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Melanotic neuroectodermal tumor of infancy, analysis of 5 cases

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Introduction: Melanotic Neuroectodermal Tumour of Infancy (MNTI) is an extremely rare fast-growing neoplasm of infants with a biphasic population of neuroblastic cells and pigmented epithelial cells. The usual site affected is the craniofacial area.

Methods: We analyzed 5 cases of MNTI from 2012 to 2014. The specimens of 2 cases were examined grossly and fixed in 10% formalin. Tissue samples from 3 patients were from other hospitals. The specimens were embedded in paraffin and stained with haematoxylin and eosin and examined histologically. Immunohistochemical evaluation was performed with HMB-45, panCK (AE1/AE3), Synaptophysin, Desmin, S-100 Protein and CD99.

Results: The male/female ratio was 4/1. One tumour was congenital, the other 4 were revealed before the age 7 months of life. Tumours of four children were localized in the craniofacial area (3 in the maxilla, 1 in the left occipital bone). In the 7-months-girl the tumour was observed in the soft tissue of the left femur without any relation to the bone. The duration of symptoms was no longer than 4 months. The maximum diameter was 5 cm (occipital bone lesion). The tumours of all 5 patients were completely surgically excised. We have no data about local recurrences in our patients. Radiographically MNTI is the expansive radiolucency lesion, usually with poorly demarcated borders, and tendency to have locally invasive growth. Grossly the tumour is poorly encapsulated, firm, whitish-gray in color with black patches insight. Histologically MNTI consists of small neuroblastic cells and larger melanin-containing epithelial cells in a vascularized dense fibrous stroma. The epithelial component was reactive with HMB-45 and panCK (AE1/AE3) antibodies. The small cells expressed Synaptophysin and focally CD99. Neoplastic cells were negative for Desmin and S-100 Protein. MNTI should be differentiated with alveolar rhabdomyosarcoma, Ewing sarcoma, lymphomas, metastatic neuroblastoma, immature teratoma, and malignant melanoma.

Conclusion: We described 5 extremely rare cases of MNTI in children, one case in an unusual site (soft tissue of the left femur). MNTI should be differentiated with others children's tumours (alveolar rhabdomyosarcoma, Ewing sarcoma, lymphomas, metastatic neuroblastoma and immature teratoma).