

Round Cell Tumor of Infratemporal Fossa Presenting as Intraoral Mass

Mohan Baliga¹, Dani Mihir Tusharbhair^{2*}, Joanna Baptist³ and Aananya Mishra²

¹Professor, Department of Oral and Maxillofacial Surgery, Manipal College of Dental Sciences, Mangalore, Manipal Academy of Higher Education, India

²Postgraduate, Department of Oral and Maxillofacial Surgery, Manipal College of Dental Sciences, Mangalore, Manipal Academy of Higher Education, Mangalore, Karnataka, India

³Associate Professor, Department of Oral and Maxillofacial Surgery, Manipal College of Dental Sciences, Mangalore, Manipal Academy of Higher Education, Mangalore, India

***Corresponding Author:** Dani Mihir Tusharbhair Postgraduate, Department of Oral and Maxillofacial Surgery, Manipal College of Dental Sciences, Mangalore, Manipal Academy of Higher Education, Karnataka, India.

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Abstract

Malignant small round cell tumours represents a heterogeneous group of malignant neoplasms characterized by uniform, round to oval undifferentiated cells, present predominantly in children and young adults. It imposes a diagnostic challenge and their differential diagnosis is subjective to the age of the patient, their site of occurrence and their degree of differentiation. All the tumours types have specific treatment modalities and the prognosis is dependent on the exact histological diagnosis confirmed by the adjuvants like immunohistochemistry and cytogenetics.

Keywords: Malignant Small Round Cell Tumours (MSRCT); Malignant Neoplasms

Introduction

Malignant small round cell tumours (MSRCT) are group of tumours which predominantly pose a challenge in diagnosis due to their undifferentiated or primeval character, such group of tumours although rare in oral cavity are highly aggressive in nature, that are depicted by relatively small, round, and monotonous undifferentiated cells with a solitary hyper chromatic nucleus and increased nuclear-cytoplasmic ratio [1]. Among these group of tumours the most common ones include Ewing's family of tumours, non-Hodgkin's lymphoma, rhabdomyosarcoma, synovial sarcoma, peripheral neuroectodermal tumour, retinoblastoma, nephroblastoma and neuroblastoma [2]. Differentiating such lesions from others still remains an enigma, and accurate diagnosis requires a integration of immunophenotypic and genetic analysis along with immunohistochemistry. The role of Fine needle aspiration cytology (FNAC) is been highly debated in recent times [3].

Case Report

21 year old male patient reported with a gradually increasing painless growth with an ulcer in the centre, in the left retro molar trigone region since last 6 months. History revealed that the patient visited the family dentist for opinion for the ulcer for which he underwent extraction of left upper third molar. As the ulcer did not heal spontaneously after extraction, it was referred to our unit for further management. On examination a 3 x 2 cm non-tender fibrous mass involving the left retro molar trigone without any reduction in mouth opening was noted. There was a 1 x 1 cm ulcer over this fibrous mass (Figure 1). There was no facial asymmetry extra orally and condylar movements were within normal limits. Regional lymphadenopathy was present, involving level 1b, level 2, and level 5. The lymph nodes were palpable but non tender in nature.



Figure 1: Intraoral picture showing the lesion on left retro molar trigone area.

CT-scan revealed heterogeneously enhancing mass lesion extensive in nature in the left infra temporal region medial to the left ramus of mandible involving the left retro molar trigone measuring 5.4 x 4 x 5.8 cm (AP x TR x CC), anteriorly the lesion was causing cortical erosions of the alveolar arch of the left half of maxilla and abutting the lateral wall of left maxillary sinus. Posteriorly it was abutting the left internal carotid artery and external carotid artery along with causing compression of left internal jugular vein, medially the lesion was involving the left parapharyngeal space and laterally the lesion was infiltrating the ramus of left mandible and abutting the left condyle of mandible, while superiorly the lesion was infiltrating the left medial and lateral pterygoid muscle with cortical erosions of medial and lateral plate of pterygoid bone, inferiorly it was extending till the left submandibular gland. Multiple enlarged lymph nodes in level 1b, level 2 and level 5 with largest measuring 10 x 12 mm in left level 2 was noted (Figure 2).



Figure 2: Axial section showing the extent of the lesion.

USG abdomen, chest radiograph and routine investigations were within normal limits.

An incisional biopsy was done under local anaesthesia which on histopathological examination showed the presence of a hyperplastic stratified squamous epithelium, overlying a connective tissue stroma. The fibro-cellular stroma showed the presence of sheets of small, round to angulate, hyper chromatic and dis-cohesive cells with entrapped muscles, blood vessels and chronic inflammatory cell infiltrate suggestive of malignant round cell neoplasm (Figure 3). Immunohistochemistry was advised for further evaluation. Immunohistochemical staining was performed on the formalin-fixed, paraffin-embedded tissue sections. The tumour cells revealed strong immunoreactivity with anti-CD20 antibody (Figure 4) and were negative for CD3 and CD5 that confirmed it to be B cell lymphoma. The patient was referred to the medical oncologist for further management and during the presentation of this case patient had undergone two cycles of chemotherapy.

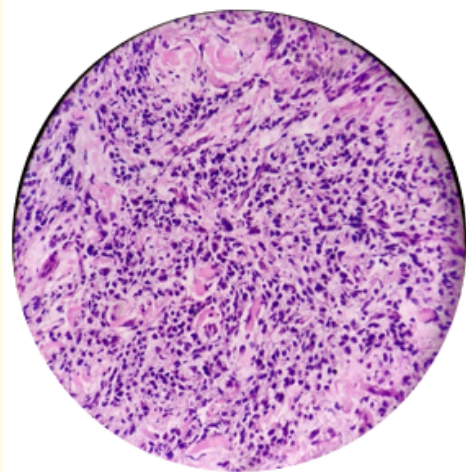


Figure 3: Histopathological picture.

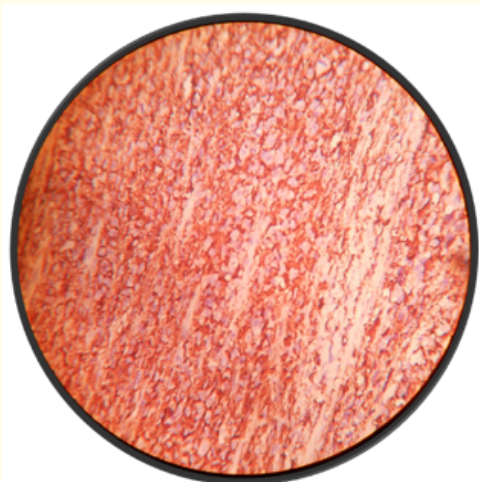


Figure 4: Immunohistochemical picture.

Discussion

Round cell tumours of oral cavity involves epithelial, mesenchyme, muscle, reticulo- endothelial, neural and varied tumours arising in soft tissue/bone. Also cytologically, these particular tumours consists of a meagre, basophilic cytoplasm with closely uniform population of oval to round cells, while the cells appear blue in colour due to existence of large hyper chromatic nuclei along with a thin rim of cytoplasm, it is also named as round blue cell tumour [4].

Lymphomas are malignant neoplasm of the lymphocyte cell origin that comprises of 50% to 59% of head and neck malignancies. They are mainly classified as either Hodgkin's or non-Hodgkin's lymphoma (NHL), among which non-Hodgkin's lymphomas (NHL) have a far greater predilection to disseminate to extra nodal site and they are much less predictable than Hodgkin's disease [5]. Lymphomas arising within the oral cavity covers less than 5% of all oral malignancies, of which 85% of lesions involve tonsils and hard palate [6]. Amongst these types diffuse B cell lymphomas (DLBCL) is the most predominant one seen. Solomides., *et al.* [7] in his study reported 68% of 71 cases as diffuse large B cell lymphoma (DLBCL). Van der Waal., *et al.* [8] in his series of 40 cases also showed that 50% were diffuse large B cell lymphoma (DLBCL). This neoplasm is characterized by a diffuse proliferation of large neoplastic B cells with nuclear size equal to or greater than normal macrophage nuclei, or sometimes it can be double the size of a normal lymphocyte, with variable clinical, morphologic, immunophenotypic, cytogenetic, and genetic features [9,10]. Although, it has greater prevalence in the oral cavity, it is due to its tendency to present itself extra nodal sites. These lesions are usually symptomatic and usually present as a rapidly enlarging mass [5].

As the curability of this lymphoma depends on the initial chemotherapy protocol, selection of the proper protocol is of critical importance, therefore the Ann Arbor staging system is essential, along with a work-up to identify the patient's risk factors.

The selection of chemotherapeutic agents represents a myriad of possible combinations and is in the province of the medical oncologist. Multi-agent chemotherapy is the first line of management for these types of tumours. The combination of cyclophosphamide, hydroxy daunomycin, Oncovin [vincristine] and prednisone (CHOP protocol) is used [11].

The role of surgery is very limited in the treatment of these tumours, as early stage tumours responds well to chemotherapy or combination of chemotherapy and radiotherapy [12].

Five pre-treatment prognostic factors identified to relate well with outcome are graded based on age, tumour stage, number of extra nodal sites, patient response to chemotherapy and serum lactic dehydrogenase levels, based on that, risk factors are determined ranging from low to high, in which 5 year cure rates for low tumour are 87% while for high grade it is 26% [13].

In our case initial histopathology report suggested to be malignant round cell tumour and accurate diagnosis of B cell lymphomas was arrived with use of immunohistochemistry, with use of CD20 marker, since lymphomas responds well to chemotherapy presently patient is undergoing chemotherapy under the supervision of medical oncologists.

Conclusion

As these tumours are morphologically similar, a combination of histopathology, immunohistochemistry and cytogenetic aids in precise diagnosis, careful histological diagnosis of round Cell tumours is of paramount important.

Conflict of Interest

The authors declare that they have no conflicts of interest.

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