Case Report

Dedifferentiated Laryngeal Chondrosarcoma: Combined Morphologic and Functional Imaging With Positron-Emission Tomography/Magnetic Resonance Imaging

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Chondrosarcoma of the larynx is a rare, low-grade malignancy in terms of histology and clinical behavior. We present an unusual case of laryngeal chondrosarcoma, which developed a large dedifferentiated component on recurrence after primary surgery. The diagnosis of dedifferentiation was suggested in view of the morphological and metabolic findings on hybrid positron-emission tomography/magnetic resonance imaging (PET/MRI) and was subsequently confirmed surgically. Whole-organ, slice-by-slice radiologic–histologic correlation revealed excellent delineation of the well-differentiated and dedifferentiated tumor components with PET/MRI. PET/MRI can provide additional functional information to supplement the morphological mapping and histopathology of these tumors.

Key Words: Positron-emission tomography/magnetic resonance imaging; dedifferentiated chondrosarcoma; larynx.

INTRODUCTION

Chondrosarcomas are rare tumors accounting for about 0.5% of all laryngeal neoplasms. Unlike their counterparts in other head and neck locations, laryngeal chondrosarcomas are slow growing, low-grade tumors, and local excision is curative in most cases.1–3 We describe a rare case of dedifferentiation or malignant mesenchymal component within a laryngeal chondrosarcoma. Although there are a few previous reports describing the pathological findings in dedifferentiated laryngeal chondrosarcomas, to our knowledge, this is the first report that describes the findings and role of multimodality imaging with hybrid positron-emission tomography/magnetic resonance imaging (PET/MRI) including diffusion-weighted MRI (DW MRI) in the diagnosis of dedifferentiation in laryngeal chondrosarcoma.

CASE REPORT WITH IMAGING AND PATHOLOGY

A 69-year-old male patient presented to our hospital in 2012 with a long-standing history of dysphonia, which had increased in severity over 2 months. He also complained of increasing respiratory stridor. Computed tomography (CT) study showed a 3.0 × 1.6-cm-sized, well-circumscribed, submucosal mass arising from the right cricoid cartilage with expansile lytic destruction and stippled calcifications. There was no involvement of the vocal cords or esophagus and no cervical adenopathy. Imaging findings and endoscopic biopsy were consistent with a grade 1–2 chondrosarcoma. The patient underwent cricotracheal resection (4.8-cm craniocaudal length), with sacrifice of the right recurrent laryngeal nerve and tracheostomy. Histologic evaluation of the resected specimen revealed a grade 2 chondrosarcoma with a microscopic focus of dedifferentiation. Five months later, he presented with slightly increasing dyspnea and peritracheostomal bleeding. Endoscopy showed right vocal cord palsy, a submucosal glottic bulge covered by intact mucosa, and posterior commissure synechia. Endoscopic biopsy was negative for tumor, and the posterior synechia was resected. Despite the negative biopsy and in view of the previous history, informed consent was obtained and the patient underwent a total body PET/MRI examination on a Philips Ingenuity TF scanner as part of an ongoing clinical study protocol. PET/MRI showed a large lobulated mass involving the larynx and trachea. The mass showed two distinct components (Fig. 1): a small, well-circumscribed, T1 hypointense, T2 hyperintense component involving the right ala of the
thyroid cartilage, as well as a larger, poorly marginated, lobulated component on the left. The left-sided component showed intermediate T1 and T2 signal intensity and heterogeneous contrast enhancement. It involved the left ala of the thyroid cartilage, with infiltration into the left pretracheal soft tissue, strap muscles, and thyroid gland as well as the left pyriform sinus and proximal esophagus. On DW MRI, the two components of the mass showed distinct behavior (Fig. 1c): the right-sided component showed high apparent diffusion coefficient (ADC) values, whereas the left component showed low ADC values. Although the smaller right-sided component appeared consistent with a classic chondrosarcoma, the large left-sided component was considered to be suspicious for a more aggressive high-grade tumor. Fused fluorodeoxyglucose (FDG)/positron-emission tomography (PET) images showed very high FDG uptake (standardized uptake value [SUV]mean = 24; SUVmax = 32) within the left component (asterisk) and very low FDG uptake (SUVmean = 0.945; SUVmax = 9.7) within the right component (white arrows). (d) Axial fluorodeoxyglucose (FDG)/positron-emission tomography (PET) image shows very high FDG uptake (SUVmean = 24; SUVmax = 32) within the left component (black arrow) in keeping with very high tumor metabolism. In contrast, moderate uptake was seen in the region of the right component (asterisk, SUVmean = 3.7; SUVmax = 4.7). (e) Fused axial PET/MRI shows very high FDG uptake within the large left-sided component (white arrow). This is in sharp contrast to the small, round, right-sided component (asterisk), which shows minor FDG uptake. (f) Axial whole-organ histologic slice of the surgical specimen obtained at the same level as images 1a through 1e shows the biphasic tumor with a well-differentiated, low-grade chondrosarcoma component (asterisk) and the large dedifferentiated component (arrows). [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]

Fig. 1. (a) Axial T2-weighted magnetic resonance imaging (MRI) shows a laryngeal mass with a small, rounded, well-circumscribed, T2hyperintense component on the right side (asterisk) and a large, lobulated, intermediate signal-intensity component on the left (white arrows). (b) Contrast-enhanced axial T1-weighted MRI shows nonspecific enhancement of the small right-sided component (asterisk) and of the lobulated left-sided component (white arrows). Note the tumor spread into the prelaryngeal strap muscles and hypopharynx. (c) Axial apparent diffusion coefficient (ADC) map shows high ADC values (ADCmean = 2.11 × 10 \(-2\) mm\(^2\)/s) within the right-sided component (asterisk) and very low ADC values (ADCmean = 0.945 × 10 \(-2\) mm\(^2\)/s) within the left component (white arrows). (d) Axial fluorodeoxyglucose (FDG)/positron-emission tomography (PET) image shows very high FDG uptake (SUVmean = 24; SUVmax = 32) within the left component (black arrow) in keeping with very high tumor metabolism. In contrast, moderate uptake was seen in the region of the right component (asterisk, SUVmean = 3.7; SUVmax = 4.7). (e) Fused axial PET/MRI shows very high FDG uptake within the large left-sided component (white arrow). This is in sharp contrast to the small, round, right-sided component (asterisk), which shows minor FDG uptake. (f) Axial whole-organ histologic slice of the surgical specimen obtained at the same level as images 1a through 1e shows the biphasic tumor with a well-differentiated, low-grade chondrosarcoma component (asterisk) and the large dedifferentiated component (arrows). [Color figure can be viewed in the online issue, which is available at www.laryngoscope.com.]
DISCUSSION

Laryngeal chondrosarcomas are rare tumors accounting for <0.2% of all head and neck malignancies and about 0.5% of all laryngeal tumors. Unlike other head and neck chondrosarcomas, laryngeal chondrosarcomas are low-grade tumors (grade 1 and 2) that generally follow an indolent clinical course. The cricoid cartilage is involved more often than the thyroid cartilage, and the tumors are typically submucosal. They commonly present with hoarseness, dysphagia, or a neck mass in older men who have had symptoms for about 2 years. On MRI, laryngeal chondrosarcomas appear similar to chondrosarcomas in other body locations, with the tumor matrix showing high T2 signal corresponding to hyaline cartilage. Small areas of intraleisional low signal correspond to intratumoral stippled calcifications, seen even more elegantly on CT. The enhancement pattern of chondrosarcomas is variable. Laryngeal chondrosarcomas portend an excellent overall prognosis, and conservative voice-sparing surgery is usually curative.

Rarely, an additional aggressive malignant mesenchymal component or dedifferentiation may develop within a chondrosarcoma. Dedifferentiation occurs in about 5% to 10% of all chondrosarcomas and has been more commonly described in bones, where the dedifferentiated components are osteosarcomas, fibrosarcomas, and Ewing sarcomas. Although integrated morphological and functional imaging is often performed for evaluating bony chondrosarcomas, its role in laryngeal chondrosarcomas has not yet been described. We report a case of dedifferentiated laryngeal chondrosarcoma, and discuss the role of integrated functional imaging with PET/MRI in its diagnosis.

CONCLUSION

Dedifferentiation within a laryngeal chondrosarcoma is a rare entity. We describe the role of integrated morphological and metabolic imaging with PET/MRI in its diagnosis. PET/MRI can provide additional functional information to supplement the morphological mapping and histopathology of these tumors.

BIBLIOGRAPHY


