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Update on paraneoplastic neurological syndromes

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Paraneoplastic neurological syndromes (PNS) are remote effects of cancer that may involve any part of the nervous system. Rarity hinders their diagnosis and management and at least 60% of cases do not present a tumor at neurological symptoms onset. An important diagnostic element is detection, in patients' serum or cerebrospinal fluid of onconeuronal antibodies which recognize antigens expressed by the nervous system and by neoplastic cells during de-differentiation. Their detection also implies that PNS have autoimmune origin and that immunomodulation could be an effective treatment. The lack of clinical trials due to the rarity of PNS makes it hard to test the efficacy of immunomodulatory therapy but dividing the diseases into two groups' permits preliminary analysis. A humoral immunoresponse prevails in group one and antibodies seem to have a pathogenetic role indicating antibody removal strategies. Group two are mainly PNS of the central nervous system with autoantibodies directed against intracellular antigens, probably involving a cell-mediated mechanism. Immunotherapy with steroids or cytotoxic immunosuppressive agents may be useful here. Immunomodulatory treatment is always indicated when a tumor is not found but neurological symptoms are progressing, since first line treatment is tumor identification and where possible removal.

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Posterior fossa abscess secondary to dermal sinus associated with dermal cyst in children

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Introduction: Dermal sinus occurs in the cranial vault and the occipital region. Rarely, is localized in the posterior fossa, particularly in the midline position or in the cavity of the fourth ventricle. It could communicate with the skin through a fistula with potential risk of deeper abscesses. Posterior fossa abscess secondary to dermal sinus associated with intracranial dermal cyst is an uncommon pathology (0.1 to 0.7 %).

Methods: A 24-month old girl was admitted to our institution with headache but no focal neurological signs. Was present a cutaneous fistula in the midline of the occipital region. MR showed a sottotentorial intra diploic cyst with peripheral enhancement and edema. The mass was hyper intensity on T1-weighted sequences with lower signal on T2-weighted images. On STIR images the lesion showed low signal intensity. Sagittal images demonstrate the typically oblique stalk that linked cyst and skin. A sub-occipital craniotomy was performed with evacuation of the abscess and an excision of the visible capsule with a total removal of a 3 cm whitish, midline and encapsulated cystic mass with hair component.

Results: The histologic examination confirmed the diagnosis of abscess associated with dermal cyst and dermal sinus. Post-operative course was uneventful. The patient experienced a sudden improvement of cephalalgia and 15 days after the microsurgical excision was discharged. On postoperative RM imaging, it was found the total removal of the neoplasm; a 12 month follow-up no evidence of recurrence.

Discussion: Posterior fossa dermoid cyst should be considered in all children with a cutaneous fistula. Early neurosurgical treatment of these benign tumors should be performed to prevent the development of severe intracranial infection. Best results were obtained in cases of early diagnosis and the complete removal of the abscess. The reported case passes through a review of similar cases reported in the literature confirm the rarity of the case report.

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