International Journal of Otorhinolaryngology and Head and Neck Surgery
Shetty SS et al. Int J Otorhinolaryngol Head Neck Surg. 2018 Jan;4(1):276-277
http://www.ijorl.com
pISSN 2454-5929 | eISSN 2454-5937

DOI: http://dx.doi.org/10.18203/issn.2454-5929.ijohns20175641

Case Report

Primitive neuroectodermal tumor of thyroid: a rare presentation

Shravan S. Shetty*, Dhairyasheel N. Savant, Hozefa Lokhandwala, Akanksha Chichra

Department of Surgical Oncology, Somaiya Ayurvihar Asian Cancer Institute, Mumbai, Maharashtra, India

Received: 06 September 2017
Revised: 06 December 2017
Accepted: 07 December 2017

***Correspondence:**
Dr. Shravan S. Shetty,
E-mail: drshravanshetty@gmail.com

ABSTRACT

Extraskeletal Ewing's sarcoma (EES) commonly arises in the soft tissues of trunk or extremities. EES is rare in the head and neck region; most having been documented in nasal cavity, paranasal sinuses and neck. Head and neck PNETs have an intermediate prognosis. We report the case of a 12 year old boy who presented with primitive neuroectodermal tumor of the thyroid and was treated with multimodal treatment including surgery, chemotherapy and radiotherapy. The patient is alive and fit with a functional larynx. Major drug regimens use vincristine, doxorubicin, ifosfamide or cyclophosphamide, dactinomycin and/or etoposide. Complete surgical excision is undertaken whenever possible to improve long-term survival. However, the relative radiosensitivity of tumors of the Ewing family, suggest multimodal treatment including adjuvant radiotherapy in case of positive margins or poor response to chemotherapy rather than resection with 2-3 cm margins, which would imply laryngeal sacrifice for thyroid tumors.

Keywords: Extra skeletal, Ewing's sarcoma, Thyroid, Multimodal

INTRODUCTION

Primitive neuroectodermal tumors (PNETs) belong to the Ewing family of tumors and represent 1% of sarcomas.[1,2] Ewing's family of tumor is the second most common primary bone malignancy of childhood. Extra skeletal Ewing's sarcoma is rare.[3] Head and neck PNETs have an intermediate prognosis between abdominopelvic disease (who fare much worse) and extremities.[4] Ewing's thyroid cases mostly occur in patients ≤30 years.

CASE REPORT

A 12 year old boy presented with a short history of cough and on & off nasal blockage. On examination, the patient had a good performance status with clinically palpable thyroid swelling and no cervical lymphadenopathy. PET CT showed hypermetabolic lesion (SUV max 9.3) involving thyroid gland suggestive of primary neoplasm, hypermetabolic bilateral level II nodes, posterior nasopharynx and bilateral tonsillar areas suggestive of inflammatory etiology and no evidence of FDG avid lesion elsewhere (Figure 1). CT guided biopsy of thyroid revealed malignant round cell tumor. The presence of strong membrane expression of CD 99 and nuclear expression of ERG suggested that this is a primitive neuroectodermal tumor with a possible EWS/ERG translocation. The possibilities considered were metastatic undifferentiated carcinoma from a possible primary in nasopharynx (in view of strong cytokeratin and p63 positivity and high uptake in the nasopharynx and tonsil on PET) and primitive neuroectodermal tumor/Ewing's sarcoma (in view of MIC2 positivity, although Fli-1 is negative). On FISH test, EWSR1 gene rearrangement was not detected. The patient received 4 cycles of chemotherapy (Vincristine, Ifosfamide, Doxorubicin and Etoposide) as per EURO-EWING protocol. Post 3 cycles of chemotherapy PET-CT showed good response (SUV max 6.22) in the primary and no new lesions elsewhere in the body (Figure 2). He

underwent total thyroidectomy with central compartment clearance and bilateral neck dissection (Level II-IV) and tracheal resection. Intraoperatively retrosternal extension was noted and the tumor was infiltrating in to the anterior wall of trachea and encasing the left recurrent laryngeal nerve. Both the recurrent laryngeal nerves were preserved and the trachea was closed primarily after resection of three rings. Postoperatively the patient developed hypocalcaemia which was managed conservatively. He was discharged with an intact voice. Histopathology report revealed 60% residual viable PNET involving both lobes of thyroid, LVE+, 1/6 perithyroidal nodes positive and 1/10 right neck nodes positive. Immunohistochemistry studies revealed FLI-1+/CD 99+ which confirmed the diagnosis of primitive neuroectodermal tumour. Post-surgery, the patient received 9 more cycles of adjuvant chemotherapy and adjuvant radiotherapy. The patient is alive and fit with a functional larynx.

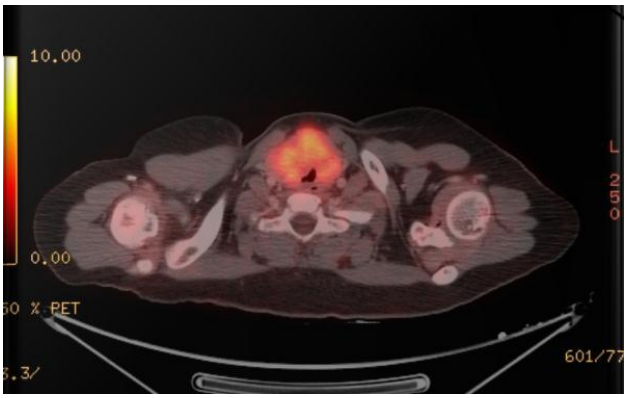

Figure 1: PET CT (pre chemotherapy): hypermetabolic lesion involving the thyroid gland (SUV Max 9.2) and bilateral neck nodes.

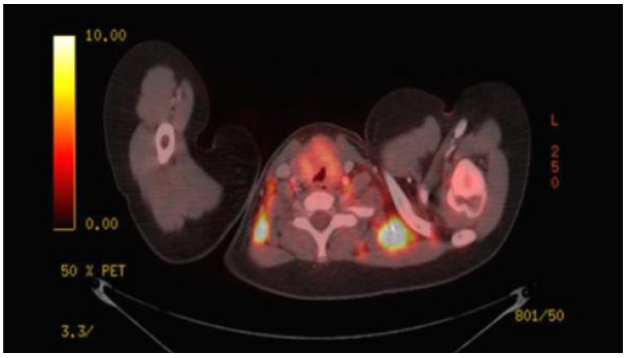

Figure 2: PET CT (post chemotherapy): good response in the primary (SUV Max 6.22).

DISCUSSION

Extraskeletal Ewing's sarcoma (EES) commonly arises in the soft tissues of trunk or extremities.[3] Ewing and PNET sarcomas share a similar histological appearance of small round blue cell tumor, immunounohistochemical markers, cytogenetic translocation t (11;22) (q24;q12) and MIC 2 gene expression (both present in more than 90% of cases). An absence of neural differentiation supports a diagnosis of Ewing's sarcoma rather than PNET. Diagnosis is based on immunostaining with at least 2 neural markers, ultrastructural examinations and evidence of an abnormal t (11;22)(q24;q12).

Based on a review of Ewing's sarcoma (bone or soft tissue, head and neck cases), the optimal treatment of PNETs consists of neoadjuvant chemotherapy, surgery and radiation based both on response to neoadjuvant chemotherapy and tumor site. Complete surgical excision is undertaken whenever possible to improve long term survival.[5] Radiotherapy is indicated in Ewing sarcomas whenever there are close or positive margins or poor response to chemotherapy (necrosis<90%). It is associated with improved local control and improves survival. Chemotherapy (neoadjuvant or sequential) has predominant part in the multimodal treatment of Ewing's sarcoma. Multimodal treatment yield five year survival rates of about 60% (52-88%) as reviewed in series including atleast one case of Ewings sarcoma / PNET of the head and neck.[6,7]

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Kim E, Bae TS, Kwon Y, Kim TH, Chung KW, Kim SW, et al. Primary malignant teratoma with a primitive neuroectodermal tumor component in thyroid gland: a case report. J Korean Med Sci. 2007;22:568-71.
2. Adapa P, Chung TW, Popek EJ, Hunter JV. Extraosseous Ewing sarcoma of the thyroid gland. Pediatr Radiol. 2009;39:1365-8.
3. Rick A, Brian M, Mary D, Bruce R. Extra skeletal Ewing's sarcoma. Cancer. 1999;85:725-31.
4. Kimber C, Michalski A, Spitz I, Pierro A. Primitive neuroectodermal tumors: anatomic location, extent of surgery and outcome. J Pediatr Surg. 1998;33:39-41.
5. Craver RD, Lipscomb JT, Suskind D, Velez MC. Malignant teratoma of the thyroid with primitive neuroepithelial and mesenchymal sarcomatous components. Ann Diagn Pathol. 2001;5:285-92.
6. Allam A, El-Husseiny G, Khafag Y, Kandil A, Gray A, Ezzat A, et al. Ewing's sarcoma of the head and neck : a retrospective analysis of 24 cases. Sarcoma. 1999;3:11-5.
7. Elomaa I, Blomqvist C, Saeter G, et al. Chemotherapy in Ewing's sarcoma. The Scandinavian sarcoma group experience. Acta Orthop Scand Suppl. 1999;285:69-73.

Cite this article as: Shetty SS, Savant DN, Lokhandwala H, Chichra A. Primitive neuroectodermal tumor of thyroid: a rare presentation. Int J Otorhinolaryngol Head Neck Surg 2018;4:276-7.