



# Atypical Marginal Zone Hyperplasia (MZH) Is a Mimic for Marginal Zone Lymphoma (MZL) in Pediatric patients



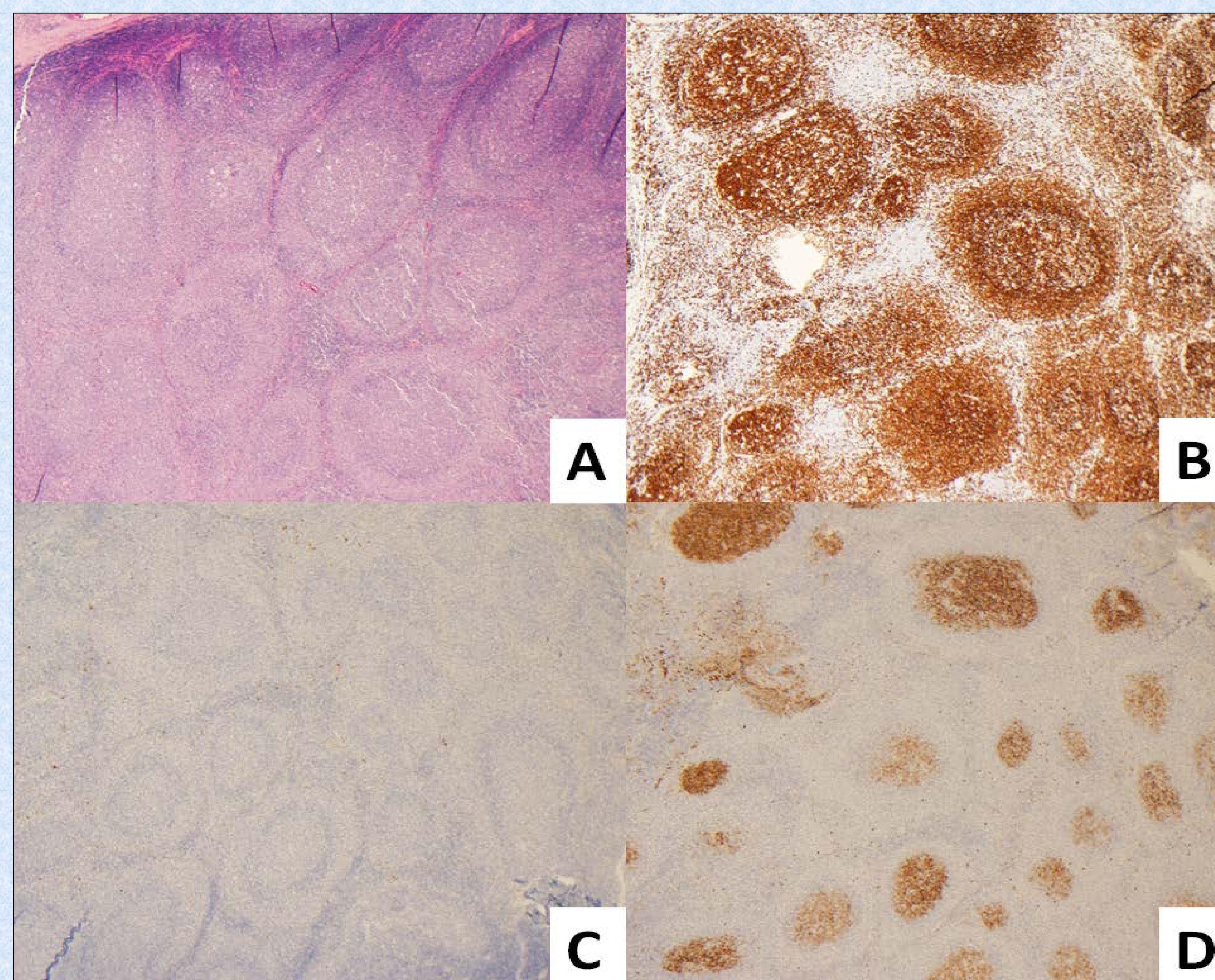
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## Case History:

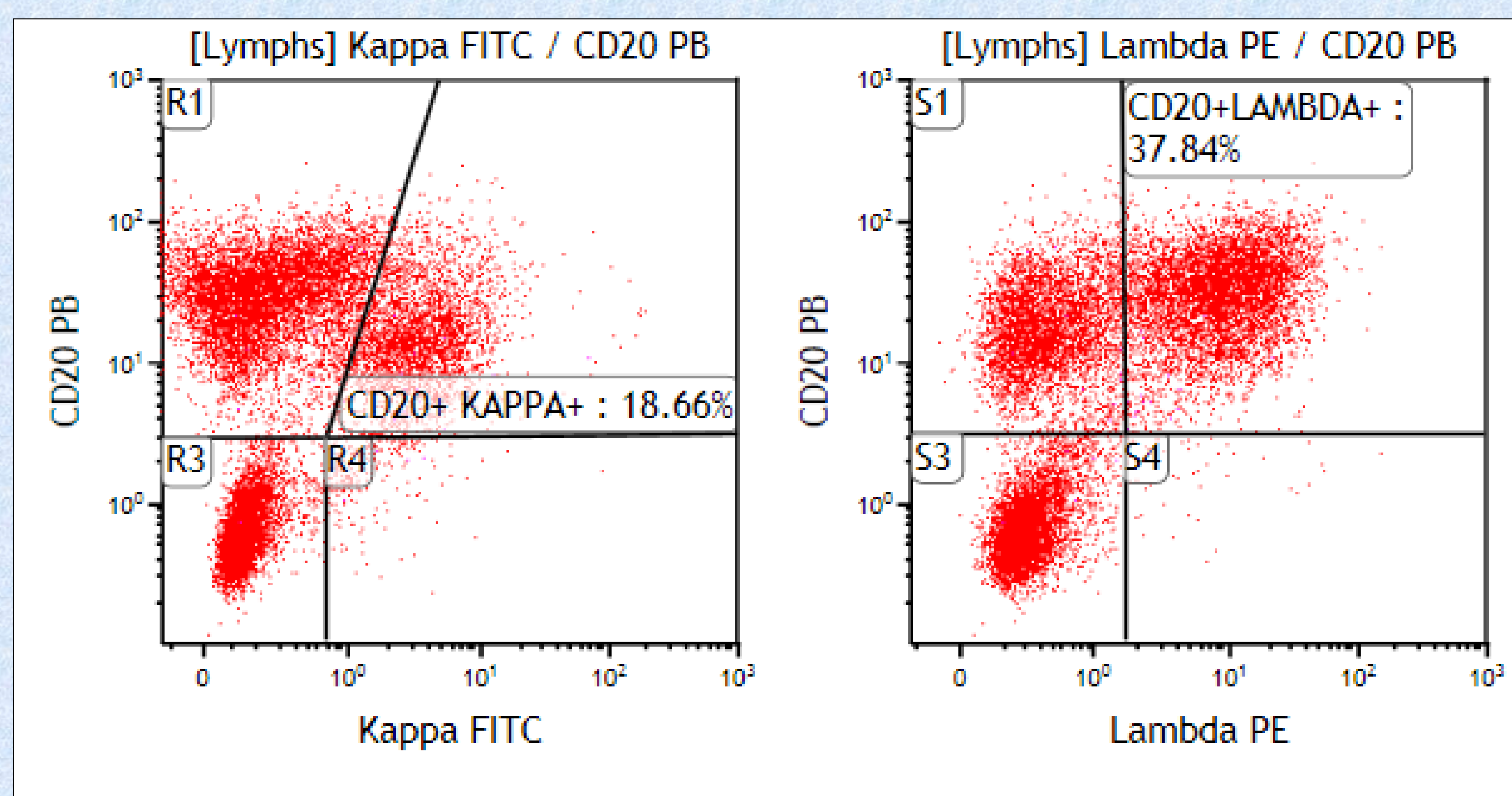
This is a rare case of a four year old female with no significant past medical history with the chief complaint of snoring and day time napping. Physical examination revealed bilateral tonsillar hypertrophy and partial obstruction of the posterior pharynx. Histological examination of the tonsils revealed an atypical nodular proliferation with expanded marginal zones and an intact overlying squamous epithelium (Figure 1A) .

## Case Work-up:

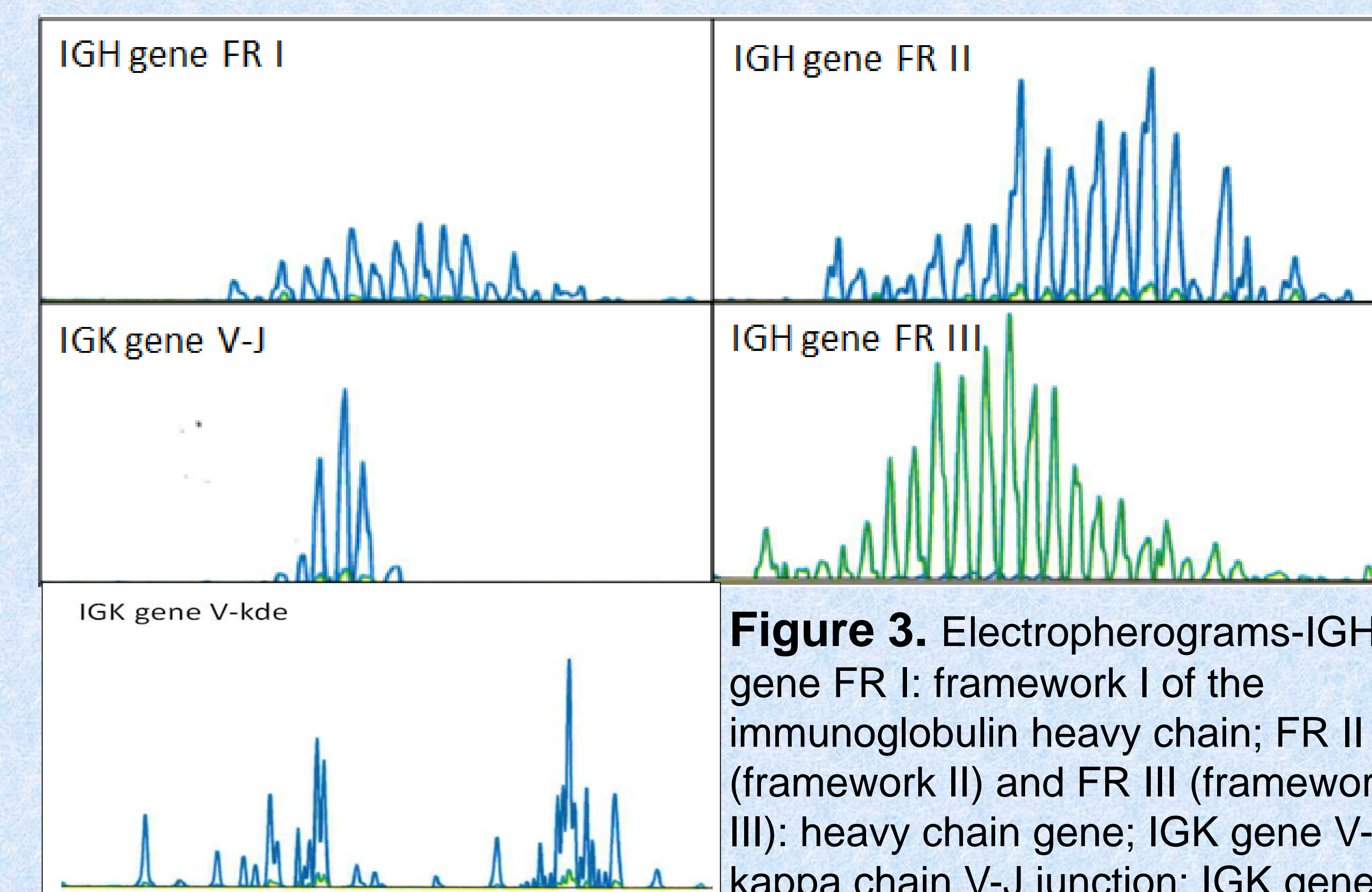
The lymphocytes expanding the marginal zone were shown to be B-cells (CD20+, CD5, CD10-, CD79a+, PAX-5+, BCL-1-, BCL-2+) predominantly small to intermediate sized with scant eosinophilic cytoplasm. These lymphocytes showed slight lambda light chain excess by flow cytometry (kappa to lambda ratio of 1:2) (Figure 2) and immunohistochemistry. No expansion or colonization of the germinal centers were noted by CD23 staining and Epstein-Barr studies were negative. The above findings raised suspicion for a neoplastic process. However, the follow up B-cell gene rearrangement studies showed no evidence of clonality (Figure 3) which was consistent with a non-neoplastic process (in light of no distinct monotypia by flow/immunohistochemistry and the patient's age and histologic findings).



**Figure 1.** H&E section of the tonsil showing follicular hyperplasia with expansion of the mantle zones (Figure 1A). The lymphocytes within the mantle zone are B-cells that are PAX-5 positive (Figure 1B), BCL-1 negative (Figure 1C) and CD10 negative (Figure 1D).



**Figure 2.** Flow cytometry showing CD20 positive B-cells with lambda excess.



**Figure 3.** Electropherograms-IGH gene FR I: framework I of the immunoglobulin heavy chain; FR II (framework II) and FR III (framework III); heavy chain gene; IGK gene V-J: kappa chain V-J junction; IGK gene V-Kde:V-Kde junction.

## Discussion:

Atypical MZH is a relatively newly described entity that appears related to local antigenic stimulation with B-cell expansion which can mimic MZL, but are non-clonal proliferations and based on current evidence have no risk of progression to lymphoma. Interestingly, they may be lambda light chain restricted or have lambda light chain excess and may also show a moderately high proliferative index which raises suspicion for a clonal process. Despite the suspicious presentation, marginal zone hyperplasia is a benign process and must be confirmed by molecular testing. Large scale studies are required to further understand the pathobiology and risk factors that may predispose one to MZH and MZL. Overall, this rare case represents an instance of marginal zone hyperplasia with lambda excess which is a diagnostic pitfall. The pathologist and pediatrician must be aware of these entities to prevent misguided therapy.