Commentary: As is our pathology, so is our practice

The above comment was made by the legendary clinician William Osler. In recent years, clinical acumen and investigative modalities have become quite a bit advanced, particularly imaging.[1] Pathology has also advanced a lot as it is no longer morphologic or pattern diagnosis. Immunohistochemistry with several molecular markers have revolutionized today's pathology.

In the 1980s, we pathologists used to diagnose lymphoid inflammatory lesions in the orbit into two types, monomorphic or pleomorphic. Monomorphic lymphoid infiltrate would be labeled as lymphoma whereas pleomorphic lymphoid infiltrate was called the benign or idiopathic inflammatory disease of the orbit. Now, any lymphoproliferative disease needs to be subjected to immunohistochemistry to identify the clonality. Lymphoma again needs to be subdivided into B-cell or T-cell lymphoma and it is also important to distinguish between idiopathic orbital inflammatory disease and malignant lymphoma. Molecular diagnosis by gene expression profiling helps to differentiate the various forms of orbital inflammatory disease.[2]

In addition, any inflammatory lesions of the orbit should be investigated to rule out infective etiology like tubercular, fungal, or parasitic inflammations. As histopathology may not often be characteristic with caseation necrosis with granulomatous inflammation, special staining for acid-fast bacilli should be done. A polymerase chain reaction for mycobacterial tuberculosis DNA from the paraffin section of the biopsy specimen can also be done if there is suspicion of tuberculous infection. Similarly, GMS staining for fungus should also be carried out when suspected fungal infection of the orbit.

Idiopathic orbital inflammations are often a challenge to diagnose. It accounts for approximately 5% of all orbital mass lesions. Histopathology reveals chronic inflammatory cellular infiltrate composed of small mature lymphocytes (predominantly T-cells), plasma cells, neutrophils, eosinophils, and occasionally histiocytes and macrophages. The infiltrates are sometimes focally organized in lymphoid follicles with reactive germinal centers. Stromal changes may include edema and proliferative fibrosis and sclerosis. When connective tissue fibrosis predominates over inflammatory cells, diagnosis changes to idiopathic sclerosing orbital inflammation. Some feel it as the end stage of the histological continuum of the orbital pseudotumor while some feel it be a unique form of idiopathic orbital inflammation.[3]

It is quite interesting to note that authors in their series found IgG4-related disease in one case with Mikulicz's disease.[4] There are several reports of IgG4-related eye disease in the orbit.[5] IgG4-related diseases are systemic syndromes characterized by elevated serum levels of IgG4 and IgG4-positive lymphoplasmacytic infiltrative lesions in the body. Histopathology of orbital IgG4-related disease includes different degrees of lymphoplasmacytic infiltrates with dominant sclerosing lesions or reactive lymphoid follicles. Eosinophilic infiltrates are also observed. Immunohistochemistry shows IgG4-positive cells and plasma cells and can distinguish from other inflammatory conditions of the orbit. The diagnostic criterion for IgG4-related diseases are elevated serum IgG4-related concentration >135 mg/dl and >10 IgG4+ plasma cells per high power field of the biopsy sample and the ratio of IgG4+/IgG+ plasma cells being >40% of IgG+ plasma cells.[6]

The distinction between various forms of idiopathic orbital inflammation and its systemic associations, *per se* is challenging for clinicians. However, histopathological distinction helps comment upon disease types, prognosis and, hence, treatment modalities.

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