Perineuriomas are benign peripheral nerve sheath neoplasms composed of perineurial cells with characteristic immunohistochemical and ultrastructural features. They have been traditionally classified into two main types according to their location–intraneural and extraneural–and overlap histologically with many other tumors, which may be diagnostically challenging to general surgical pathologists.

The correct identification of perineuriomas is important to avoid unnecessary overtreatment. The histologic diagnosis should be confirmed through immunohistochemical studies (including epithelial membrane antigen, S100 protein, and more recently described antibodies such as claudin-1 and GLUT1) or electron microscopy. Cytogenetic and molecular genetic studies are still of limited value for the diagnosis of perineuriomas but may play a fundamental role in excluding important differential diagnoses and also in helping elucidate the biology of these poorly known neoplasms.