



Primary desmoplastic small round cell tumor of upper cervical lymph nodes

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Desmoplastic small round cell tumors (DSRCT) are rare malignancies that typically arise in the abdominopelvic cavities. They are very uncommon in the head and neck region. We present a case of an 11-year-old Caucasian male with a primary cervical lymph node tumor in the neck. Fine-needle aspiration cytology, histopathologic examination, immunohistochemical staining, and molecular genetic testing led to the diagnosis of DSRCT. Due to the very limited number of cases reported and the lack of staging criteria, the preferred management approach remains uncertain. (*Oral Surg Oral Med Oral Pathol Oral Radiol* 2015;120:e4-e10)

Desmoplastic small round cell tumors (DSRCT) are uncommon and highly aggressive tumors that usually arise in the abdominopelvic cavities. DSRCT usually affect young adults, with a striking male predominance.^{1,2} These tumors are very rare in the head and neck region, and to the best of our knowledge, this is the first case of a primary DSRCT involving the cervical lymph nodes reported in the English literature. Most cases of the head and neck involve the major salivary gland. Most cases involve the major salivary gland.³ A very unusual presentation of primary DSRCT involving the upper cervical lymph nodes, including the cytology, histopathology, immunohistochemistry, and molecular analysis that are characteristic for this disease, is discussed here. A literature review of head and neck DSRCT is summarized in [Table I](#).³⁻⁹

CASE HISTORY

We present the case of an 11-year-old, previously healthy Caucasian male who presented with a left neck mass of 4 months' duration. The mass was gradually increasing in size, reaching 4.0 × 4.0 cm. It was located in the left upper cervical region just below the arch of

the mandible. No other masses were palpable. Imaging including computed tomography (CT) of the thoraco-abdomino-pelvic region, and whole body positron emission tomography (PET) confirmed no tumors elsewhere. Fine-needle aspiration cytology (FNAC) was performed, followed by excisional biopsy of the involved cervical lymph node. A panel of immunohistochemical staining and molecular analysis was carried out confirming the diagnosis of a primary upper cervical lymph node DSRCT.

The patient was started on Euro-Ewing's 99 trial (localized disease protocol) of chemotherapy. The therapy consisted of vincristine (1.5 mg/m²/d), ifosfamide (3.0 g/m²/d), doxorubicin (20 mg/m²/d), etoposide (150 mg/m²/d), and mesna (1.0 g/m²/d I.V push before ifosfamide + 3.0 g/m²/d intravenous infusion). He received four cycles of chemotherapy. Follow-up magnetic resonance imaging (MRI) of the neck showed disease regression. The patient underwent adenoidec-tomy and left tonsillectomy, and no evidence of malignancy was observed. Thereafter, the patient underwent left modified radical neck dissection which revealed the following:

- Level 1: 3/3 positive lymph nodes.
- Level 2: 3/3 positive nodes.
- Level 3: 3/14 positive nodes.
- Level 4: No lymph nodes involved.

This was followed by external beam therapy (EBT) for a period of 48 days. Photons with energy of 6 MV were applied, based on three-dimensional conformal planning therapy with CT guidance. He received a total dose of 59.4 gray (Gy), fractionated to 33 fractions, and 1.8 Gy per fraction. The patient is currently on regular follow-ups and has been disease free for the past 39 months.

Imaging

Initial CT of the head and neck showed multiple enlarged lymph nodes in the cervical region, the largest lymph node measuring 2.4 cm in the left level II-B ([Figure 1](#)).

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Table 1. Literature review of head and neck desmoplastic small round cell tumors

| Author | Age | Gender | Nationality | Clinical features | Site | Size | Histopathology | Immunostaining features (Positive) | Molecular analysis | Follow-up | Treatment |
|-----------------------------------|-----|--------|-------------|----------------------------------|---|----------|---|--|--|---|--|
| Faras et al. (current case) | 11 | M | Kuwaiti | Gradually growing mass ×4 months | Left upper cervical lymph node | 4 × 4 cm | 1. Cohesive islands and nests separated by fibrocollagenous bands 2. Vesicular chromatin patten in nuclei 3. Scanty cytoplasm | CK (AE1/AE3) Pankeratin EMA Vimentin NSE Desmin | FISH: EWSR1 rearrangement RT-PCR: EWS–WT1 gene fusion | 39 months no EDR | Chemotherapy + left modified radical neck dissection + radiotherapy |
| Pang et al. ³ | 41 | M | Chinese | Mass | Left SMG | 5 cm | 1. Nests of tumor cells separated by desmoplastic stroma 2. Cells show fine granular chromatin with scanty cytoplasm | CK (AE1/AE3) Desmin WT1 EMA CD56 | FISH: EWSR1 rearrangement RT-PCR: EWS–WT1 gene fusion | 5 weeks postoperatively patient died | Left submandibular gland excision + radical ipsilateral neck lymph node dissection |
| Santos Gorjon et al. ⁴ | 36 | M | NS | Pain and swelling ×8 months | Right SMG | 4 × 3 cm | 1. Large nests of round cells 2. Areas of central necrosis | CAM 5.2 EMA Vimentin Desmin EGFR ENE P53 NM23 | RT-PCR: EWS–WT1 gene fusion | 10 months no EDR | Right submandibular gland excision + radiotherapy |
| Yin et al. ⁵ | 24 | M | NS | Rapidly growing mass ×1 month | Right SMG | 4 cm | 1. Variable-sized nests in a desmoplastic stroma 2. Tumor cells are medium-sized with hyperchromatic nuclei and moderate eosinophilic cytoplasm 3. Coagulative necrosis | CK (CK8/CK18) NSE Desmin P53 | FISH: EWSR1 rearrangement RT-PCR: EWS–WT1 gene fusion | 7 month no EDR | Right submandibular gland excision + chemotherapy + radiotherapy |
| Cho et al. ⁶ | 26 | M | Korean | Mass ×10 months | Left SMG | 4 × 3 cm | 1. Small round triangular islands of small undifferentiated cells 2. Abundant collagenous stroma | CK Vimentin Desmin NSE | RT-PCR: EWS–WT1 gene fusion | 25 months postoperatively patient died | Left submandibular gland excision + neck dissection + radiotherapy + chemotherapy |
| Mihok et al. ⁷ | 16 | M | NS | Firm mass ×5 days | Medial left supraclavicular lymph node (metastasis) | 3.5 cm | 1. Irregularly shaped nests separated by fibrotic desmoplastic stroma 2. Necrotic foci (occasional) 3. Cells with prominent nucleoli and moderate amount of pale cytoplasmic vacuoles | CK cocktail NSE Desmin | NS | 7 months postoperatively, evidence of metastasis | Surgical excision + chemotherapy |

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