Commentary

Plexiform Schwannomas an Update

Schwannomas exhibit histologic features that may overlap with those of benign and malignant peripheral nerve sheath tumors (MPNSTs). This can be the challenging in making accurate histopathological diagnosis there by influence clinical management.[1]

Schwannomas show an interesting histologic variety despite being composed of a limited array of cellular constituents. Key issue for the pathologist is a definitive diagnosis in this histologic diversity for better management. Peripheral nerve sheath neoplasms exhibit histologic features that overlap with those of many other benign and malignant soft-tissue tumors.[2] The correct diagnosis relies mainly on histologic findings and immunohistochemistry profile, but ultrastructural studies can be used in difficult cases. Key issue for the pathologist includes distinguishing schwannomas from neurofibromas and MPNSTs from cellular schwannoma or neurofibromas Plexiform schwannoma is a rare variant of schwannoma that accounts for only 5% of all schwannomas and typically shows a plexiform or multinodular growth pattern. It was first described by Harkin and Reed in 1978, and very few cases had since been reported.[3]

It is believed that these lesions arise from proliferation of Schwann cells at a point inside the perineurium, which causes a displacement and compression of the surrounding normal nerve tissue and trauma and neurofibromatosis type 2 are well-recognized risk factors for these lesions.[4]

Differential diagnosis of plexiform schwannoma poses a challenge and includes mucocele, granular cell tumor, lipoma, hemangioma, eosinophilic granuloma, epidermoid and dermoid cysts, epithelial hyperplasia, and benign salivary gland tumors. In some cases, glandular malignant processes, squamous cell carcinoma, sarcomas, and MPNSTs may share similar clinical features plexiform schwannoma from plexiform neurofibromas and MPNSTs because plexiform schwannoma follows a benign clinical course, with complete surgical excision being curative. Plexiform schwannoma can have a multinodular or plexiform appearance, although the term has been used interchangeably by many researchers.[5]

Solitary plexiform schwannomas occur in patients with no significant predisposing factor, however, multiple tumors are more frequent in individuals with neurofibromatosis Type II, schwannomatosis, Gorlin–Koutlas syndrome, and patients with positive family history or history of trauma.[6]

No sex predilection has been reported for plexiform schwannoma. Malignant transformation of this benign neoplasm is very rare. There are some case reports of benign solitary schwannomas with a consequent malignant transformation.[7]

Although plexiform schwannoma has a superficial resemblance to the plexiform neurofibroma characterized by multinodular growth, it is composed purely of Schwann cells with nuclear palisading and verocay bodies without myxoid changes of stroma. Whereas S100 protein staining identifies a purely and strongly positive Schwann cell population in plexiform schwannoma, variable expression of this antigen is observed in plexiform neurofibroma.[8] To exclude the malignant counterpart, the combination of pathologic findings (cellularity, mitotic activity, and at typia), as well as immunohistochemical study for S100 protein (most MPNSTs show negative or weak staining) should be considered.[9]

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