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Sporadic malignant peripheral nerve sheath tumour (MPNST) in a 3-year-old girl: A diagnostic challenge

Zawawi, Muhamad Syahrul Fitri[a, b, c]; Ismail, Wan Faisham Nu'Man Wan[b, c]; Mustapar, Nurulhasanah[a, d]; Mohamad, Norsarwany[c, e]; Nasir, Ariffin[c, e]; Sharif, Sharifah Emilia Tuan[a, c]

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[a] Department of Pathology, School of Medical Sciences, Universiti Sains Malaysia, Health Campus, Malaysia
[b] Department of Orthopaedics, School of Medical Sciences, Universiti Sains Malaysia, Health Campus, Malaysia
[c] Hospital Universiti Sains Malaysia, Kelantan, Kubang Kerian, 16150, Malaysia
[d] Department of Pathology & Laboratory Medicine, International Islamic University Malaysia, Pahang, Malaysia

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Introduction: Malignant peripheral nerve sheath tumour (MPNST) is an uncommon malignant neoplasm of childhood with unfavourable prognosis. Only a limited number of cases have been reported in children less than 12 years of age, and approximately one-half arise from a benign peripheral nerve sheath tumour , especially in the background of neurofibromatosis type 1 (NF1). Primary MPNST in children is even rarer. Case report: A 3-year-old Malay girl presented with painful right axillary swelling for six months, initially treated as axillary lymphadenitis and she defaulted follow up. She came back four months later with enlargement of the swelling. The previous biopsy was reported as Schwannoma, which correlates with a benign peripheral nerve sheath tumour 's MRI findings. The final diagnosis after debulking surgery was consistent with MPNST . She succumbed to death 20 months after her initial diagnosis of advanced MPNST and lung metastasis. Pathological findings: Grossly, a huge partly circumscribed soft tissue mass was noted arising from a nerve with a solid greyish yellowish myxoid cut surface. Spindle-shaped cells arranged in a herringbone pattern alternated with areas of myxoid hypocellular areas exhibited marked pleomorphism, brisk mitosis, and extensive necrosis are seen microscopically. Immunohistochemistry shows patchy S100 protein staining with loss of expression of H3K27me3. Conclusion: Although MPNST is rare in the paediatric age group, the diagnosis should be considered in children without NF1 with a rapidly evolving and painful mass in the peripheral nerve distribution. In this case, the diagnosis was delayed and made after surgery. Due to its morphologic heterogeneity and lack of specific immunohistochemical markers, MPNST remains a diagnostic challenge . © 2022, Malaysian Society of Pathologists. All rights reserved.

Author keywords

children; diagnostic challenges; malignant peripheral nerve sheath tumour

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Sharif, S.E.T.; Department of Pathology, School of Medical Sciences, Universiti Sains Malaysia, Health Campus, Kelantan, Kota Bharu, Malaysia; email:dremilia@usm.my

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