

Schwannoma With Extensive Vascular Hyperplasia in the Breast Mimicking Angiosarcoma

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1. Abstract

Schwannoma with extensive vascular hyperplasia in the breast is extremely rare. It is typically benign and slow growing. Here, we describe a case of 54-year-old woman underwent complete resection of a tumor measuring 2.5×2.0 cm in the breast. The tumor was composed with spindle cell component and abundant blood vessel component. The subcapsular cellular component was composed of spindleshaped cells arranged in bundle and palisading pattern. There was vascular hyperplasia, anastomoses, congestion and cellulose effusion in the central area. The spindle-shaped cells possess modest amounts eosinophilic cytoplasm, no discernible cell borders, and normochromatic elongated tapered nuclei. Focal cells were epithelioid with mild atypia in vascular hyperplasia area. This case was misinterpreted as a low grade angiosarcoma in the initial diagnosis. Immuno histochemical stains demonstrated that spindled cells were immuno reactive for S100 and Sox10. The focal atypical cells were negative for S-100, Sox10, CD34, ERG, CD31, SMA and desmin. The proliferative index Ki-67 was 3%. These findings confirmed to be schwannoma. The patient has been followed up for 6 months postoperatively with no evidence of tumor recurrence. We report an extremely rare intramammary schwannoma with extensive vascular hyperplasia, along with a review of relevant literature.

2. Introduction

Schwannoma is a nerve sheath tumour composed entirely or nearly entirely of differentiated neoplastic schwann cells [1]. Schwannomas of the breast are rare, accounting for <3% of all schwannomas and <1% of all primary breast neoplasms. Schwannomas are typically benign, slow growing, and asymptomatic uncommon <http://acmcasereports.com>

neurogenic tumors occurred in breast [2]. In particular, schwannoma with extensive vascular hyperplasia occurred in the breast is extremely rare. Recently, we experienced a case of intramammary schwannoma with extensive vascular hyperplasia mimicking angiosarcoma that developed in a middle-aged woman. We report this case which can be confused with angiosarcoma.

3. Case Report

A 54-year-old woman was referred to the Breast Surgery Department due to a slow growing palpable lump at right breast for 5 years. On physical examination, a firm, mobile, smooth, tender mass measuring 2.5×2.0 cm was palpated in the inner lower quadrant of right breast. Nipple discharge, abnormal skin findings and axillary adenopathy were not found. Ultrasonography revealed a regular, well delineated, isoechoic mass of right breast measuring 2.1×1.3 cm (Figure 1), with abundant blood stream signals. A complete surgical excision of the nodule was performed.

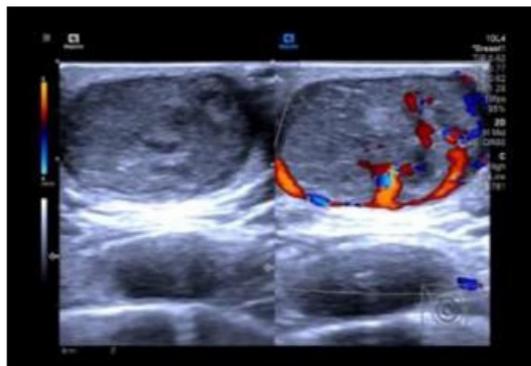


Figure 1: Ultrasonography revealed a regular, well delineated, isoechoic mass of right breast measuring 2.1×1.3 cm, with abundant blood stream signals.

3.1. Pathological findings

Grossly, the tumor was an encapsulated, well-circumscribed, solid mass measuring about 2.5×2.0×1.2cm (Figure 2A). The cut surface of the mass was grayish white, with moderate hardness, hemorrhage was seen (Figure 2B). Histological examination revealed a well-defined tumor surrounded by a fibrous capsule. The tumor was composed with a cellular component (dark area) and abundant blood vessel component (Figure 3A). The subcapsular cellular component was composed of spindle-shaped cells arranged in bundle (Figure 3B) and palisading pattern (Figure 3C). There was vascular hyperplasia, anastomoses, congestion and cellulose effusion in the central area (Figure 3D). Some vascular were hyalinization. Lymphocytic and multinucleated giant cells, hemosiderin were seen. The spindle-shaped cells posed modest amounts eosinophilic cytoplasm, no discernible cell borders, and normochromat-

ic elongated tapered nuclei (Figure 3E). Focal cells were epithelioid, with mild atypia in the vascular hyperplasia area mimicking angiosarcoma (Figure 3F). Mitotic figures were rare. This case was misinterpreted as low grade angiosarcoma in the initial diagnosis.

Immuno histochemical stains demonstrated that spindle cells were immuno reactive for S-100 (Figure 4A,B), Sox10 (Figure 4C), vimentin. No immuno reactivity was obtained with pan-CK, p63, CD34, ERG, CD31, ER, PR, HMB45, Melan-A, SMA, desmin and SMHHC in spindle cells. The focal atypical cells were negative for S-100, Sox10, CD34 (Figure 4D), ERG (Figure 4E), CD31, SMA, desmin. The focal atypical cells were positive for CD68. The proliferative index of tumor cells, detected with Ki-67, was 3% (Figure 4F), as well as in focal atypical cells. The immuno histochemical findings supported a diagnosis of degenerated (ancient) schwannoma with extensive vascular hyperplasia.



Figure 2: Gross tumor specimen of breast's schwannoma. (A)The mass was an encapsulated, well-circumscribed, solid mass measuring about 2.5×2.0×1.2cm. (B) The cut surface of the mass was grayish white, with moderate hardness, hemorrhage.

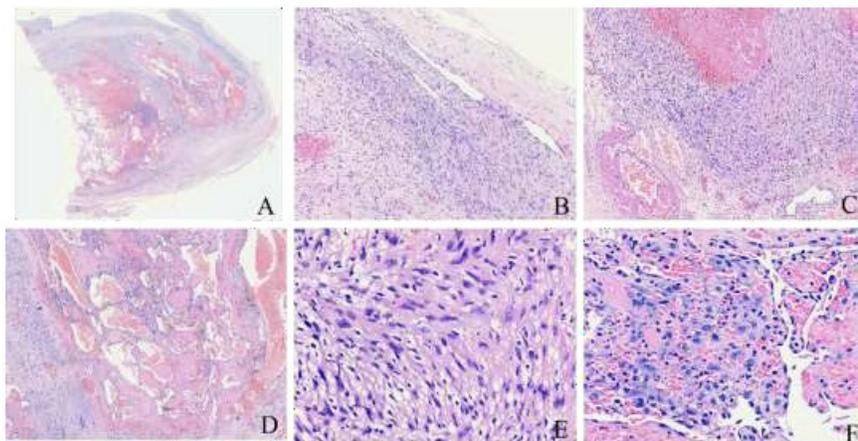


Figure 3: Histological findings of schwannoma of the breast. (A) A well-defined tumor surrounded by a thin fibrous pseudocapsule (slice scanning, 0.35×). (B) The sub capsular spindle-shaped cells were arranged in bundle (100×). (C) Paravascular spindle-shaped cells were arranged in palisading pattern (100×)(D)There were vascular hyperplasia, anastomoses, congestion and cellulose effusion in the central area (40×)(E)The spindle-shaped cells were modest amounts eosinophilic cytoplasm and normochromatic elongated tapered nuclei (400×). (F) Focal cells were epithelioid, with mild atypia in the vascular hyperplasia area (400×).

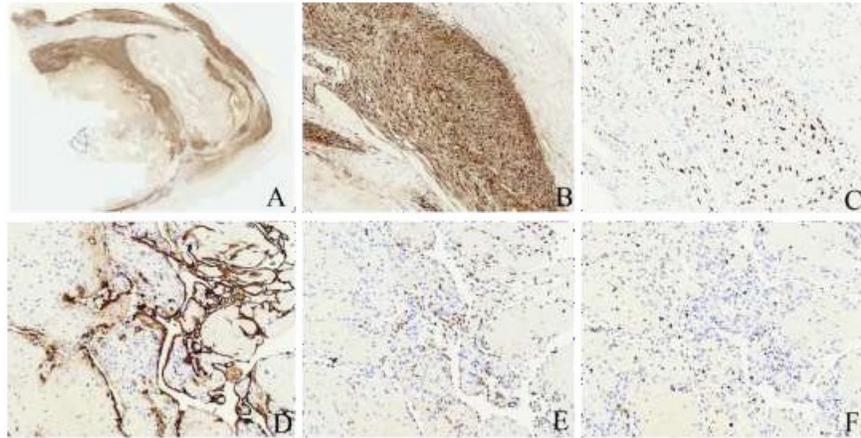


Figure 4: Immuno histochemical findings of schwannoma of the breast. (A) The cellular component was immuno reactive for S100 (slice scanning, 0.35 \times). The spindle -shaped cells were immuno reactive for S100 (B) and Sox10(C) (100 \times). The focal atypical cells were negative for CD34 (D), ERG (E). (F) The proliferative index Ki-67 was 3%.

4. Discussion

Benign and malignant peripheral nerve sheath tumors can involve the breast, presenting as masses in the dermis, deep breast parenchyma or axillary soft tissue [3]. Nearly 40 cases with schwannoma occurred in the breast have been reported up to 2020. The size of those masses was between 0.7 and 11 cm with a mean of 3.7 cm. The age range was 13–83 years with a mean of 48.6 years [4]. There are several subtypes including ancient schwannoma, cellular schwannoma, plexiform schwannoma, epithelioid schwannoma, microcystic/reticular schwannoma. The recognition of the wide morphologic spectrum exhibited by schwannoma, is crucial to avoid confusion with other benign or malignant breast lesions. Ancient schwannoma differs from the conventional schwannoma only by its presence of scattered atypical to bizarre-appearing nuclei, a feature that is often considered degenerative. Such cases may show extensive hyalinization or central ischemic changes [1]. Ancient schwannoma, characterized by degeneration due to long course, is rare and can be mistaken for malignancy due to heterogeneous intensity and degeneration evident on MRI and nuclear atypia on histopathology [5], so the ancient schwannoma may represent a diagnostic pitfall. The degenerative changes in schwannoma include pseudo glandular cystic spaces [6], significant cystic change [7], gross calcification [8], widespread foam cell infiltration [9], extensive myxomatous degeneration and multiple vessel thrombosis [10]. In our case, extensive vascular hyperplasia arranged in anastomoses, focal mild moderate atypical cells, simulating a low grade angiosarcoma. This case was misinterpreted as a low grade angiosarcoma in the initial diagnosis. However, a slow-growing tumor for 5 years, the well-circumscribed borders and smooth external surface of the mass, and the absence of mitoses were features against malignancy. Immuno reactivity with S100 and Sox10 in spindled cells supported tumour originating from schwann cells. The focal atypical cells were negative for CD34, ERG and CD31, which don't support these cells originating from

endothelial cells. The focal atypical cells were also negative for SMA, desmin, which don't support these cells originating from fibroblastic/myofibroblastic cells. The focal atypical cells were positive for CD68, supporting these cells were histiocyte. Extensive vascular hyperplasia may be a degenerative change. In addition, most schwannomas are cytokeratin negative, but focal cytokeratin labeling has been reported, posing a potential diagnostic pitfall with spindle cell carcinoma [11].

The differential diagnosis of breast schwannoma includes neurofibroma and breast spindle cell lesions such as fibromatosis-like metaplastic carcinoma, myofibroblastoma, fibromatosis, pseudo angiomatous stroma hyperplasia, nodular fasciitis, solitary fibrous tumor, leiomyoma, phyllodes tumor, low-grade sarcoma, reactive changes after injury [11], that of ancient schwannoma includes malignant peripheral nerve sheath tumor (MPNST). Conventional MPNSTs are hyper cellular, with atypical, pleomorphic spindled cells, a brisk mitotic rate, and a herringbone or storiform architecture. In our case, lack of mitotic and cell pleomorphism, a low proliferative index confirms a benign tumour. Palisaded myofibroblastoma of the breast closely mimicked schwannoma [12]. Palisaded myofibroblastoma were diffusely immuno reactive for vimentin, desmin, α -smooth muscle actin (SMA), calponin, estrogen receptor (ER), progesterone receptor (PR), no immuno reactivity was obtained with S100 and Sox 10.

5. Conclusion

In conclusion, we reported an extremely rare case of schwannoma with extensive vascular hyperplasia, mimicking angiosarcoma, which was located at an unusual site. The recognition of degenerative changes in schwannoma is vital to avoid mistaking schwannoma for sarcomas or other soft-tissue neoplasms.

6. Conflict of Interest

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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