

Primary lymphoepithelial carcinoma of the nasopharynx

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Abstract— We presented a case of a 32-year-old Chinese lady who presented with intermittent epistaxis and nasal blockage for 2 years duration. Magnetic resonance imaging revealed a large well defined symmetrical soft tissue mass lesion arising from the mucosa of the nasopharynx. Endoscopic excision of the tumor was performed and the final histological examination was compatible with sinonasal lymphoepithelial carcinoma. The patient was then referred to the Oncology team for further management in view of postoperative radiotherapy.

Keywords— epistaxis, lymphoepithelial carcinoma, nasopharyngeal carcinoma.

1. Case presentation

A 32-year-old Chinese lady presented with 2 years history of intermittent epistaxis and nasal blockage.

Magnetic resonance imaging was performed which revealed a large soft tissue mass lesion arising from the mucosa of nasopharynx extending to post nasal space bilaterally. The lesion was symmetrical measures about 3.5x2.6x2.5cm with no spread elsewhere.

Following the imaging study, patient was then referred to our centre for further management and subsequently underwent endoscopic excision of nasopharyngeal tumour and the biopsy of the tumour was sent for histological examination.

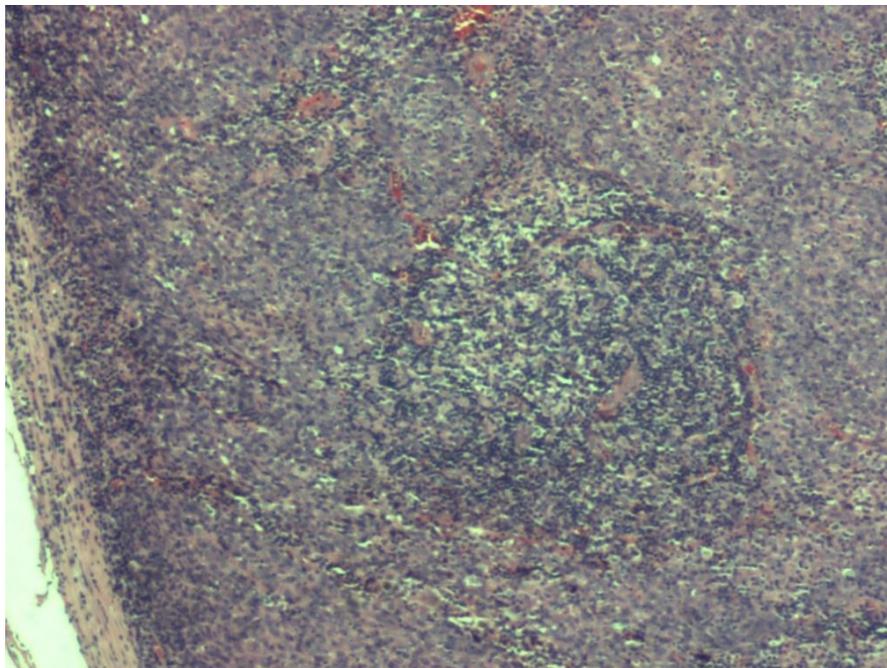


Figure 1: Tumour surrounding a lymphoid germinal centre (Haematoxylin & eosin, original magnification x40)

Histological examination of the biopsy demonstrated polymorphous lymphoid stroma (mixture of lymphocytes, plasma cells, and neutrophils) infiltrated by irregular islands of malignant cells with mild to moderate nuclear pleomorphism. Focal area however shows severe nuclear pleomorphism. The tumour cells have vesicular nucleus, large prominent eosinophilic nucleolus and indistinct cell borders, i.e. syncytial appearance. Tumour necrosis and focal individual cell keratinization are seen, but there are only occasional mitosis and hardly any stromal desmoplasia (Figures 1 & 2).

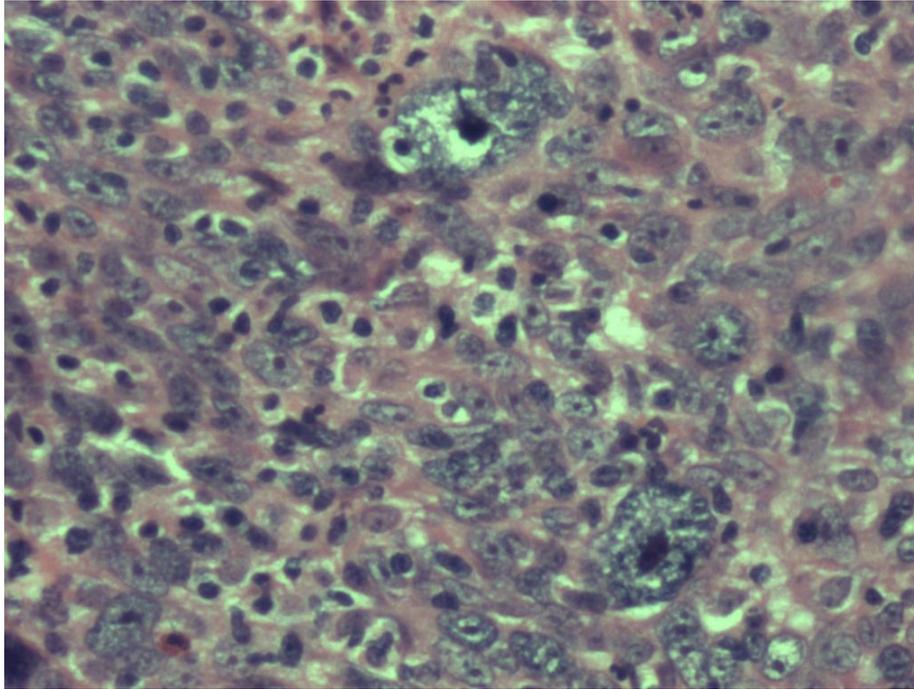


Figure 2: Occasional highly pleomorphic tumour cells amidst a background comprising syncytial sheet of tumour cells. (Haematoxylin & eosin, original magnification x200)

Immunostaining demonstrated that tumour cells are positive for MNF116, with occasional tumour cell nucleus expressing positivity for Epstein-Barr encoded RNA (EBER).

2. DISCUSSION

Lymphoepithelial carcinoma (LEC) subtype has been identified as an extremely rare form of tumour. It affects adults in the fifth to seventh decades, and there is a male predominance of approximately 3:1. Sinonasal lymphoepithelial carcinoma are more common in the nasal cavity compared to the paranasal sinuses, although the disease can occur at both sites simultaneously. The more aggressive tumour may invade locally into palate, orbit and base of skull.

The most common symptoms related to sinonasal lymphoepithelial carcinoma are epistaxis, nasal blockage or blood-stained sputum. The least common symptoms would be proptosis and cranial nerve palsy once the tumour has invaded intracranially. However, these symptoms are quite broad and can be related to any types of nasal pathology either benign or malignant. A high index of suspicion is needed in diagnosing with this rare subtype of malignancy.

Histology and further immunostaining play an important role in confirming this disease. From the previous study, lymphoepithelial carcinoma (LEC) has been strongly associated with Epstein-Barr virus (EBV) just like nasopharyngeal carcinoma (NPC) [5]. However, lymphoepithelial carcinoma (LEC) is separated from nasopharyngeal carcinoma (NPC) by its location [6] and its clinical outcome [7]. Interestingly, patients with lymphoepithelial carcinoma (LEC) in Western Europe and the USA are usually Epstein-Barr virus negative [6].

It is also crucial to distinguish between sinonasal lymphoepithelial carcinoma (LEC) and the more aggressive sinonasal undifferentiated carcinoma (SNUC) because the latter carry poor prognosis despite radical treatment. Histologically, sinonasal undifferentiated carcinoma (SNUC) is characterised by tumour cells with nuclear pleomorphism, high mitotic rate and the presence of necrosis. EBV status also can be used to distinguish between sinonasal lymphoepithelial carcinoma (LEC) and sinonasal undifferentiated carcinoma (SNUC) since the latter is mostly EBV negative except in very rare case.

In this study, the further immunostaining test are positive for Epstein-Barr encoded RNA (EBER) which confirm the diagnosis of lymphoepithelial carcinoma (LEC) in the sinonasal region.

5. Conclusion

Lymphoepithelial carcinoma of the sinonasal is a very rare malignant tumor with a very little report in the literature. A high index of suspicion is needed in diagnosing this disease. Furthermore, histological and immunostaining test are also needed to differentiate further in between this rare malignant tumour with the other malignancies.

6. References

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