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Cellular Neurothecoma: A Case Report of a 56-Year-Old Man

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Abstract

Cellular neurothecoma is a rare benign skin tumor, which must be distinguished from certain malignant tumors, in particular fibro-histiocytic plexiform cell tumors, clear cell sarcomas ... We report a new case of a 56-year-old man, who consulted for a skin lesion in the lumbar region, measuring approximately 2 cm in diameter. Histological examination showed a tumor proliferation located in the dermis and arriving at the superficial part of the hypodermis, made up of bundles of spindle cells and epithelioids with abundant eosinophilic cytoplasm, oval nucleus and fine chromatin. These bundles were separated by fibrous trabeculae isolating the tumor from the epidermis. The cells expressed CD10, CD99 and CD34 and were negative for PS100, GFAP, desmin, EMA, AML and STAT6. Thus, the diagnosis retained was that of a cellular neurothecoma.

Keywords: Neurothecoma • Benign skin tumor • Rare

Introduction

Cellular neurothecoma is a rare benign tumor, it preferentially affects young women. It is usually localized on the trunk, head and neck [1,2]. This tumor rarely recurs, especially in case of incomplete excision [3]. Its morphological appearance and immunohistochemical profile help to distinguish it from other tumors, including myxoid neurothecoma, Spitz's nevus, plexiform fibrohistiocytic tumor, piloleiomyoma and clear cell sarcoma.

Case Presentation

We report a new case of a 56-year-old man, who consulted for a tumor in the lumbar region that had been developing for less than a year. Clinical examination revealed a tumor lesion approximately 2cm in diameter, located in the lumbar region. The lesion was lumpy, solitary, superficial and soft (Figure 1). On its surface ran fine telangiectasias. It was painless. The lymph node areas were free. Complete surgical excision was performed. The histological study of the excisional biopsy has showed a poorly defined lesion, located in the dermis and reaching the superficial hypodermis, with a fasciculate architecture made up of spindle-shaped cells and epithelioids with abundant eosinophilic cytoplasm, with an oval nucleus, fine chromatin (Figure 2) and 2 mitoses for 10 fields were observed at high magnification. These bundles were separated by fibrous spans isolating the tumor from the epidermis. In immunohistochemical study, cells expressed CD10 (Figure 3), CD99 and CD34. They were negative for PS100 (Figure 4), GFAP, desmin, EMA, AML and STAT6. Thus, the diagnosis retained was the cellular neurothecoma.

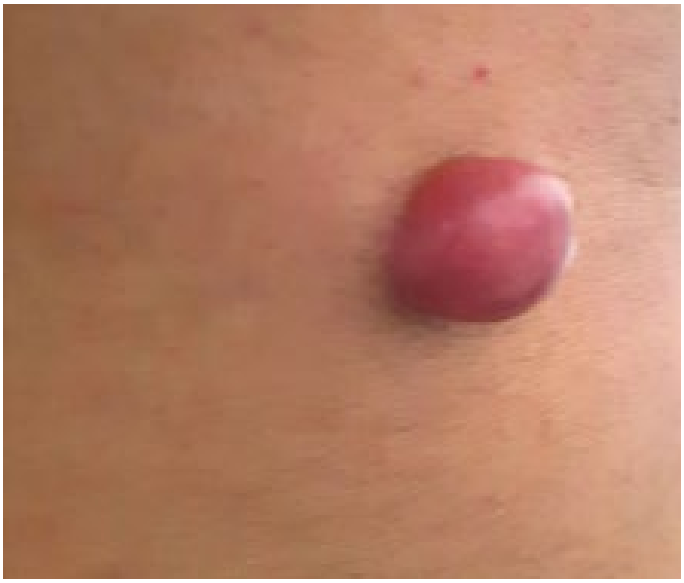

Figure 1. A picture showing the skin lesion macroscopically.

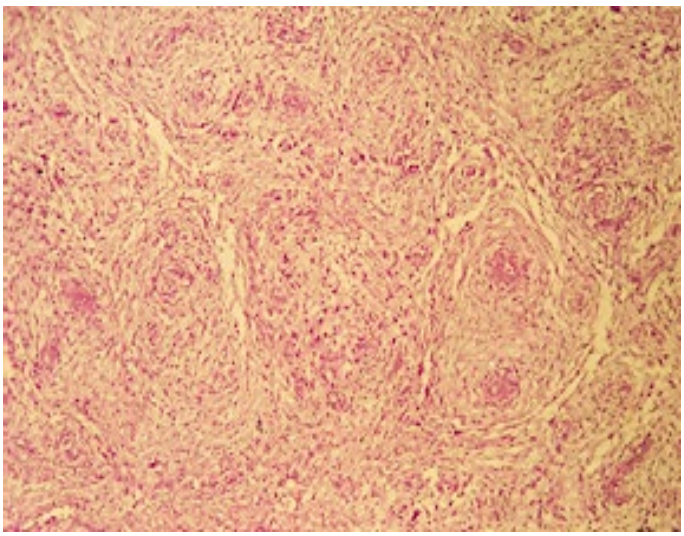

Figure 2. Fasciculated architecture made up of spindle-shaped cells and epithelioids (H.E 40X).

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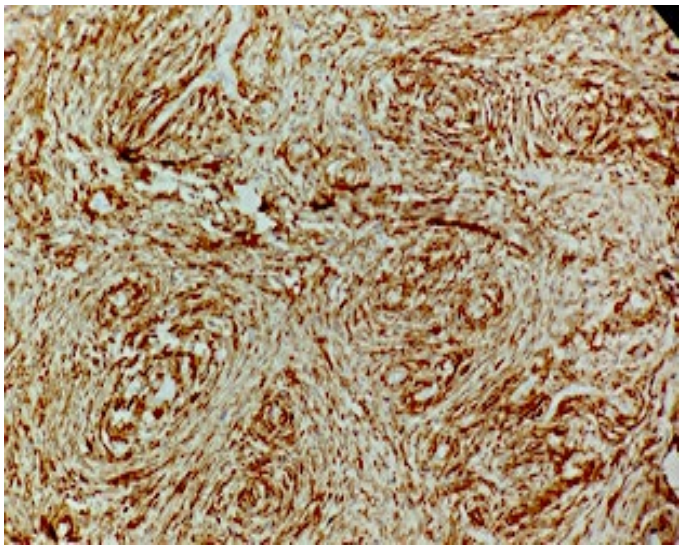

Figure 3. Positivity of CD10 in tumor cells.

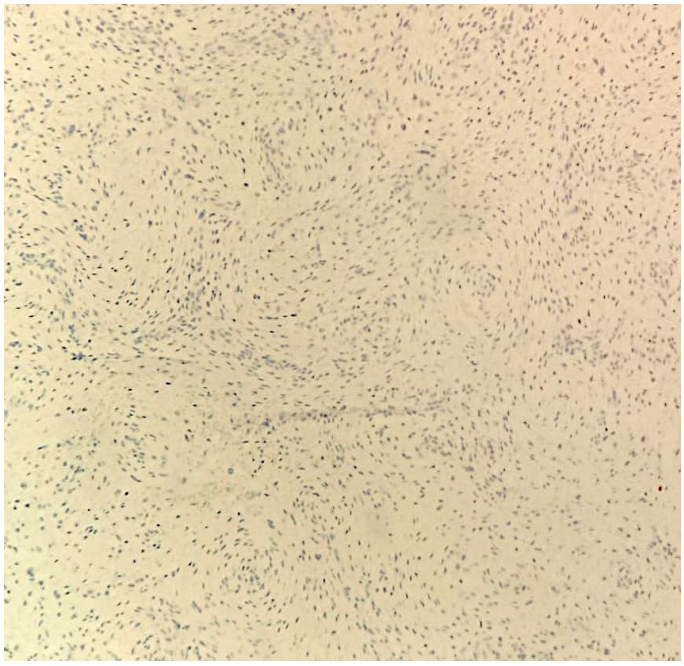

Figure 4. Negativity of S100 protein in tumor cells.

Results and Discussion

Cellular neurothecoma is a rare skin tumor of uncertain histogenesis [4]. It was first described by Gallager and Helwig in 1980, who proposed the term to reflect the presumed origin of the lesion from the nerve sheath [5]. Then in 1986 Rosati LA, et al. [6] distinguished it from neurothecoma myxoid by histological, immunohistochemical and ultrastructural characteristics. The diagnosis of this tumor is histological. Cellular neurothecoma is more common in women (sex ratio F/M=1.6[3] to 4.3[7]). The average age at diagnosis is 23 years (the extreme ages are: 1 to 65 years [3]). It preferentially reaches the head, the neck and the trunk [7], cellular neurothecoma is a tumor of the cervico-facial and scapulo-humeral dermis of the young woman. It takes the form of a solitary firm induration, usually less than 1 cm, without epidermal ulceration. An unusual case is reported by Chow LTC, et al [8]: the 6.5 cm tumor was hypopharyngeal.

Some rare localization can be seen such as the oral mucosa (cheeks, gums, palate and lips), vulva and exceptionally para-areolar, spinal, subdural, intracranial, or maxillary sinus. The three characteristics of our clinical case are: Elder age, male sex and localization on the lumb. Three cases of localization in the chin have been reported by Bonte E, et al. [9], in a 48-year-old woman. Neurothecoma chin myxoid was also described by Pasquier B, et al. [7] in a 16-year-old girl. And Brix M, et al. [10] describes a case of Chin localisation in an 11-year-old child. Two cases of Cellular neurothecomas have been reported in 14 years old girls, in the right shoulder and arm [3].

Macroscopically, the tumor nodule is colorless, or lightly red, with a soft consistency, it usually measures 1 cm in diameter. It is sometimes lobulated and may have myxoid sectors and more rarely central calcification, surrounded by a gelatinous crown. Histologically, the neurothecoma is characterized by a well-defined tumor proliferation, respecting the epidermis and spreads the dermal collagen bundles and can infiltrate the hypodermis; lobules that are compact or dissociated by myxoid imbibition, juxtaposed within fibrosis; large tumor cells, often spindle-shaped; epithelioid-looking cell clumps; nuclear polymorphism with atypical nuclei. Cellular neurothecoma is made up of lobules of cohesive cells, often epithelioid, in a stroma predominantly fibrous.

In immunohistochemistry, the presence or absence of cell marking makes it possible to differentiate the forms myxoid and cellular neurothecoma and ensure the differential diagnosis. Tumor cells are positive for NKI-C3 in 100% of cases (melanocyte marker), NSE, CD10 and vimentin. Most often, the marking by anti-S100 protein stain is negative, however in the case of Brix M, et al. [10] it was positive, but this does not exclude the diagnosis, as the markers: NSE, vimentin and NKI-C3 were typically positive.

The clinical features can eliminate a hemangioma, a dermal naevus, a sebaceous cyst or a dermatofibroma. A case has been reported of an association of a myxoma with a neurinoma [11] and neurothekoma with a typical neurofibroma [12] within the same tumor nodule. The cellular neurothecoma must be differentiated from the naevus of spitz Plexiform, neurotropic malignant melanoma, clear cell sarcoma, schwannoma, neurofibroma plexiform. It is the immunohistochemical study that confirms the

diagnosis in the absence of S100 protein expression and HMB45 by tumor cells. The tumor progresses slowly. It can progress during several months or even several years before diagnosis. After resection, the evolution is favorable. Cellular neurothecoma rarely recurs, it seems only in case of incomplete excision [13], in 3 cases of a series of 123 [12]. The treatment of cellular neurothecoma is the complete surgical excision ensured by histological control.

Conclusion

Neurothecoma will probably come out of the chapter on nerve tumors when its histogenesis is clearly elucidated. Despite its poorly limited and infiltrating nature, the possibility of atypia and mitosis, it is a benign tumor which must be completely removed to avoid recurrence.

References

1. Hornick, Jason L. and Christopher DM Fletcher. "Cellular neurothekeoma: Detailed characterization in a series of 133 cases." *Am J Surg Pathol* 31 (2007): 329-340.
2. Fetsch, John F., William B. Laskin, James R. Hallman and George P. Lupton, et al. "Neurothekeoma: An analysis of 178 tumors with detailed immunohistochemical data and long-term patient follow-up information." *Am J Surg Pathol* 31 (2007): 1103-1114.
3. Chatelain, D., J. Ricard, M. Colombat and C. Ghighi, et al. "Cellular neurothekeoma, a rare cutaneous tumor. Anatomo-clinical and immunohistochemical study of 2 cases." *Ann Pathol* 20 (2000): 225-227.
4. Argenyi, Zsolt B., Philip E. LeBoit, Daniel Santa Cruz and Paul E. Swanson, et al. "Nerve sheath myxoma (neurothekeoma) of the skin: Light microscopic and immunohistochemical reappraisal of the cellular variant." *J Cutan Pathol* 20 (1993): 294-303.
5. Gallager, Richard L. and Elson B. Helwig. "Neurothekeoma a benign cutaneous tumor of neural origin." *Am J Clin Pathol* 74 (1980): 759-764.
6. Rosati, L. A., F. C. Fratamico and V. Eusebi. "Cellular neurothekeoma." *Appl Pathol* 4 (1986): 186-191.

7. Pasquier, B., R. Barnoud, M. Peoc'h and N. Pinel, et al. "Neurothekeoma: General review apropos of an anatomoclinical case with immunohistochemical and ultrastructural study." *Arch Anat Cytol Pathol* 42 (1994): 133-140.

8. Chow, L. T. C., T. K. F. Ma and W. H. Chow. "Cellular neurothekeoma of the hypopharynx." *Histopathology* 30 (1997): 192-194.

9. Bonte, E., S. Fraitag and N. Brousse. "Un nodule cutané du menton." In Annales de pathologie (Paris) 16(1996) 137-138.

10. Brix, M., E. Ranfaing, F. Lesne and B. Ricbourg. "Cellular neurothekoma in an 11-year-old child." *Rev Stomatol Chir Maxillofac* 106 (2005): 356-359.

11. Reed RJ and Harkin JC. "Perineurial myxoma (nerve sheath myxoma). In: Tumors of the peripheral nervous system." *Atlas Tumor Pathol* (1983): 16-18.

12. FM, Enzinger. "Neurothekeoma (nerve sheath myxoma)." Soft tissue tumors (1995).

13. Barnhill, Raymond L., G. Richard Dickersin, Volker Nickeleita and Atul K. Bhan, et al. "Studies on the cellular origin of neurothekeoma: Clinical, light microscopic, immunohistochemical and ultrastructural observations." *J Am Acad Dermatol* 25 (1991): 80-88.

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