Primitive Neuro Ectodermal Tumor (PNET) in infant diagnosed by histopathology and immunohistochemistry techniques

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Abstrak

Kasus ?Primitive Neuro Ectodermal Tumor? (PNET) sangat jarang dan sangat sukar didiagnosis. Sebuah kasus PNET didiagnosis dengan teknik histopatologi dan pemeriksaan imunohistokimia. Seorang bayi laki-laki umur 4 bulan diperiksakan ke rumah sakit dengan benjolan pada dinding dada sejak bayi tersebut berumur 3 hari. Benjolan tersebut makin lama makin membesar hingga akhirnya mencapai diameter ± 10 cm, selanjutnya penderita dibawa ke klinik. Benjolan tersebut terfiksir pada dinding dada dengan batas tidak tegas, pada kulit diatas tumor tampak dua ulkus. Selanjutnya tumor tersebut didiagnosis sebagai suatu hemangioma. Secara makroskopis tumor berukuran 17 x 13 x 5,5 cm, berbatas tidak tegas, berwarna putih dan lunak. Secara mikroskopis massa tumor terdiri atas sel-sel berukuran kecil yang tidak berdiferensiasi, berbentuk bulat-oval, dengan inti hiperkromatik, dan sebagian membentuk struktur roset, Homer-Wright di antara bagian lainnya yang difus. Mitosis 7/10 HPF, nekrosis minimal kurang dari 25%. Gambaran ini sesuai dengan suatu ?malignant small round sel tumor?, Pada pemeriksaan imunohistokimia dengan panel antibodi meliputi Vimentin, NSE, Chromogranin dan CD99 menunjukkan Vimentin positif lemah-sedang, NSE negatif-positif lemah, Chromogranin negatif-positif lemah dan CD99 positif lemah-sedang. Secara keseluruhan, berdasarkan pemeriksaan makroskopis, histopatologik, dan imunohistokimia disimpulkan sebagai suatu ?Malignant Small Round Cell Tumor? yang sesuai dengan PNET / ES (Ewing?s sarcoma) yang perlu di konfirmasi dengan pemeriksaan sitogenetik. (Med J Indones 2007; 16:108-12).
<hr>Primitive Neuro Ectodermal Tumor (PNET) is rare and difficult to diagnose. A case of PNET was diagnosed based on histopathological and immunohistochemical findings. A 4-month-old infant was admitted to the hospital with a tumor on the midline of his chest wall since he was 3 days old. The tumor was fixed on the chest wall and had ill-defined margin, enlarged over time and reached more than 10 cm in diameter when he was brought to a clinician. Two small ulcers were seen on the skin overlying the tumor. It was diagnosed as soft tissue tumor suggestive of a hemangioma. The tumor was 17 x 13 x 5.5 cm in size, white colored and firm to the touch. Microscopic examination revealed malignant small round cells with round to ovoid nuclei, coarse chromatin and scanty cytoplasm. Most cells were arranged in a solid pattern with scattered Homer-Wright rosettes. The mitotic count was 7/10 HPF, and necrosis was minimal (less than 25%). On immunohistochemical examination, the cells showed weak to moderate immunoreactivity to Vimentin and CD99, but showed negative to weak positive reactivity to NSE and Chromogranin. Based on the clinical features, gross findings, histopathologic and immunohistochemical examinations, the case was diagnosed as a malignant small round cell tumor consistent with PNET / ES (Ewing?s Sarcoma). To confirm the diagnosis, cytogenetic examination is suggested. (Med J Indones 2007; 16:108-12).