum, thigh, and kidney.11,12

In the case we present, our patient was found to have an OFMT in the parapharyngeal space. The differential diagnosis of OFMT in the parapharyngeal space neoplasms includes pleomorphic adenoma (40%), paraganglioma (20%), neurogenic tumor (including schwannoma?) (14%), malignant salivary gland tumor (13%), followed by miscellaneous tumors such a lipoma, AV malformation, sarcoma, and lymphoma. Approximately 80% of tumors in this region are benign, while 20% are malignant.12 To the author’s knowledge a case of OFMT, although previously found in the head and neck region, has never been reported specifically in the parapharyngeal space. This singular finding demonstrates the need to expand our differential diagnosis of parapharyngeal space tumors to include OFMT. In addition to its unique location, the occurrence of yet another case of the malignant form of this rare tumor underscores the need to further research it so that we may improve our management of it. Considering that malignant forms of the tumor are more likely to metastasize it is particularly important to address management of this possible complication. Currently, there is paucity of information regarding treatment of the malignant OFMT. The only study which does specifically address management of OFMT is that by Suehiro et al which involved a case of mediastinal and pulmonary metastases.13 Surgical management is one option for treatment of this tumor and the one utilized for our patient. However, chemotherapy and radiation treatment should be evaluated as alternative options, especially given that not every patient may be a candidate for surgery. In their study, Suehiro et al found inconclusive evidence for the use of radiotherapy or chemotherapy, again emphasizing the need to further investigate the role of this treatment.13 Although the ossifying fibromyxoid tumor is rare, in its malignant form recurrence rates are substantial. Current literature on both features of the tumor itself and its management are limited and as such it remains a malignancy that demands further study.

CONCLUSION

This case presents the first reported malignant OFMT in the parapharyngeal space. It should be added to this differential diagnosis of parapharyngeal space neoplasms. Considering the paucity of current literature on this malignancy and its treatment coupled with is substantial rate of recurrence, further research and development is certainly needed. Hopefully with advancements in the understanding of this malignancy, treatment can be streamlined to more specifically target it and effectively manage it in the event of metastases.

REFERENCES