Case Report

Left neck undifferentiated sarcoma with synchronous left kidney renal cell carcinoma

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ABSTRACT

Parapharyngeal space sarcomas are rare malignancies and most in the reported literature are synovial sarcomas. Here we present the case of a 27-year-old female with a painless, left-sided neck mass who underwent computed tomography and magnetic resonance imaging evaluation which demonstrated a benign appearing mass that upon resection unexpectedly proved to be a high-grade undifferentiated sarcoma with rhabdoid features. Staging computed tomography revealed a synchronous renal cell carcinoma with clear cell and papillary features. Despite an extensive genetic work-up no underlying cause linking these two cancers was discovered.

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1. Introduction

Sarcomas constitute approximately 2% of all head and neck cancers with undifferentiated sarcomas accounting for about 38% of this group. Approximately 1000 new cases of head and neck sarcoma are diagnosed each year in the United States, with the median age of diagnosis at 50-54 years [1]. On the other hand, renal cell carcinoma (RCC) accounts for 80%-85% of primary kidney tumors with 64,000 new cases diagnosed in the United States each year. The median age of diagnosis for primary RCC is 64 years [2].

2. Case presentation

A 27-year-old white female presented with left submandibular swelling with associated dysphagia and hoarseness. She had no significant past medical history. There was a family history of cancer in all grandparents but no first degree relatives with cancer.

Computed tomography (CT) neck demonstrated a well-circumscribed, hypodense but vascularized mass in the left parapharyngeal space sarcomas (PPS) (Fig. 1). Contrast-enhanced magnetic resonance imaging showed an avidly enhancing, Short TI Inversion Recovery (STIR) hyperintense, well-circumscribed mass in the left PPS just cephalad to the carotid bifurcation (Figs. 2, 3a, and b). The mass displaced the carotid sheath posteriorly and the submandibular gland anteriorly with no apparent invasion of adjacent structures. There was a borderline enlarged left level IIa node but no additional lymphadenopathy.

The clinical teams believed the mass to be a paranglioma given the benign imaging appearance and avid enhancement in conjunction with the young age of the patient. An ultrasound-guided fine needle aspiration (FNA) demonstrated some spindle cells but due to the difficulty in the loca-
Fig. 1 – Axial Contrast enhanced computed tomography (CECT) demonstrates a hypodense mass with internal vascularity in the left parapharyngeal region.

Fig. 2 – Sagittal T2WI demonstrates the mass just anterior to the carotid bifurcation. No splaying of the Internal carotid artery (ICA) or External carotid artery (ECA).

Fig. 3 – (a) STIR sequence demonstrates a well-circumscribed, avidly enhancing mass in the left parapharyngeal space displacing the carotid sheath posteriorly. (b) Fat saturated contrast-enhanced T1-weighted image showing same mass in the left parapharyngeal space.

tion of the mass, the specimen was paucicellular. Stains on the sample were negative for S100 and synaptophysin; however, this was also attributed to possible sample error. Additionally, the 24-hour urine metanephrines and normetanephrines were within normal limits.

The patient was taken to the OR under the continued assumption that the mass represented either a paragangioma
or perhaps myoepithelial tumor from the deep lobe of the parotid gland. Frozen section intraoperatively demonstrated malignant cells, and a gross total resection and nodal dissection were performed. Permanent section reported positive margins with no nodal disease and no perineural spread.

Stain for sarcoma (Vimentin) was diffusely positive; however, stains for rhabdomyosarcoma (Desmin and myogenin) were negative. Stains for malignant solitary fibrous tumor (CD34, CD99, and BCL2); angiosarcoma (CD31 and CD34); synovial sarcoma (CD99, S100, and EMA); melanoma (S100, SOX-10); and dendritic sarcoma (CD45, CD68, and CD21) were all negative. The pathology department believed malignant fibrous tumor and synovial sarcoma to still be high in the differential despite negative staining. However, both send out tests to ARUP including Signal transducer and activator of transcription (STAT) 6 for malignant solitary fibrous tumor by immunohistochemistry and SS18 (SYT) (18q11) gene rearrangement for monophasic synovial sarcoma by FISH were also negative. The final diagnosis was high-grade undifferentiated sarcoma with focal rhabdoid features.

CT of the chest, abdomen, and pelvis performed for staging purposes and to rule out a potential primary tumor demonstrated a 4 cm heterogeneously enhancing, solid mass in the left kidney as well as borderline enlarged ipsilateral retroperitoneal lymph nodes (Fig. 4). FNA was consistent with RCC with clear cell and papillary features. Subsequently, positron emission tomography (PET) CT showed avid radiotracer uptake by the renal mass but no nodal disease.

The patient received adjuvant radiation to the neck and suffered mild mucositis, fatigue, and weight loss but otherwise no significant complications. Following completion of radiation therapy, she underwent partial nephrectomy for resection of the synchronous RCC. Final pathology diagnosis for the renal tumor was grade 3 RCC unclassified with clear cell and papillary features. The margins were clear and all nodes were negative.

An extensive genetic workup including a STAT workup for pathogenic variants of 9 genes associated with increased risk for breast cancer were all negative, including TP53 which has been linked to several malignancies including sarcomas. A larger panel of 17 genes associated with an increased risk for renal cell cancer was also negative. A variant of uncertain significance in the FLCN tumor suppressor gene was identified, which has been associated with increased risk of some clear cell RCC and colon cancers [3], but it is unclear whether this variant was the cause of this patient’s condition.

### 3. Discussion

PPS neoplasms are rare accounting for approximately 0.5% of head and neck tumors, and within this group approximately 70%-80% are benign and 20%-30% are malignant [4]. Malignancies within the PPS can include sarcomas, lymphoreticular malignancies, minor salivary gland tumors, and metastases [5]. Much of the reported literature on sarcomas of the parapharyngeal space has focused on synovial sarcomas [5–8]. Our case was ultimately diagnosed as undifferentiated sarcoma, as stains for rhabdomyosarcoma and angiosarcoma were negative and synovial sarcoma was excluded by molecular analysis.

Our case shares similarities with other reported synchronous sarcoma cases and highlights some of the difficulties others have reported in diagnosing and managing this disease. First, the relative rarity of the disease and potentially benign appearance on imaging can result in the consideration of malignancy being overlooked. Secondly, the relative inaccessibility of the PPS can make biopsy difficult which can further complicate diagnosis. A case series of five PPS’s, reports misdiagnosis by FNA in two cases, with one case correctly diagnosed by intraoral open biopsy [9]. The median patient age in this series was 35 years. In additional series of four different PPS malignancies reports, only one was correctly diagnosed preoperatively by FNA [3]. In retrospective discussion with our ENT colleagues, it was found that had the diagnosis been known preoperatively, the preferred surgical approach given the size of the tumor would have been the more morbid transmandibular approach in order to maximize the possibility of complete extirpation. As clean surgical margins were not achieved, radiation to the tumor bed was given postoperatively.

The discovery of a second mass in the left kidney during staging CT further complicated matters as it was not known whether this represented a metastasis from the neck, a primary sarcoma, or a synchronous renal malignancy. FNA of the renal mass revealed an RCC histologically unrelated to the disease in the neck. RCC is the third most common malignancy to metastasize to the head and neck region, behind breast and lung cancers, accountable for 8%-15% of head and neck metastases [10]. It is believed that head and neck carcinomas have a 2%-3% incidence of secondary primary lesions, but these malignancies occur most often in the upper aerodigestive tract possibly due to field cancerization or common clonal origin [11].

Additionally, Merimsky et al. have reported a 7.5% frequency of second and third primary tumors among patients in whom one of the tumors was a soft tissue sarcoma (STS), most often occurring incidentally [12]. In agreement with their findings, our case appears incidental as there were no known
cancerinogenic influences or susceptibility factors (an extensive genetic workup was negative and no known first degree relatives with cancer). In their series, approximately 21% (6/28) of patients had synchronous primary tumors with the STS diagnosed first in exactly 50% as in the current case. Unlike their series, however, our patient had a high-grade sarcoma and was younger than the median age of their patients (47 years). Of note, of the 14 patients with STS as their primary tumor, approximately 28% (4/14) had RCC as their secondary tumor.

Head and neck sarcomas tend to have a higher rate of local recurrence after treatment because the anatomical structures nearby render complete extirpation difficult. This failure of complete resection is associated with a relatively poor prognosis due to local recurrence and metastasis. The treatment consists of surgical removal, followed by radiation therapy postoperatively, and possibly adjuvant chemotherapy based on the histologic type. The 5-year survival rate for head and neck sarcoma is 49%-55% [1]. This patient's low stage I RCC has a 5-year survival chance of more than 90%. The definitive treatment for RCC without metastasis is surgery [13]. In conclusion, we report a rare case of high-grade PPS undifferentiated sarcoma with apparently coincidental synchronous RCC. Our patient underwent gross total resection of the PPS with positive margins and subsequent adjuvant radiation to the tumor bed, and partial nephrectomy for the synchronous RCC. At the time of this writing, no long term follow-up is available.

Consent

Informed consent for the case to be published (including images, case history and data) was obtained from the patient(s) for publication of this case report.

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The authors certify that they have no affiliation with any organization with any financial or non-financial interest in the subject matter discussed.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi: 10.1016/j.radcr.2018.04.027.

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