Case Report

Teratocarcinosarcoma of the tongue with bilateral neck nodal metastases

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When confronted with a tongue and neck mass in a child, a specific diagnosis can be difficult to make with a differential diagnosis often provided. Teratocarcinosarcoma of the tongue with metastases to the neck would be considered an extremely rare specific diagnosis to correctly make prospectively. This study presents the clinical appearance, as well as the CT, MRI, PET and pathologic findings in this rare entity. With this information and constellation of findings, it is believed that this rare entity should be included in the differential diagnosis.

CASE REPORT

A 14 year old male presented with 4 months history of an enlarging tongue lesion and enlarging right neck mass. Physical examination revealed a focal fullness of the right side of the tongue with an apparent submucosal mass, but no mucosal abnormality. A 12 cm right neck mass extruded through the skin (image G) with several additional enlarged multilevel cervical nodes bilaterally. CT (Image 1), MRI (Image 2), and PET/CT (Image 3) confirmed the right neck mass (long arrow), tongue mass (short arrow) and left neck nodes (double arrows). Bilateral modified neck dissection, right hemiglossectomy and superficial parotidectomy were performed.

Pathology revealed metastatic teratocarcinosarcoma in 23 of 44 nodes and the parotid. The largest nodal metastasis was 12 cm, with extranodal extension (Image 3F). Teratocarcinosarcoma formed a 5.2 × 3.0 × 2.7 cm mass in the tongue musculature. Left level I-V lymph node dissection revealed metastatic teratocarcinosarcoma in 6 of 26 nodes: (3 of 5 level IIA, 3 of 17 level III); largest 2.8 cm level IIA, with extranodal extension.

There was metastatic teratocarcinosarcoma in a single left retropharyngeal lymph node. The tumor demonstrated a mixture of heterogeneous elements including undifferentiated carcinoma, undifferentiated spindle cell sarcoma, primitive neuroepithelial elements, differentiating ganglioneuronal/glial elements (Image 4A to E). The morphologic and immunophenotypic features were consistent with teratocarcinosarcoma. The patient completed treatment with Cisplatin and 27 of 30 fractions radiotherapy (5400 of 6000 cGy). PET/CT demonstrated widespread metastases including neck, mediastinum, lumbar spine, and iliac region. The patient was lost to follow-up 16 months after presentation.

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Image 1. Neck mass on contrast enhanced CT.

Image 2. MRI of right tongue mass (long arrow), right neck mass (short arrow) and left neck nodes (double arrows).

Image 3. PET/CT of right tongue mass (long arrow), right neck mass (short arrow) and left neck nodes (double arrows).
DISCUSSION

Teratocarcinosarcoma arises from pluripotential stem cells, often with an immature appearance, explaining its heterogeneous microscopic appearance. There are benign neural elements and various benign and malignant epithelial and mesenchymal components (Smith et al., 2008). There are portions of squamous cell or adenocarcinoma with sarcomatous components of spindle cell, smooth muscle, skeletal muscle, cartilage, and/or bone origin (Mondal et al., 2012). Treatment includes a combination of surgery, chemotherapy and radiation. Because the tumors consist of multiple malignant cell lines, chemotherapy should be individualized to the specific pathology present (Nitsche et al., 2005). The rare presentation and varied combination of histopathological features make it difficult to choose an optimal treatment strategy (Budrakkar et al., 2010). Teratocarcinosarcoma is an aggressive neoplasm with a poor prognosis (46% 5 year survival). Lesions typically arise in the sinonasal tract or nasopharynx, becoming symptomatic after invading surrounding tissues. As such, they present at an advanced stage (Budrakkar et al., 2010), typically in adult males. Most commonly, progression is due to local/regional failure, with distant and nodal metastasis less common. Behavior varies depending on the type of carcinoma and sarcoma within the tumor. This case is unique in that it represents an uncommon presentation of an already rare tumor due to the patients’ young age, the atypical location of the primary lesion, oral cavity and the unusual clinical presentation (early aggressive nodal and distant metastasis). There are only 3 prior reports in patients under 18, including the sphenoid sinus in a 10 year old (Budrakkar et al., 2010), oral cavity in a 10 year old (Crazzolara et al., 2004), and nasal cavity in a 15 year old (Agrawal et al., 2012). This case is also important in that it demonstrates the importance of performing high quality biopsies in multiple locations in a heterogeneous tumor. While heterogeneity can be related to necrosis, tissue invasion, or other factors; complex lesions such as this often require multiple aggressive biopsies for diagnosis and timely optimal management.

Conflict of interest

The authors have not declared any conflict of interest.

REFERENCES

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