CASE REPORT

Malignant rhabdoid tumor of the tongue: A rare occurrence

Munlima Hazarika, Tashnin Rahman[1], Anupam Sarma[2], Manigreeva Krishnatreya[1]
Department of Medical Oncology, [1]Head and Neck Oncology, [2]Pathology, Dr. Bhubaneswar Borooah Cancer Institute, Guwahati, Assam, India

Address for correspondence:
Dr. Munlima Hazarika,Assistant Professor, Department of Medical Oncology, Dr. Bhubaneswar Borooah Cancer Institute, Guwahati - 781 016, Assam, India.
E-mail: drmunlimahazarika@yahoo.com

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ABSTRACT

Malignant rhabdoid tumors (MRTs) are highly aggressive neoplasms that most commonly occur in the kidneys of young children. Malignant rhabdoid tumor of the tongue is an extremely rare entity and very few have been reported in the literature. The course of extra-renal MRT is short and its prognosis is very poor. A 19-year-old female presented with a progressive swelling and restricted mobility of the tongue for over 3 months duration. We present here a locally advanced case of MRT of the tongue, its diagnosis, management and review of the literature related to it.

Key words: Extra-renal rhabdoid tumor, malignant rhabdoid tumor, tongue

INTRODUCTION

Malignant rhabdoid tumors (MRTs) are highly aggressive neoplasms that most commonly occur in the kidneys of young children. A few cases of primary MRT occurring in extra-renal sites have been reported, particularly in the soft tissues. Malignant rhabdoid tumor was first described by Beckwith and Palmer in 1978 as a rhabdomyosarcomatoid variant of Wilms tumor.[1] But due to the absence of ultra-structural or immunohistochemical evidence of myogenic differentiation, the term rhabdoid was later adopted for these neoplasms.[2,3] Malignant rhabdoid tumors usually occur during infancy and the median age is about 11 months.[4] However, MRTs have been reported in patients from the age of 3 weeks to 50 years.[5-8] The tumors have a male preponderance with a male to female ratio of 1.5:1.[7] The course of extra-renal MRT is short and its prognosis is very poor. The 5-year survival without tumor recurrences are less than 50% regardless of the therapy used.[9-11] We present here a case of locally advanced case of MRT of the tongue.

CASE REPORT

A 19-year-old girl presented to our outpatient department with a swelling of the tongue of 1-year duration, which was initially slowly growing but later started progressing rapidly in the past 3 months. She was complaining of inability in the movement of the tongue leading to difficulty in speaking and chewing of food. This also resulted in nasal regurgitation of food. She was not suffering from any co-morbid conditions.

On examination, her general health was fair (Eastern Cooperative Oncology Group performance status was 1). On intra-oral examination, there was a lobulated swelling involving the entire tongue with an intervening ulcer and there was restriction of the tongue movement. Multiple bilateral level 1A and 1B nodes of varying sizes were palpable and which were fixed to the underlying structures. The largest node was around 3 cm in size, which was on the left side (clinical staging was T4bN2cM0).

Examination of the skin, abdomen, respiratory system, cardio vascular and the central nervous systems were found to be normal. Magnetic resonance imaging (MRI) of the tongue showed an enhancing mass of more than 5 × 4 cm in its greatest dimension. The mass involved both sides of intrinsic as well as extrinsic muscles with loss of definition of lingual septum [Figure 1a]. Extension of the mass was seen into the right-sided sublingual space with encasement of the neurovascular bundle which was also extending anteriorly to the tip of tongue. There was no bony erosion. No extensions were seen into the carotid and bucco-masticator spaces. Magnetic resonance imaging of the neck showed metastatic cervical lymphadenopathy at the level I, II, III, IV on the left and level II on the right [Figure 1b]. Clinically, the diagnosis of the lobulated swelling with an ulcer on the tongue, was that of an infiltrative epithelial malignancy or a minor salivary gland malignancy. Limitation of punch biopsy in this case was the presence of a limited intervening area with ulceration on the tongue swelling, which may not be representative of the presence of definite malignant cells. However, punch biopsy from the ulcerative lesion showed solid sheets of moderately

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pleomorphic tumor cells with round to oval, central to eccentric nuclei and eosinophilic cytoplasm suggestive of undifferentiated malignant neoplasm with features of rhabdoid differentiation [Figure 2a]. Immunohistochemistry (IHC) analysis was done to detect the expression of vimentin, desmin, pan cytokeratin (CK) and S-100 protein. Immunohistochemistry was positive for vimentin [Figure 2b] but negative for the expression of desmin, pan CK and S-100. The diagnosis of MRT was established after histopathological examination and IHC analysis. In this case, the patient was not considered for upfront surgery in view of the growth involving the whole of the tongue extending up to the vallecula; hence, there would be an absence of clear margins for resection. The patient was treated with six cycles of palliative chemotherapy with doxorubicine (50 mg/m^2, D1) and ifosphamide (1500 mg/m^2, D1–D5). Palliative chemotherapy was followed by palliative external beam radiotherapy (EBRT). There was no response to the treatment after completion of chemo-radiotherapy and the patient expired after 3 years from the date of diagnosis.

DISCUSSION

Rhabdoid tumor was first described in the kidneys with a poor prognosis.[1] Over the past several years, research have shown that it can also occur in the central nervous system (brain and spinal cord), as well as other locations outside the kidneys such as the liver, muscle, heart, lung, soft tissues, skin, uterus and the thymus.[5-8,12] Tongue is an extremely rare site. To the best of our knowledge, there have been two case reports of MRT of the tongue in the English literature. Patron *et al.* have reported MRT of the tongue in a 10-day-old male child.[13] Another case of MRT of the tongue was reported by Tomasovic *et al.* in an 18-year-old female.[14]

Extra-renal MRT develops mostly in children with a slight male preponderance.[7] The cases have been reported from new born to teenagers and adults.[5-8] In our case, the patient was a 19-year-old female who presented with indurations and swelling of the tongue for a duration of 1 year. The clinical presentation of our case was similar to the case reported by Tomasovic *et al*.[14]

In the histology of MRT polygonal and eosinophilic cells with globular hyaline inclusion, open nuclei and prominent nucleoli are the typical findings, but to establish the diagnosis, clinical evaluation and histopathological feature along with ultra-structural and IHC findings are necessary. Histopathology of MRT can be heterogeneous, so the differential diagnosis can be poorly differentiated carcinoma, melanoma, anaplastic large-cell lymphoma and rhabdomyosarcoma. In this case, histopathology from the tongue lesion showed sheets of neoplastic cells with abundant eosinophilic cytoplasm and eccentric mildly irregular vesicular nuclei with lobulation. Most of the cells showed vimentin immunoreactivity and it was negative for the expressions of desmin and S-100. To arrive at a definitive diagnosis in this case, we had to take the help of IHC. Possibility of adenocarcinoma, anaplastic large-cell lymphoma, melanoma and rhabdomyosarcoma were ruled out by the IHC findings. As pan CK was negative, the diagnosis of adenocarcinomas and other epithelial tumors were excluded, the diagnosis of lymphoma was excluded by vimentin positivity and so, further lymphoma IHC study was not carried out. Melanoma was excluded due to S-100 negativity and finally rhabdomyosarcoma diagnosis was excluded based on morphology [histopathology (HPE)] by the absence of strap cells and cross-striations, which are seen in rhabdomyosarcoma; furthermore, this case was negative for desmin and CK. So, in the light of morphology and IHC study, rhabdomyosarcoma was excluded.

The imaging characteristics of soft-tissue rhabdoid tumors are yet to be determined. No specific imaging features were observed that could prove diagnostic for these tumors.[12,15] However, these tumors show a heterogeneous hyperintensity on T2-weighted (W) MRI.[15] This was a consistent finding in our case as well.

Extra-renal MRT demonstrates a rapidly progressive evolution, with metastasis occurring in most patients from 2 to 15 months after diagnosis.[16,17] In some of the reported cases, there were disseminated disease to the lungs, lymph nodes and liver at the time of diagnosis, which are the common sites of metastasis.[18,19] In our case, the patient received palliative chemotherapy and

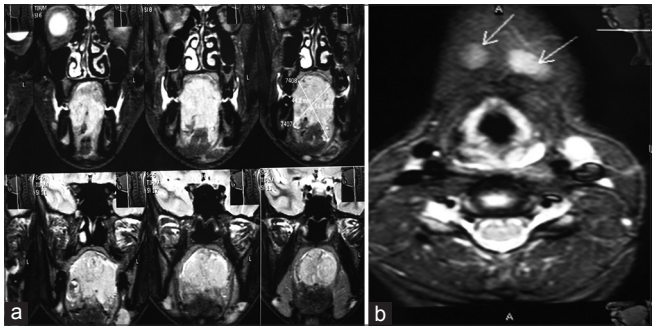

Figure 1: (a) Magnetic resonance imaging (MRI) of the tongue showing heterogeneous hyper intensity on T2-weighted (W) image. (b) Magnetic resonance imaging of the neck showing metastatic cervical lymphadenopathy

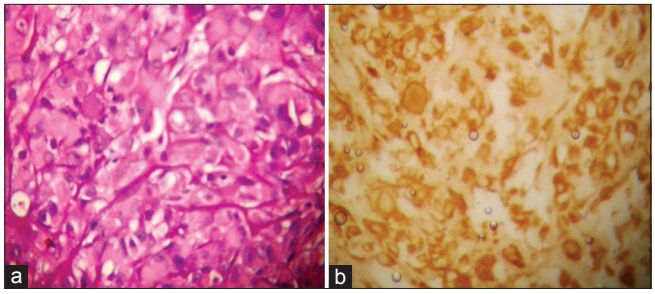

Figure 2: (a) Photomicrograph showing solid sheets of moderately pleomorphic tumor cells with round to oval and central to eccentric nuclei along with an eosinophilic cytoplasm (H&E stain, x400). (b) Photomicrograph showing tumor cells to be positive for vimentin (IHC stain, x400)

EBRT. There was no response to chemo-radiotherapy in this case, but the patient survived for a considerable period of time, unlike the poor survival of paravertebral rhabdoid tumors seen in infancy as reported by Lynch *et al*.[18]

In conclusion, authors would like to add this extremely rare case of MRT of the tongue to the literature. Malignant rhabdoid tumor of the tongue poses a therapeutic challenge to the oncologist and no consistently effective regimen has yet been established for the treatment of MRT. Although MRTs are rare, extra-renal rhabdoid tumor should be considered in the differential diagnosis of tumors of the tongue.

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