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CASE REPORT

Nasopharyngeal Malt Lymphoma- A Case Report Of Rare Entity With Review Of Literature

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ABSTRACT

Mucosa associated lymphoid tissue confers the first line of defense in case of membranes which are directly exposed to the external environment. MALT lymphomas arising at these sites present diagnostic and therapeutic challenges, especially if arising from areas such as the nasopharynx, where adenoid hyperplasia is very commonly present as a mass. In the present case report, a 55 year old male presented to the medical emergency department with complaints of severe respiratory distress. Radiological and clinical evaluation revealed a mass; the biopsy was taken and sent for histopathological examination, which suggested a possibility of MALT lymphoma. This was subsequently proven on immunohistochemical studies, which showed positive staining for CD 20 and negative staining for CD 3, CD 5 and CD 43. Hence, a final diagnosis of low grade B-cell MALT lymphoma was made. A quick and comprehensive diagnosis of MALToma is desirable, as it is amenable to chemotherapeutic therapy.

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INTRODUCTION

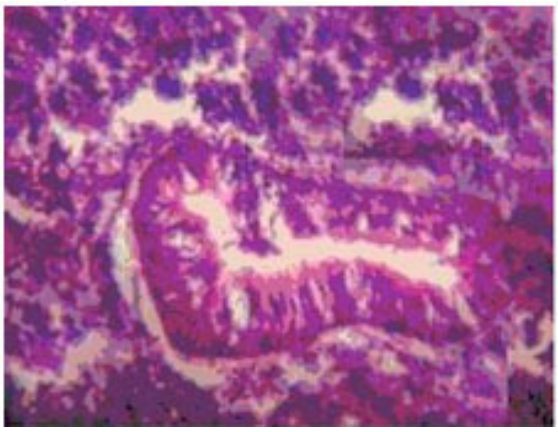
Mucosa associated lymphoid tissue (MALT) is that part of the immune system which provides protection to the freely permeable surface of the gastrointestinal tract and other mucosal membranes which are directly exposed to the external surface, such as Waldeyer's ring and the nasopharyngeal area. [1] The lymphomas arising from these sites are known as MALTomas, which exhibit characteristic histopathological and immunohistochemical features. We present a case of nasopharyngeal MALToma which caused clinical as well as histological dilemma, but with a planned algorithmic approach, was finally diagnosed as low grade B cell MALToma.

CASE REPORT

A 55 year old obese male presented to the emergency department with respiratory distress, peripheral cyanosis and altered sensorium. The patient was immediately intubated and was put on ventilator support. The arterial blood gas (ABG) analysis revealed features of respiratory acidosis with hypoxaemia. This was the first episode of distress which was incurred by the patient, although he had a history of gradual increase in activity and frequency of snoring, with difficulty in breathing while lying down,

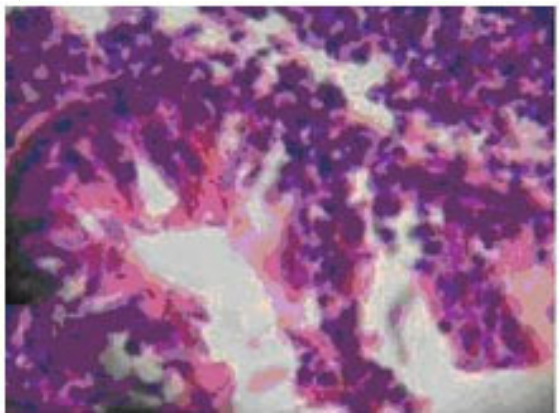
for the past one year. No other history of any past illnesses including drug intake was there. He was advised by a physician to lose weight and to visit an otorhinolaryngealologist, which he did not heed to.

[Table/Fig 1]: Lympho-epithelial lesion (H & E 200 X)

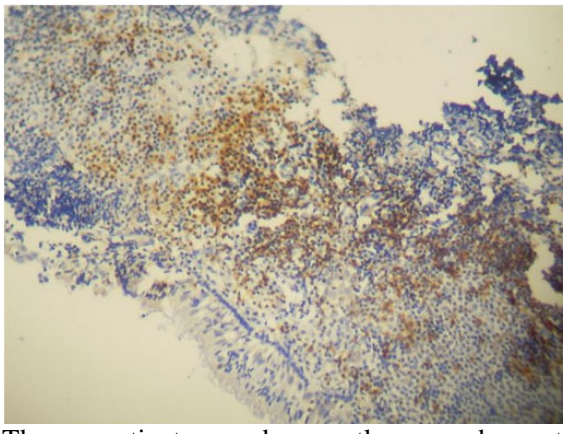


The patient was investigated thoroughly and apart from a deranged ABG report, the rest of his haematological and biochemical parameters were within normal limits, including an unremarkable electrocardiogram (ECG) and chest skiagram. Indirect laryngoscopy revelaed abulging mass in the nasopharyngeal area. CT scan was advised, which revealed a soft tissue mass (4 x 2.5 cm) involving the left aspect of the pharyngeal space, bulging into the nasopharyngeal airway. Nasal endoscopy was done and biopsy was taken from the mass, which appeared to be irregular in shape, friable and highly vascular. Multiple soft tissue pieces all together measuring 1.5 x 0.5 cm in size, were sent for histopathological examination and nasal packing was done, followed by tracheostomy. Histopathology revealed a lining of pseudo stratified columnar epithelium, beneath which dense diffuse lymphoid infiltrates were present. These lymphoid cells were seen to be infiltrating around and invaginating the underlying glandular epithelium. A provisional diagnosis of low grade mucosa associated (MALT) lymphoma was suggested. I Immunohistochemical (IHC) studies were done, which showed positive staining for CD 20 and negative staining for CD 3, CD 5 and CD 43. Hence, a final diagnosis of low grade B-cell MALT lymphoma was given.

[Table/Fig 2]: Lymho-epithelial lesion (H & E 400 X)

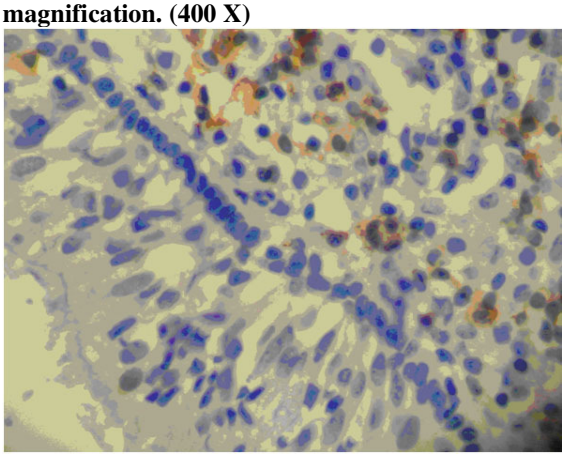


[Table/Fig 3]: CD 20 Positivity seen in the lymphoid infiltrate just beneath the respiratory epithelium. (200 X)



The patient subsequently underwent chemotherapy. On follow up after one year, nasal endoscopy and MRI showed nearly complete resolution of the nasal tumour. Also, the clinical symptoms of respiratory distress, snoring and discomfort on lying, had completely disappeared.

[Table/Fig 4]: CD 20 Positivity on higher magnification. (400 X)



DISCUSSION

Mucosa associated lymphoid tissue is present in the mucosal lining at various sites in the body, predominantly in the gastrointestinal tract and in the mucosa of the head and neck region. MALT lymphomas arising at these sites present diagnostic and therapeutic challenges. Often, by light microscopy, it is difficult to diagnose them if a clear cut evidence of lympho-epithelial lesions is not seen. The situation is often piquant in the region of the nasopharynx, where it is difficult to differentiate it from reactive lymphoid hyperplasia. In all such settings, IHC analyses are helpful in confirming a diagnosis, because of the characteristic negative staining for CD 3, CD 5 and CD 43 and positive staining for CD 20 and monotypic staining for either the kappa or the lambda light chains. [1]

Many researchers have given a detailed commentary on the role of the nasopharyngeal lymphoid tissues. It has been noted that though nasopharyngeal MALT tissue morphologically resembles the gut, MALT tissue is significantly different. lymphoid migration patterns and the binding properties of their high endothelial venules. [2]

Lymphomas are the second most common neoplasms of the head and neck region after squamous cell carcinoma. Among the lymphomas; a maximum number of cases are those of non-hodgkin's lymphoma (NHL). [3] In the head and neck region, the maximal cases of primary lymphomas are seen in the Waldeyer's ring, which comprises of palatal, lingual, tubal and the nasopharyngeal lymphoid tissues. The tumours in the Waldeyer's ring itself accounts for 5 % to 10 % of all the lymphoma cases. [4], [5] Of all the NHLs arising from the Waldeyer's ring, the tonsil is their most frequent site, followed by the nasopharynx and the base of tongue and the soft palate. [5], [6]

The treatment options for MALT lymphomas are varied and there are many regimes which are quoted in literature from Rituximab to low dose cyclophosphamide and multi chemo regimes. Localized MALT lymphomas can be treated effectively with radiotherapy or surgery. The stress however, is on the early diagnosis of extragastric MALT lymphomas (seen in the present scenario), so as to prevent their conversion into disseminated disease, which is seen in 45-50% of the cases. [7] This can be done by utilizing IHC in all the cases of nasopharyngeal "masses", where all such cases of MALT lymphomas can be sifted out. [1]

This case is worth reporting due to its rarity and also to underline the importance of IHC in confirming the diagnosis between the MALT-lymphomas and the reactive lymphoid hyperplasias of the nasopharynx. This is important, as a quick start to chemotherapy can lead to the complete resolution and the non dissemination of the disease in all such settings. [7]

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