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Primitive neuroectodermal tumours (PNETs) located in the spinal canal; the relevance of classification as central or peripheral PNET

Case report of a primary spinal PNET occurrence with a critical literature review

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Summary Intraspinal location of central PNET (cPNET) is very rare. We present a case, critically review all publications of primary intraspinal cPNET occurrence and discuss tendencies in clinical presentation. In several previous attempts to summarise, authors often confused cPNET with peripheral PNET (pPNET). cPNET and pPNET are different entities with different immunohistochemical profiles and genetic backgrounds. Clinically, they are both aggressive tumours, but exhibit different characteristics in their local manifestation and metastatic spread. Survival rates are quite similar provided that treatment is applied according to the established protocols. Protocols in cPNET treatment differ from those for pPNET as regards the order of the treatment sub-modalities, specific chemotherapeutic regimen and intensity, radiation dose and its extent and consequently, the side effects. Therefore, failure to distinguish cPNET from pPNET leads to clinical guidance and treatment proposals based on false assumptions, which might effect outcomes. Often, distinguishing between cPNET and pPNET is easy, because they occur in different location. In the case of intraspinal

tumour location, however, the differentiation is crucial because both primary cPNET and pPNET can occur intraspinally, even though this is rare. Nowadays, demonstrating the expression of MIC2 glycoprotein by immunocytochemical staining (CD99) showing the specific EWS-FLI1 chimeric gene presence in pPNET, offers an easy way of making a differential diagnosis between cPNET and pPNET.

Keywords central and peripheral primitive neuroectodermal tumour - EWS-FLI1 chimeric gene - immunocytochemical staining - intraspinal - review

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References secured to subscribers.