



Desmoplastic Round Small Cell Tumor of the Submandibular Gland: A Case Report and Review of the Literature

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BACKGROUND

Desmoplastic Round Small Cell Tumor (DSRCT) is a rare, aggressive neoplasm most commonly found intra-abdominally with a 4:1 male to female predominance¹. Typically, at the time of diagnosis, these tumors are large and advanced; thus, hold a poor prognosis. Extra-abdominal DSRCT is exceedingly rare with only seven known cases of major salivary gland DSRCT. Of those cases, nodal metastasis was a poor prognostic indicator; however, data is limited². Here we present a case of a 27-year-old male with a rapidly enlarging, firm, painless neck mass ultimately confirmed by pathology and immunohistochemical staining to be DSRCT. PET/CT showed no FDG uptake beyond the neck. The patient received surgery along with adjuvant chemotherapy.

CASE PRESENTATION

- 27-year-old male who presented with an enlarging right submandibular mass for three months. No recent infections, or systemic symptoms.
- Physical Exam: firm, mobile and non-tender right submandibular mass
- Computed tomography (CT) neck with contrast: enlarged lymph node adjacent to the right submandibular gland versus submandibular mass
- FNA: epithelial cells with basaloid features
- Excisional biopsy: intraoperatively appeared scarred and adherent to surrounding submandibular gland
- Surgical pathology: small undifferentiated cells arranged in nests of various sizes and shapes embedded within desmoplastic stroma with mitotic figures and central necrosis
- Immunohistochemistry:
 - Positive CK7, panCK, WT1, and desmin. TLE1 and FLI1 show patchy and weak nuclear staining. Fluorescence In Situ Hybridization testing for EWSR1 is positive.
 - Negative staining for myogenin, MYoD1, synaptophysin, chromogranin, NUT, SOX10, CK5/6, TTF1. CD99 was essentially negative.
- Positron emission tomography (PET)/CT: FDG uptake in right level II neck concerning for ipsilateral adenopathy, otherwise no abnormal FDG uptake
- Returned to OR: submandibular gland resection, completion right level Ib, II-III neck dissection
- Surgical pathology: positive level IIa lymph node measuring 0.5-2.5 cm in greatest dimension with extranodal extension



Figure 1. Computed tomography (CT) scan of the neck with contrast (coronal view) showing submandibular gland asymmetry, right > left.

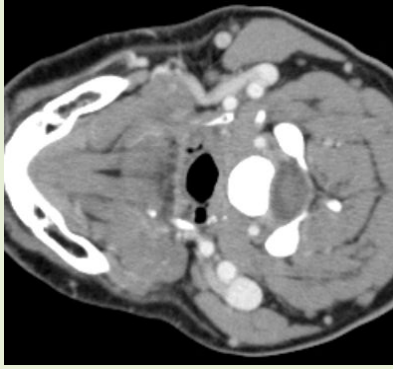


Figure 2. Computed tomography (CT) scan of the neck with contrast (axial view) showing submandibular gland asymmetry, right > left.

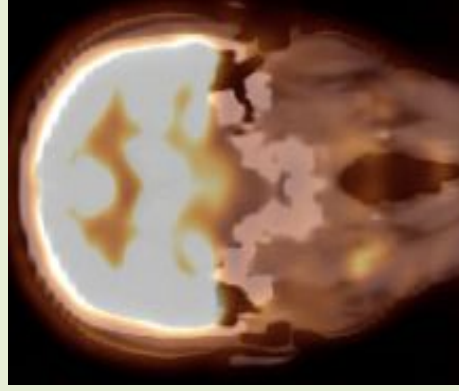


Figure 3. PET/CT scan (coronal view) showing a right submandibular neck mass measuring 1.6 x 1.9 cm.

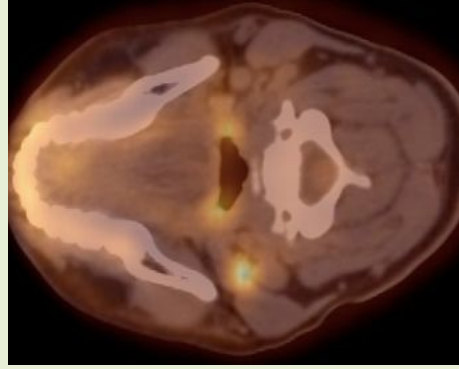


Figure 4. PET/CT scan (axial view) showing a right submandibular neck mass, peak SUV 7.9.

DISCUSSION

- DSRCT is aggressive and rare with limited data; only seven other cases reported in the major salivary glands, including three in the submandibular gland
- Treatment varies in the literature, with no specific, established protocol regarding optimal treatment of head and neck DSRCT
 - May involve both surgical and non-surgical treatment options
- DSRCT has a shared oncogene activation pathway with Ewing sarcoma
- Ewing sarcoma data used to design treatment plan for our patient
- According to the first intergroup Ewing sarcoma study (IESS-1), Vincristine, Dactinomycin, Cyclophosphamide, and Doxorubicin were associated with better five-year relapse-free survival (60% vs. 24%)³
- Beneficial five-year relapse-free survival with the addition of Ifosfamide and Etoposide alternating with the addition of Dactinomycin, Cyclophosphamide, and Doxorubicin backbone (69% vs. 54%)⁴
- Our patient's adjuvant treatment: seven alternating cycles of the above chemotherapies

CONCLUSION

- Desmoplastic Small Round Cell Tumor is an aggressive mesenchymal neoplasm typically found in the abdominal cavity with few cases reported in the head and neck
- Nodal metastasis is a poor prognostic indicator
- DSRT has no accepted treatment protocol, but has a shared oncogene activation pathway with Ewing Sarcoma
- Our patient underwent surgery with adjuvant chemotherapy based on the Ewing Sarcoma data, the outcome of which will hopefully shed light on how to treat this rare, aggressive disease in the head and neck

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