Primitive neuroectodermal tumor of the orbit: A case report.

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Abstract:

Primitive neuroectodermal tumor (PNET) is a small round cell malignant tumor of neuroectodermal origin.It is seen in young adults, rarely involves the orbit and affects males and females equally. Most of the PNETs occur in the central nervous system (CNS). PNETs recognized outside of CNS are diagnosed as peripheral PNET (pPNET). pPNET shows characteristic small round cell tumor with rosette or pseudo-rosette, positive immunohistochemistry (IHC) and in some cases ultra structural findings of neurosecretory granules. It expresses MIC-2 gene (CD99) and is said to be least aggressive after complete tumor resection. We report a case of a twelve-year old male child presenting with unilateral eccentric proptosis diagnosed as primary peripheral primitive neuroectodermal tumor (pPNET) on histopathology and immunohistochemistry. We describe its distinguishing features with emphasis on multimodal and aggressive treatment approach which ensures appropriate management of these cases.

Keywords: - CD 99, Ewings sarcoma, orbit, Primitive Neuroectodermal tumor.

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