Unilateral Tonsillar Enlargement: A Challenge To The Traditional Thinking
B Wei, S O'Leary, K Opeskin

Citation

Abstract
A case of unilateral tonsillar enlargement as a benign hyperplastic reaction to a Hodgkin's lymphoma at level II in the ipsilateral cervical lymph nodes is reported. In the traditional teaching of head and neck surgery, unilateral tonsillar hypertrophy is presumed to be neoplastic until proven otherwise. This case demonstrated that the absence of neoplasia in an enlarged tonsil does not exclude the possibility of a local-regional tumor, and therefore further investigation may be required to exclude this possibility.

INTRODUCTION
Neoplasms, most commonly squamous cell carcinoma or Non Hodgkin's lymphoma, may present as an enlargement of the palatine tonsil. Rarer tumors of the tonsils presenting in this way include extramedullary plasmacytomas, Hodgkin's disease, leukemia, and metastatic neoplasms. Unilateral tonsillar enlargement with regional lymph node hypertrophy further strengthens the suspicion of a primary malignancy in the tonsil when other causes of the unilateral tonsillar enlargement, such as acute infection, have been excluded.

It is a standard practice to perform a diagnostic tonsillectomy to exclude malignancy when tonsillar asymmetry is encountered. Our case demonstrates that the absence of an occult malignancy in an asymmetrically enlarged tonsil does not rule out a local-regional primary lymphoma. The tonsillar enlargement may be a reactive response to a nearby primary tumor.

CASE REPORT
A 15-year-old male presented for an opinion concerning right tonsillar enlargement, which had persisted for 6 months. The patient was well with no previous history of tonsillitis, dysphagia, odynophagia or haemoptysis. The examination findings revealed a marked tonsillar asymmetry, with the enlarged right tonsil, measuring 4x3x3cm. The tonsillar mucosal surface was normal. The left tonsil was of normal size and appearance. The examination of the neck revealed an enlarged right tonsillar node of 3x3x3cm, which was mobile and smooth. The rest of the ENT examination was unremarkable. The patient's routine blood tests were normal.

A right tonsillectomy was performed, and the histopathological examination revealed marked reactive changes only (reactive follicular lymphoid hyperplasia). Repeated histopathological analysis including flow cytometry, southern blot analysis, polymerase chain reaction (PCR)- DNA analysis found no evidence of B-cell lymphoma. Culture of the tonsil grew actinomyces, but the histopathological appearance of the tonsil indicated that this was a commensal organism.

One month later the cervical lymphadenopathy was unchanged. A CT scan of the head and neck confirmed the ipsilateral cervical lymphadenopathy. Fine needle aspiration (FNA) of the cervical lymph node did not reveal any pathology. An excisional biopsy of the node was therefore performed for diagnostic purposes. The histology of the node revealed a tumor mass with the histopathological features of a lymphocyte predominant Hodgkin's lymphoma. Subsequent investigation confirmed that the disease was localized to the neck.

DISCUSSION
Unilateral tonsillar enlargement is considered to be a sign of a potential tonsillar malignancy. In a series from the University of Iowa, Beaty et al. reported that the strongest risk factor for predicting tonsillar malignancy was tonsillar asymmetry, which occurred in 21(84%) of 25 patients with a tonsillar neoplasm. The experience of the House Ear
Institute is that the incidence of occult malignancy in patients with incidental unilateral tonsillar enlargement and an otherwise normal tonsillar mucosa and neck examination, is about 5%.[4] Therefore the traditional teaching of otolaryngology is to consider unilateral tonsillar hypertrophy neoplastic until proven otherwise and diagnostic tonsillectomy is recommended [American Academy of Otolaryngology-Head and Neck Surgical Indicator Compendium].

Primary malignant tumors of the tonsil have been said to represent 12% of oral cavity neoplasms. Squamous cell carcinoma accounts for 85% to 90% of tonsillar malignancies, and the remaining 10% to 15% are lymphomas and other rarer tumors. Primary lymphoma of the tonsil is the most frequent form of primary extra nodal lymphoma in the head and neck.[7, 8, 9, 10] In contrast to squamous cell carcinoma, lymphoma usually presents first as an unilateral globular tonsillar enlargement without ulceration of the overlying mucosa. Secondary malignancy of the tonsil is very rare.[11] Therefore, most surgeons consider the risk of a local-regional neoplasm to be low if excision biopsy of a tonsil proves negative for a malignancy.

However, our observations challenge the traditional teaching relating to unilateral tonsillar enlargement. It demonstrates that despite the absence of malignancy in the unilaterally enlarged tonsil, there is still a possibility of a local-regional lymphoma and further investigations may need to be carried out to exclude this possibility. In this case presentation, the presence of the cervical lymph node hypertrophy and the non-diagnostic fine needle aspirations prompted the excision biopsy of the pathological lymph node. Further more, needle aspirations or needle biopsies are not adequate for the primary histologic diagnosis of Hodgkin's disease or any other lymphoma.[13]

An alternative approach to diagnosis may have been to use positron emission tomography (PET) imaging. Metabolic imaging with PET using fluorodeoxy-D-glucose (FDG) provides information on the functional characterization of tissues without dependence on morphologic criteria. It allows the detection of foci of increased glycolysis, which is an important sign of tumour metabolism.[14] FDG-PET is a well established tool for staging for various malignancies. In particular, it has the ability to image lymphomatous involvement in Hodgkin's disease and Non-Hodgkin's lymphomas. It also has an impact on the detection of the primary as well as the disseminated disease in patients with cancer of unknown primary.[15, 16] In this case, PET scanning may have shown more significant uptake in the cervical lymph nodes than the unilaterally enlarged tonsil, aiding the choice of the site for initial tissue sampling.

Benign reactive hyperplasia of the local-region lymph nodes in response to local malignancy is well documented.[17, 18, 19] It is not an uncommon event in head and neck cancers and has been described in response to squamous cell carcinoma. However, reactive enlargement of lymph nodes in response to a regional lymphoma is rare. For this to occur in the tonsils is an even rarer event. Our case demonstrates that the tonsil can become hyperplastic, like regional lymph nodes, in response to a nearby lymphoma.

The exact mechanism of the benign reactive hyperplasia in response to regional tumor is still unknown. The most widely accepted theory is that of an immunological response to the tumor antigens.[18, 20] This is supported by the fact that lymphocytes and macrophages of the hyperplastic node are specifically cytotoxic to autologous tumor cells.[20, 21] The specific tumor antigens for lymphoma have not yet been identified.[22, 23, 24, 25, 26]

A possible explanation for the rarity of the benign tonsillar reactive hyperplasia in response to a local lymphoma is that, unlike lymph nodes, the lymphoid tissue of Waldeyer's ring has no afferent lymphatic channels.[27] In our case, we consider the most likely mechanism of the reactive tonsillar changes to be the obstruction of lymphatics in the tonsillar lymph nodes by the Hodgkin's disease with retrograde flow of lymph to the tonsil. In this way, the tonsillar lymphoid tissue could be exposed to antigens from the Hodgkin's lymphoma that had risen in the neck.

CONCLUSION

Unilateral tonsillar enlargement may be a benign hyperplastic reaction to a localized Hodgkin's lymphoma in the ipsilateral cervical lymph nodes. Therefore, the absence of tumors in an enlarged tonsil does not exclude the possibility of a local-regional primary neoplasm. Further investigation to exclude a local-regional lymphoma may be required, particularly in the presence of enlarged ipsilateral lymph nodes. The initial tonsillectomy may not provide the diagnosis and FNA of enlarged cervical lymph nodes may not be definitive, necessitating a diagnostic excision biopsy of the node.

CORRESPONDENCE TO

Dr Benjamin P.C. Wei Department of Otolaryngology
University of Melbourne Royal Victorian Eye & Ear Hospital 32 Gisborne Street East Melbourne 3002 E-mail: bwei@bionicear.org Fax: (03) 9663 1958

References
Author Information

Benjamin P. C. Wei, MBBS
Department of Otolaryngology, The Royal Victorian Eye & Ear Hospital, The University of Melbourne

Stephen O'Leary, Ph.D, F.R.A.C.S.
Associate Professor, Department of Otolaryngology, The Royal Victorian Eye & Ear Hospital, The University of Melbourne

Kenneth Opeskin, M.D. F.R.C.P.A.
Department of Anatomical Pathology, St Vincent's Hospital