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Photo Vignette

Erythematous nodular lesion on the chest of an infant

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Abstract

An 11-month-old girl presented with an erythematous nodule on the chest, which had been growing for 8 months. The tumor was composed of uniform polygonal and spindle-shaped cells, forming nodules and fascicles. The diagnosis of neurothekeoma was based upon the histology and immunohistochemistry.

Keywords: Cellular neurothekeoma; Juvenile xanthogranuloma; Neurofibroma; Neuroblastoma; Spitz nevus; Lymphoma.

Case synopsis

An 11-month-old girl presented with a cutaneous tumor located on the chest, dating from the third month of life. On examination, we observed a well-defined, 1x0.5 cm, oval and erythematous nodule located on the chest. The nodule was adherent to skin at its center (Figure 1). The regional lymph nodes were not enlarged. The nodule had gradually increased in size during the last few months. There was no history of seizures or other systemic symptoms. Blood tests showed hematological and biochemical parameters within normal limits. A chest X-ray and ophthalmological examination revealed no abnormalities. A punch biopsy was performed and later the lesion was excised under general anesthesia.



Figure 1. Erythematous, indurated nodular lesion on the chest of an infant.

Histological examination of the lesion showed a well-circumscribed and non-encapsulated tumor with a distinctly lobular pattern. The tumor was mainly composed of uniform polygonal and spindle-shaped cells, forming nodules and fascicles with some intervening collagenous stroma. No atypical mitoses were seen. Immunohistochemistry showed the tumor cells to be positive for NCI/C3. The tumor cells were negative for smooth muscle actin, S100, CD34, and EMA (epithelial membrane antigen) (Figure 2).

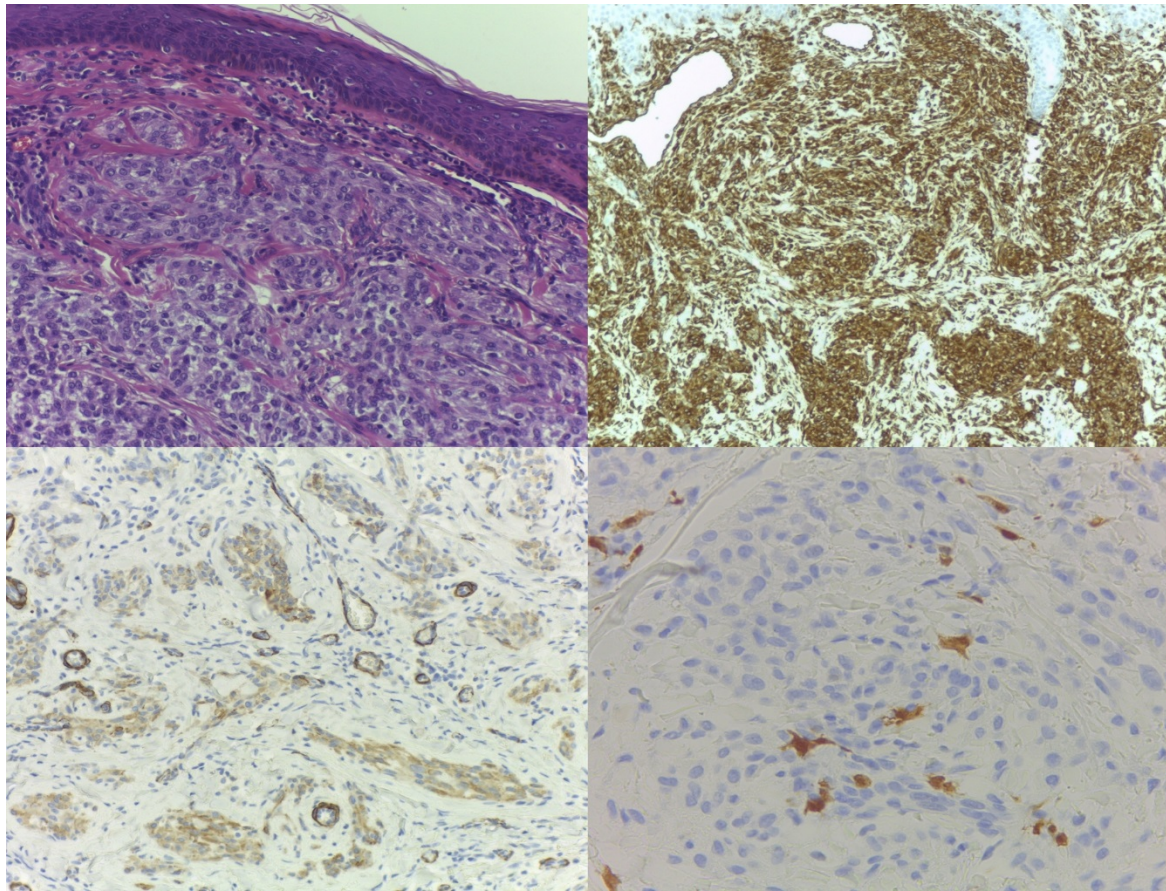


Figure 2. Histopathology and immunohistochemistry showed (a) a proliferation of uniform polygonal and spindle-shaped cells, forming nodules and fascicles with some intervening collagenous stroma (hematoxylin and eosin stain; original magnification 250x); (b) positive staining for NCI/C3 (c) negative staining for smooth muscle actin, and (d) S100 (original magnification 100x)

Diagnosis: Cellular neurothekeoma

Discussion

Cellular neurothekeoma typically presents as a solitary lesion that generally occurs in children or young adults [1-3]. Few cases mention patients having multiple tumors, usually located on the neck and/or head of young adult males [4, 5]. Neurothekeomas should be included in the differential diagnosis of dermal nodules in both children and infants. Other dermal tumors can clinically simulate neurothekeomas. Some of them include histiocytic tumors (juvenile xanthogranuloma), fibrous tumors (dermatofibroma and dermatofibrosarcoma protuberans), melanocytic tumors (malignant melanoma and Spitz nevus), lymphocytic tumors (lymphoma), muscular tumors (leiomyoma), neural tumors (neurofibroma and neuroblastoma) and sarcomatous tumors. The term "cellular neurothekeoma" was coined in 1986 by Rosati and cols. [1] to define a tumor they considered to be the cellular variant of the "myxoid neurothekeoma". Later on, mixed forms were described and the term "neurothekeoma" was considered to include the entire spectrum, from purely myxoid to predominantly cellular forms. Nevertheless, later studies showed that the clinical, immunohistochemical, ultrastructural, and even the genetic profile of both tumors are different, so that, whereas the myxoid neurothekeoma has a neural origin, the cellular neurothekeoma would have a fibrohistiocytic lineage [3]. Cellular neurothekeomas have a predominant epithelioid cell component and typically lack abundant stromal mucin. In addition, S100 protein expression is consistently lacking, although other neural and fibrohistiocytic markers, such as NSE, PGP9.5, NK1/C3, and CD163, are positive. Recently, some authors have described focal positivity for S100 protein in a minority of cases [6]. On the other hand, myxoid neurothekeoma is characterized by copious stromal mucin and a discreet proliferation of spindled and, less commonly, epithelioid cells. These lesions demonstrate evidence of Schwannian differentiation with constant positivity for S100 protein. Therefore, the diagnosis is based on histology and immunohistochemistry, to rule out other neoplasms. Although staining with NKI-C3 is always positive, this marker is not specific. Therefore, the negativity of other markers are much more relevant for the diagnosis.

Complete excision of the tumor is curative. Tumor persistence has been described when surgical excision is incomplete. Concerns have been raised about the possibility for malignant transformation of cellular neurothekeomas because some tumors contained cells with a variable degree of cytological atypia and variable mitotic activity suggesting a more aggressive

form. A study of 10 atypical cases did not find recurrence after surgery [2]. Therefore, these histological findings seem to have no prognostic significance at this time.

References

1. Rosati LA, Fratamico FC, Eusebi V. Cellular neurothekeoma. *Appl Pathol* 1986; 4:186-91. [PMID: 3297114]
2. Busam KT, Mentzel T, Colpaert C, et al. Atypical or worrisome features of cellular neurothekeoma: A study of 10 cases. *Am J Surg Pathol* 1998; 22:1067—72. [PMID: 9737238]
3. Argenyi ZB, LeBoit PE, Santa Cruz DJ, Swanson PE, Kutzner H. Nerve sheath myxoma (neurothekeoma) of the skin: light microscopic and immunohistochemical reappraisal of the cellular variant. *J Cutan Pathol* 1993; 20: 294. [PMID: 7693776]
4. Mahalingam M, Alter JN, Bhawan J. Multiple cellular neurothekeomas – a case report and review on the role of immunohistochemistry as a histologic adjunct. *J Cutan Pathol* 2006; 33: 51. [PMID: 16441413]
5. Garcia-Gutierrez M, Toussaint-Caire M, Gonzalez-Sanchez P, Ortiz-Hidalgo C. Multiple desmoplastic neurothekeomas localized to the face of a 16-year-old-boy. *Am J Dermatopathol* 2010; 32: 509. [PMID: 20571347]
6. Plaza JA, Torres-Cabala C, Evans H, Diwan AH, Prieto VG. Immunohistochemical expression of S100A6 in cellular neurothekeoma: clinicopathologic and immunohistochemical analysis of 31 cases. *Am J Dermatopathol*. Jul 2009;31(5):419-22. [PMID: 19542912]