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Rajjyoti Das, Anupam Sarma, Partha Sarathi Chakraborty, Jagannath Dev Sharma, Amal Chandra Kataki

ABSTRACT

Introduction: Malignant triton tumor (or malignant Schwanoma with Rhabdomyoblastic differentiation) is a very rare entity with poor prognosis. It isconsidered a high grade malignantneoplasm with poor outcome. In 70% of cases the neoplasm is associated with von Recklinghausen neurofibromatosis and in the remaining 30% it is the sole morbid finding. Local recurrence is frequent and distant metastasis preferentially situated in lung and brain. Surgery is the treatment of choice and post operative radiotherapy is always indicated. Histopathology and immunohistochemistry are helpful in diagnosis. We report clinical course, therapeutic approach, histopathology and immunohistochemistry of such a case.

Case Report: A 19-year-old female presented in Dr B Borooah Cancer Institute, Guwahati with a complaint of swelling in the right side of the cheek with ulceration in the oral cavity. On clinical examination there was a round swelling in left cheek of 4x4 cm size with overlying normal skin. Computed tomography scan revealed a hypo dense lesion in the right maxillary sinus with erosion of the floor. Punch biopsy was taken from palatal growth and sent for histopathological examination which revealed a malignant neoplasm with sarcomatoid features composed of spindle cells. Immunohistochemical stains showed tumor cells positive for Vimentin, S-100 protein and, Desmin. Patient was taken up for surgery followed by radiotherapy.

Conclusion: Malignant triton tumor of maxilla is extremely rare. The diagnosis must be based upon imaging study, histopathology and immunohistochemical features. Considering the aggressive nature of disease radical surgery should be followed by adjuvant chemoradiation.

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Keywords: Malignant triton tumor (MTT), Surgery, Histopathology and immunohistochemistry

Rajjyoti Das[1], Anupam Sarma[2], Partha Sarathi Chakraborty[3], Jagannath Dev Sharma[4], Amal Chandra Kataki[5]
Affiliations: [1]MBBS, MS, Assistant Professor, Department of Head & Neck Oncology, Dr. B. Borooah Cancer Institute. Guwahati, Assam, India; [2]MBBS, MD, Assistant Professor, Department of Pathology, Dr. B. Borooah Cancer Institute, Guwahati, Assam, India; [3]BDS, MDS, P.G. Fellow, Department of Head & Neck Oncology, Dr. B. Borooah Cancer Institute, Guwahati, Assam, India; [4]MBBS, MD, Professor & Head, Department of Pathology, Dr. B. Borooah Cancer Institute, Guwahati, Assam, India; [5]MBBS, MD, Director & Head, Department of Gynaecologic Oncology, Dr. B. Borooah Cancer Institute. Guwahati, Assam, India.
Corresponding Author: Dr. Rajjyoti Das, Department of Head & Neck Oncology, Dr. B. Borooah Cancer Institute.Assam, Guwahati -78016, India; Ph: +91-9435087642; Email: dr_rajjyoti@yahoo.in

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INTRODUCTION

Malignant peripheral nerve sheath tumor (MPNST) accounts for about 5–15% of all soft tissue sarcomas [1]. Malignant Triton Tumor (MTT) constitutes about 5% of all MPNSTs [2]. Common sites for MTT include head, neck, extremities and trunk [1–3]. This rare tumor with fewer cases reported in literature, generally affects adult patients but can also occur in children [4]. Morphology

and immunohistochemistry are helpful in diagnosis. A 5-year survival is low up to 10–20% indicating the high malignant nature of the disease [5]. Surgical resection of the tumor is thought to be best but due to its aggressive nature adjunct chemotherapy and radiotherapy may increases the survival [6, 7].

CASE REPORT

A 19-year-old female reported in the head and neck oncology out patient department with a complaint of swelling in the right side of the cheek since one and half months, with ulceration in the oral cavity since 25 days and pain in the right upper teeth with occasional bleeding from the mouth. On general examination she was average built. Respiratory and cardiovascular examination was normal. On ear, nose and throat clinical examination there was a round swelling in right cheek of 4x4 cm size with overlying normal skin. On palpation tenderness was present over the swelling with normal sensory finding. Intra oral examination revealed an irregular surface ulcer in the right side of hard palate with loose premolar and molar tooth. Neck examination revealed no cervical lymphadenopathy. Indirect laryngoscopic examination was normal. All other hematological examination and chest radiogram were normal.

Computed tomography (CT) scan revealed a hypo dense lesion in the right maxillary sinus with erosion of the floor, there was enhancement of the lesion in contrast study (Figure 1). Punch biopsy was taken from palatal growth and sent for histopathological examination (HPE). The HPE revealed a malignant neoplasm with sarcomatoid features composed of spindle cells (Figure 2).The malignant cells showed hyperchromatic nuclei and pale cytoplasm. Differential diagnosis for spindle cell sarcoma includes malignant peripheral nerve sheath tumor (MPNST), spindle cell carcinoma, monophasic synovial sarcoma and spindle cell form of malignant melanoma. Immunohistochemical stains showed tumor cells positive for Vimentin, subsets of tumor cells positive for S-100 protein and another subset of tumor cells was positive for Desmin (Figures 3, 4).Immunohistochemistry was negative for CK, HMB-45 and chromogranin.

Patient was taken up for surgery. With a Weber-Ferguson incision right maxilla was approached and it was found that growth eroded the anterior maxillary wall and whole tumor was removed enblockincluding the involved palatal part. Tumor free margins was obtained at the time of surgical resection macroscopically and temporary palatal prosthesis was applied. Three weeks following surgery a final prosthesis was applied.

Specimen consisted of multiple fragments of bone, fibrous tissue and tumor mass which weighed16.3 gm measuring 5x4x3 cm. On cut surface it was composed of grey firm tissue with cystic spaces.The tissue was sent for histopathological examination. HPE confirmed Malignant Triton tumor. Bone and cut margins were

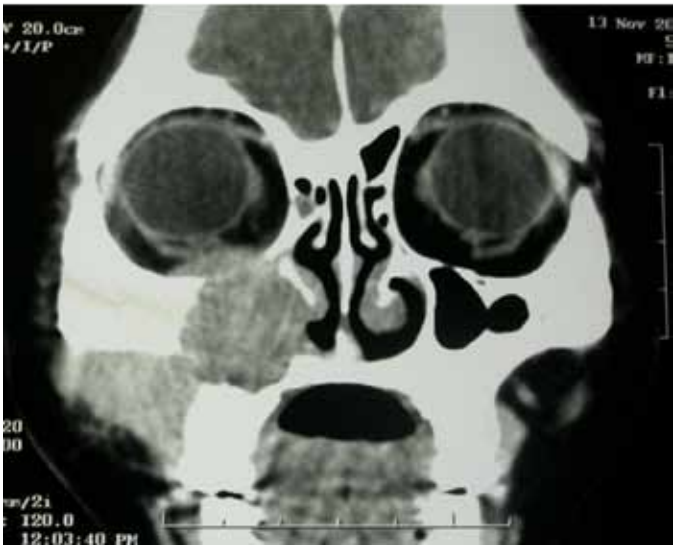

Figure 1: Computed tomography (CT) scan showing disease extension of the patient.

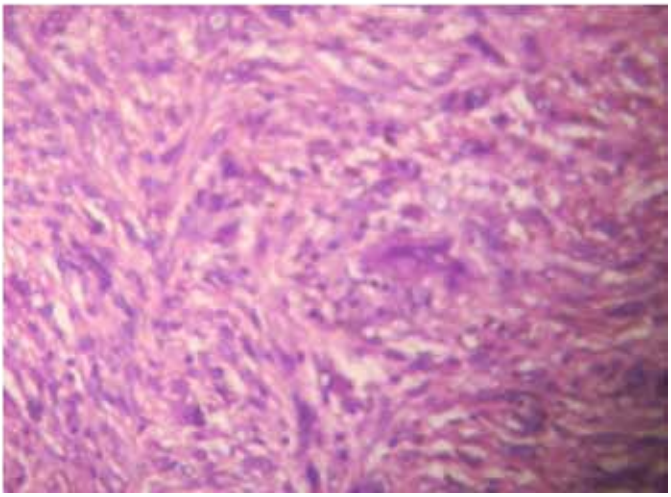

Figure 2: Photomicrograph showing sarcomatoid spindle shaped tumor cells with variably pleomorphic and hyperchromatic nuclei and mitosis. (H&E, 100X).

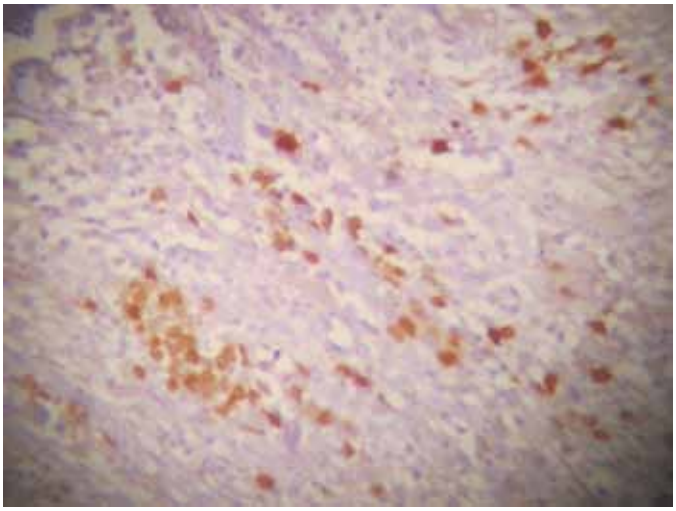

Figure 3: Immunohistochemical staining showing subsets of tumor cells are positive for S-100. (S-100,100X).

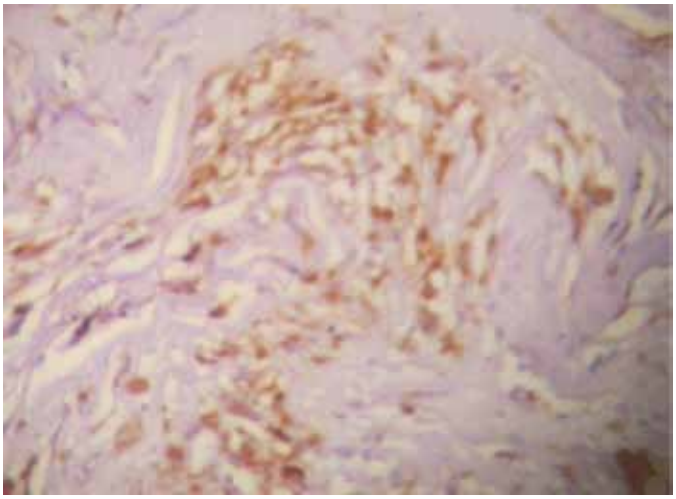

Figure 4: Immunohistochemical staining showing subsets of tumor cells are positive for Desmin, identifying muscle differentiation (Desmin, 100X).

free of tumor. Three weeks following surgery patient was subjected to radiotherapy (66 Gy in 33 fractions for 6 weeks) over maxillae including upper neck.

DISCUSSION

Masson first described a lesion that consisted of a malignant schwannoma with rhabdomyoblastic differentiation [8]. Subsequently, Woodruff et al. coined the term "Malignant triton tumor" to indicate such a neoplasm [9]. There are two types of tumor, sporadic or in association with NF-1. Those with von Recklinghausen neurofibromatosis constitute over 70% and displayed a marked male predominance, young age with a common presentation in head and neck. On the other hand, those without von Recklinghausen neurofibromatosis are mostly common in older age, female predominance and frequently located on trunk [10].

MPNST can be diagnosed on histopathology supported by positivity for S-100 protein. In morphology most areas show appearance of an extremely cellular spindle cell neoplasm with abundant mitoses. Although most tumors are quite monomorphic but heterologous elements like rhabdomyoblasts, cartilage and bone may be present in some cases. Such tumors show positivity for S-100 protein in 50-90% of cases, suggesting a nerve sheath origin. Rhabdomyoblasts are positive for immunostains such as desmin, myogenin and myo-D1 [11].

Though MPNST is non-radio-sensitive, like other sarcomas, wide local surgical excision followed by adjuvant radical radiotherapy is mostly adequate. Role of chemotherapy is still not been clearly defined. But chemotherapy might play some role in adjuvant setting in eradicating micro metastasis [6, 7].

MTT has poor prognosis with 5 years survival rate of only 5–15%, where as MPNST has a good survival of about 50-60% [12]. Poor prognosis of MTT depends on the location, grade and total radical resection [13]. Prognosis is good in head, neck, and extremities but very poor in buttock and other sites.

In spite of adequate treatment, local recurrence and distant metastasis rates approximate 25 and 48%, respectively. Survival of patients with head and neck MTT ranges between four months and 22 years [11]. Cytogenetic study has revealed some karyotypic changes associated with this tumor. Cytogenetic analysis has revealed some breakpoints which are considered as region for myogenic differentiation and probably responsible for rhabdomyoblastic differentiation [14]. Aggressive biologic behavior is may be due to amplification of c-myc oncogene [15].

CONCLUSION

Malignant triton tumor of maxilla is extremely rare. The clinical presentation may be misleading. The diagnosis must be based upon imaging study, histopathology and immunohistochemicalfeatures as it was in our case. Additional physical signs of NF-1 should also be excluded. Considering the aggressive nature of disease radical treatment of surgery should be followed by adjuvant chemoradiation.

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Author Contributions

Rajjyoti Das – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published
Anupam Sarma – Conception and design, Drafting the article, Critical revision of the article, Final approval of the version to be published
Partha Sarathi Chakraborty – Conception and design, Drafting the article, Critical revision of the article, Final approval of the version to be published
Jagannath Dev Sharma – Conception and design, Drafting the article, Critical revision of the article, Final approval of the version to be published
Amal Chandra Kataki – Conception and design, Critical revision of the article, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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ABOUT THE AUTHORS

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Rajjyoti Das is Senior Head & Neck Oncosurgeon at Dr B. Borooah Cancer Institute (Regional Cancer Center), Guwahati,Assam,India,with more than 10 years experience . His special interest in Microlaryngeal Laser & Reconstructive Oncosurgery. He is associated with DBT (India) project. He has few publications in National & International Journals. He intends to pursue post-doctoral fellowship in Head & Neck Oncology.

Anupam Sarma is Assistant Professor, Departrment of Pathology, Dr B. Borooah Cancer Institute, Guwahati,India. His area of interest is Oncopathology,Hemato-oncology,Molecular Pathology & Cancer Research. He intends to pursue some post-doctoral fellowship in the field of Oncopathology. He is associated with various research projects sponsored DBT & ICMR (Govt of India). He has more than 10 publications in International Journals.

Partha Sarathi Chakraborty is Fellow at Department of Head & Neck Oncology/ Dr B. Borooah Cancer Institute, Guwahati, Assam, India. His research interests includes Maxillofacial Oncology, He intends to pursue further training or fellowship in Head & Neck Microvascular Surgery in future.

Jagannath Dev Sharma is Professor & HOD in Department of Pathology at Dr B. Borooah Cancer Institute, Guwahati,India. He is also The Principal Investigator ,Cancer Registry, ICMR (Govt. Of India). His fields of interest are Tumor Pathology, IHC, Molecular Patholgy & Cancer Research. He has 22 international journal publications to his credit.

Amal Chandra Kataki is the Director of Dr. B. Borooah Cancer Institute (RCC), Guwahati, Assam,India, and Professor & Head of Department of Gynaecologic Oncology. He is actively involved in organizing MD & PhD programmes & Research on Molecular Biology of Cancer in addition to his routine clinical & administrative works. He is recipient of UICC fellowship. He has published 45 research papers in National & International journals.

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