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A TUMOR WHICH SHOULD BE KEPT IN MIND- CELLULAR NEUROTHECOMA

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Introduction: Cellular neurothekoma formerly considered as a nerve sheath tumor, is a dermal tumor of fibrohistiocytic origin. It usually occurs on head-neck and upper extremities of young adults, especially women. Diagnosis can be challenging in cases with atypical features. In our presentation we aimed to raise awareness to this tumor.

Case: A 6-year old boy presented with a 0,5 cm gray-yellow mass on his occipital region. The lesion had duration more than a-year without progression. He attended to pediatric surgery for excision. On microscopic evaluation the tumor extended from dermis to superficial subcutaneous adipose tissue with expansile borders. The tumor composed of nests formed by epithelioid cells with eosinophilic cytoplasm and vesicular nucleus. Between these areas few remnants of hair shafts were surrounded by multinucleated giant cells. The cells had no pleomorphism and only 1 mitosis/10 HPF was assessed. Immunohistochemistry was diffusely positive for MITF, CD68, NKI/C3 (CD63), Podoplanin (D2/40), CD10 and FXIIIa, but negative for S100.

Conclusion Neurothekoma was firstly described by Harkin and Reed as tumors of nerve sheath origin. In 1980, neurothekoma term was suggested by Gallager and Helwig. Cellular neurothekomas are often more cellular and have less myxoid stroma than neurothekomas. Plexiform fibrous histiocytoma should be considered in the differential diagnosis. Having similarities with clinical, morphological and phenotypic features, some authors suggested that they are a part of continuous spectrum. However, plexiform fibrous histiocytoma is a low grade tumor with a few cases of reported metastasis. The main points of differential diagnosis are: cellular neurothekomas are MITF, NKI/C3 (CD63) and Podoplanin (D2/40) positive while plexiform fibrous histiocytomas often have osteoclast-like multinuclear giant cells. It is important to know and make the differential diagnosis of these two tumors for clinical approach and follow-up.

Anahtar Kelimeler : cellular,neurothekoma, plexiform, fibrous, histiocytoma