EDORIUM Journals

J Case Rep Images Pathol 2021;7(1):7–10.
www.ijcripathology.com

CASE REPORT PEER REVIEWED | OPEN ACCESS

Neuroblastoma-like schwannoma: An unusual morphologic variant

Preethi Dileep Menon, Josefine Heim-Hall, Alia Nazarullah

ABSTRACT

Introduction: Neuroblastoma-like schwannoma is a rare morphologic variant of schwannoma with formation of large rosettes.

Case Report: A 29-year-old woman presented with a longstanding left chest wall soft tissue mass. Histology showed small monomorphic tumor cells radially arranged around a fibrocollagenous core forming large rosettes. No evidence of mitotic activity or necrosis was identified. Diffuse S100 reactivity by immunohistochemistry helped confirm the diagnosis as neuroblastoma-like schwannoma.

Conclusion: Awareness of this unusual variant of schwannoma will avoid misinterpretation and confusion with other benign and malignant rosette forming tumors.

Keywords: Neuroblastoma, Rosette, Schwannoma

How to cite this article

Menon PD, Heim-Hall J, Nazarullah A. Neuroblastoma-like schwannoma: An unusual morphologic variant. J Case Rep Images Pathol 2021;7(1):100049Z11PM2021.

Article ID: 100049Z11PM2021

Preethi Dileep Menon[1], MD, Josefine Heim-Hall[1], MD, Alia Nazarullah[1], MD

Affiliations: [1]Department of Pathology and Laboratory Medicine, University of Texas Health, 7703 Floyd Curl Drive, MC#7750, San Antonio, Texas, USA.

Corresponding Author: Alia Nazarullah, MD, Assistant Professor/Clinical, Department of Pathology and Laboratory Medicine, University of Texas Health, 7703 Floyd Curl Drive, MC#7750, San Antonio, Texas 78229, USA; Email: nazarullah@uthscsa.edu

Received: 08 December 2020
Accepted: 15 January 2021
Published: 06 March 2021

****

doi: 10.5348/100049Z11PM2021CR

INTRODUCTION

Neuroblastoma-like schwannoma is a subtype of schwannoma that is extremely rare with 25 previously reported cases [1]. This subtype was first described by Goldblum et al. [2] in 1994. According to the literature review conducted by KoubaaMahjoub et al., the lesion is thought to arise predominantly in females with a median age of 32.2 years [1] and most commonly involved the limbs. The rarity of this lesion and its most striking feature, rosette like formation, may lead to confusion with other benign and malignant entities characterized by rosettes and pseudorosettes, such as neuroblastoma, dendritic cell neurofibroma with pseudorosettes, hyalinizing spindle cell tumor with giant rosettes (low-grade fibromyxoid sarcoma) and Ewing sarcoma. We report a case of a 29-year-old woman with a schwannoma with neuroblastoma-like rosettes involving the chest wall soft tissue. The following case report emphasizes the need to have a high index of suspicion for this entity and the utility of morphologic clues and characteristic immunophenotype to avoid a diagnostic pitfall.

CASE REPORT

A 29-year-old woman presented with a left chest wall soft tissue lesion for 1 year. The lesion was surgically excised. Microscopic examination showed a well circumscribed, encapsulated lesion with many large rosette-like structures, composed of small round cells radially arranged around a central eosinophilic fibrocollagenous core (Figure 1A–C). These small cells were monomorphic, round to slightly elongated, with scant amount of cytoplasm, hyperchromatic nuclei, and prominent nucleoli. Perivascular hyalinized rosettes were also present. There were no mitoses or areas of necrosis identified. Immunohistochemical stain with S100 showed that these cells were strongly and diffusely positive with both cytoplasmic and nuclear staining (Figure 1D).

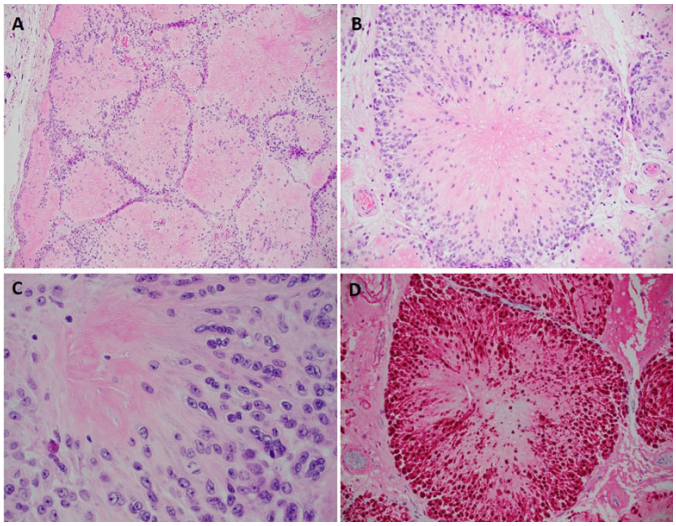

Figure 1: Light microscopy and immunohistochemical features of neuroblastoma-like schwannoma. (A)–(C) Low-, medium-, and high-power H&E stain (2×, 10×, and 40×) showing large rosette-like structures, composed of small round cells radially arranged around a central eosinophilic fibrocollagenous core. (D) Medium power S100 immunohistochemistry stain (10×) showing strong and diffuse positivity in the small round cells.

These features were consistent with schwannoma with neuroblastoma-like rosettes. No areas with features of conventional schwannoma were identified within the lesion.

DISCUSSION

Neuroblastoma-like schwannoma is a rare subtype of schwannoma, with histological features resembling neuroblastoma. This subtype was initially described by Goldblum et al. [2] in 1994 in a case series of three cases. Few case reports followed [3–8] highlighting a range of morphologic features from small, ovoid cells to epithelioid cells comprising the lesion. Although considered a sporadic lesion, two cases of neuroblastoma-like schwannomatosis have been reported [3, 9].

This lesion is characterized by a well circumscribed lesion with many large rosette-like structures, composed of small round cells radially arranged around a central eosinophilic fibrocollagenous core. However, smaller rosettes that somewhat resemble Homer-Wright rosettes have also been described in these lesions [2, 10–12]. The rarity of this lesion and its most striking feature, rosette-like formation, may lead to confusion with other entities characterized by rosettes and pseudorosettes, such as neuroblastoma, dendritic cell neurofibroma with pseudorosettes [6], hyalinizing spindle cell tumor with giant rosettes (low-grade fibromyxoid sarcoma) [6, 7] and Ewing sarcoma/primitive neuroectodermal tumor (ES/PNET).

Neuroblastoma can be distinguished from neuroblastoma-like schwannoma by the absence of capsule, presence of true Homer-Wright rosettes 3,8 that are much smaller than the rosettes found in neuroblastoma-like schwannoma and that contain neuron-specific enolase positive fibrillary structures. S100 expression is only focal in the neuroblasts and stromal cells while it is strong and diffuse in neuroblastoma-like schwannoma [8].

Dendritic cell neurofibroma with pseudorosettes is a solitary well-circumscribed, painless lesion arising in the dermis on the extremities, trunk, and head. Histologically it is composed of small hyperchromatic lymphocyte-like cells surrounding larger cells with long dendritic extensions forming the pseudorosettes. The larger cells are more strongly positive for S100 protein than the smaller lymphocyte-like cells [13]. Distinguishing features between dendritic cell neurofibroma and neuroblastoma-like schwannoma includes small pseudorosettes with large cells in their centers that stain with S100 protein and CD34 in the former in contrast to the large rosettes with acellular centers, strong diffuse staining with S100 protein and absent CD34 staining in the latter [13].

Hyalinizing spindle cell tumor with giant rosettes, a variant of low-grade fibromyxoid sarcoma, typically arises below the fascia in the extremities and trunk. It has giant rosettes composed of epithelioid fibroblasts which are S100 immunoreactive, cuffed around a central hyalinized core of collagen [14, 15]. Features that help to distinguish low grade fibromyxoid sarcoma from neuroblastoma-like schwannoma include spindled cells in a whorling growth pattern, arcades of curvilinear blood vessels, MUC4 expression, lack of diffuse S100 reactivity and fibromyxoid sarcomatous areas in the former [15–17].

Ewing sarcoma/primitive neuroectodermal tumor (ES/PNET) is also included in the differential due to the presence of small round blue cell and Homer-Wright rosettes in Ewing sarcoma. This can be differentiated from neuroblastoma-like schwannoma by the lack of encapsulation, morphology of densely packed small round blue cells, membranous staining with CD99 and specific translocations associated with Ewing sarcoma [18]. In addition, tumor cells in Ewing sarcoma are mitotically active with ill-defined glycogen rich cytoplasm and often show areas of tumoral necrosis.

Neuroblastoma-like schwannoma is considered a neoplasm with benign behavior in most cases reported. No cases of recurrence or metastasis have been reported for this variant. Hence complete excision of this lesion is recommended as the choice of treatment [12]. However, a single case of malignant peripheral nerve sheath tumor arising in a patient with neuroblastoma-like schwannomatosis has been reported [9]. Hence due to the limited number of the reported cases and rarity of this lesion, additional studies with follow-up of the patients will help to better understand the pathophysiology and prognosis of this lesion and guide with adequate treatment.

CONCLUSION

In conclusion, neuroblastoma-like schwannoma is an unusual variant of schwannoma with relatively few reported cases which may be misinterpreted as a malignant neoplasm. This error can be avoided by awareness of the existence of this entity which is characterized by the presence of large rosette-like structures with small round, monomorphic cells radially arranged around a central eosinophilic fibrocollagenous core, absence of atypia and mitosis, and intense reactivity for S100.

REFERENCES

1. Koubaa Mahjoub W, Jouini R, Khanchel F, et al. Neuroblastoma-like schwannoma with giant rosette: A potential diagnostic pitfall for hyalinizing spindle cell tumor. J Cutan Pathol 2019;46(3):234–7.
2. Goldblum JR, Beals TF, Weiss SW. Neuroblastoma-like neurilemoma. Am J Surg Pathol 1994;18(3):266–73.
3. Sulhyan KR, Deshmukh BD, Gosavi AV, Ramteerthakar NA. Neuroblastoma-like schwannoma in a case of schwannomatosis: Report of a rare case. Int J Health Sci (Qassim) 2015;9(4):478–81.
4. Fisher C, Chappell ME, Weiss SW. Neuroblastoma-like epithelioid schwannoma. Histopathology 1995;26(2):193–4.
5. Bhatnagar S, Banerjee SS, Mene AR, Prescott RJ, Eyden BP. Schwannoma with features mimicking neuroblastoma: Report of two cases with immunohistochemical and ultrastructural findings. J Clin Pathol 1998;51(11):842–5.
6. Sharma P, Chatterjee D, Das A. Neuroblastoma like schwannoma: A diagnostic challenge. BMJ Case Rep 2017;5;2017: bcr2017220721.
7. Lewis ZT, Geisinger KR, Pichardo R, Sangueza OP. Schwannoma with neuroblastoma-like rosettes: An unusual morphologic variant. Am J Dermatopathol 2005;27(3):243–6.
8. de Saint Aubain Somerhausen N, Valaeys V, Geerts M, André J. neuroblastoma-like schwannoma: A case report and review of the literature. Am J Dermatopathol 2003;25(1):32–4.
9. Carter JM, O'Hara C, Dundas G, et al. Epithelioid malignant peripheral nerve sheath tumor arising in a schwannoma, in a patient with "neuroblastoma-like" schwannomatosis and a novel germline SMARCB1 mutation. Am J Surg Pathol 2012;36(1):154–60.
10. Gutte RM. SkIndia quiz 12: A firm yellowish nodule over lip. Indian Dermatol Online J 2014;5(1):100–2.
11. Sedassari BT, da Silva Lascane NA, Cury Gallottini MH, Orsini Machado de Sousa SC, Pinto Júnior DDS. Neuroblastoma-like schwannoma of the lower labial mucosa: A rare morphologic variant of peripheral nerve sheath tumor. Oral Surg Oral Med Oral Pathol Oral Radiol 2014;118(5):579–82.
12. Kaur K, Kakkar A, Binyaram, et al. Neuroblastoma-like schwannoma of the skull base: An enigmatic peripheral nerve sheath tumor variant. Neuropathology 2016;36(6):573–8.
13. Michal M, Fanburg-Smith JC, Mentzel T, et al. Dendritic cell neurofibroma with pseudorosettes: A report of 18 cases of a distinct and hitherto unrecognized neurofibroma variant. Am J Surg Pathol 2001;25(5):587–94.
14. Folpe A, van den Berg E, Molenaar WM. Low grade fibromyxoid sarcoma. In: Fletcher CDM, Krishnan Unni K, Mertens F, editors. Pathology and Genetics Tumours of Soft Tissue and Bone. World Health Organization classification of tumours. Lyon, France: IARC Press; 2002. p. 104–5.
15. Lane KL, Shannon RJ, Weiss SW. Hyalinizing spindle cell tumor with giant rosettes: A distinctive tumor closely resembling low-grade fibromyxoid sarcoma. Am J Surg Pathol 1997;21(12):1481–8.
16. Woodruff JM, Antonescu CR, Erlandson RA, Boland PJ. Low-grade fibrosarcoma with palisaded granulomalike bodies (giant rosettes): Report of a case that metastasized. Am J Surg Pathol 1999;23(11):1423–8.
17. Farinha P, Oliveira P, Soares J. Metastasizing hyalinizing spindle cell tumour with giant rosettes: Report of a case with long survival. Histopathology 2000;36(1):92–3.
18. Ozaki T. Diagnosis and treatment of Ewing sarcoma of the bone: A review article. J Orthop Sci 2015;20(2):250–63.

****

Author Contributions

Preethi Dileep Menon – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Interpretation of data, Drafting the work, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Josefine Heim-Hall – Design of the work, Analysis of data, Interpretation of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Alia Nazarullah – Conception of the work, Design of the work, Acquisition of data, Analysis of data, Revising the work critically for important intellectual content, Final approval of the version to be published, Agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved

Guarantor of Submission

The corresponding author is the guarantor of submission.

Source of Support

None.

Consent Statement
Written informed consent was obtained from the patient for publication of this article.

Conflict of Interest
Authors declare no conflict of interest.

Data Availability
All relevant data are within the paper and its Supporting Information files.

Copyright