Peripheral primitive neuroectodermal tumors (pPNETs) are also classified as part of the Ewing family of tumors (EFTs); peripheral primitive neuroectodermal tumors (pPNETs) and Ewing family of tumors (EFTs) are often referred to interchangeably in the literature. Generally, Ewing family of tumors (EFTs) and peripheral primitive neuroectodermal tumors (pPNETs) represent different manifestations of the same tumor and have similar genetic alterations. Ewing sarcoma, however, is more common in bone, while peripheral primitive neuroectodermal tumors (pPNETs) are more common in soft tissues. Immunohistochemical and cytogenetic studies suggest that these tumors all have a common origin.

The following tumors are classified as peripheral primitive neuroectodermal tumors (pPNETs):

Ewing sarcoma (osseus and extraosseous)

Malignant peripheral primitive neuroectodermal tumors (pPNETs) or peripheral neuroepithelioma of bone and soft tissues

Askin tumor (peripheral neuroepithelioma of the thoracopulmonary region)

Other less common tumors (eg, neuroectodermal tumor, ectomesenchymoma, peripheral medulloepithelioma)

Primary spinal peripheral primitive neuroectodermal tumor

see Primary spinal peripheral primitive neuroectodermal tumor

Ewing's Sarcoma peripheral primitive neuroectodermal tumor

see Ewing's Sarcoma peripheral primitive neuroectodermal tumor

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Last update: 2024/06/07 02:58

