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CASE REPORT

Psammomatous Melanotic Schwannoma Not Associated with Carney Complex: a Case Report

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ABSTRACT —— ***Background.*** Psammomatous melanotic schwannoma is a rare neoplasm that may arise in the context of the Carney complex, a unique multiple endocrine neoplasia syndrome. ***Case.*** We herein report a case of psammomatous melanotic schwannoma not associated with the Carney complex, arising in the right paraspinal area of a 26-year-old woman. She underwent en bloc total resection of the mass and has exhibited a good clinical course for 10 years since tumor resection. ***Conclusion.*** Most melanotic schwannomas are slow-growing tumors; however, the prognosis can be unfavorable in cases with local recurrence or malignant behavior. This type of tumor is curable via resection and should be distinguished from other pigmented tumors, such as malignant melanoma.

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KEY WORDS —— Psammomatous melanotic schwannoma, Carney complex, Malignant melanoma, Thoracic

INTRODUCTION

Melanotic schwannoma (MS) is a rare nerve sheath tumor that shows evidence of melanocytic differentiation. Melanin pigments are occasionally misdiagnosed as melanoma.[1] Approximately 40-50% of MS cases have psammoma bodies (scattered concentric microcalcifications), and this type is defined as psammomatous MS (PMS).[2,3] Half of all patients with PMS are known to have the Carney complex (CNC), a multiple neoplasia syndrome.[3] Unlike schwannomas, MSs have low malignant potential. The prognosis can be unfavorable in cases with local recurrence or malignant behavior, especially when multiple lesions are present and/or appear as part of CNC.[4,5]

We herein report a case of PMS without CNC.

CASE

A 26-year-old woman was admitted due to an abnormal shadow on chest X-ray. There was no remarkable medical or familial history other than anemia. Routine laboratory data were within normal limits. Plain computed tomography (Figure 1a) revealed a 2.5-cm mass in the right paraspinal area. We initially suspected a benign tumor, such as schwannoma, and performed video-assisted thoracoscopic surgery (VATS). On an examination, the tumor appeared to be a soft, black mass (Figure 1b) and it was located on the posterior spinal roots. However, an intraoperative histopathological examination suggested the possibility of a malignancy such as melanoma. We therefore performed open thoracotomy with additional resection of the intercostal artery, vein, nerve, and peripheral pleura.

A histopathological examination revealed that the margin was negative, and the tumor comprised epithelioid cells and occasional spindle cells arranged in whorl-like nests with abundant intracytoplasmic melanin pigment (Figure 2a). In addition, the mass contained psammoma bodies, which represent a form of concentric scattered calcification (Figure 2b). The histological and immunohistochemical findings of the tumor were compatible with those of PMS. The tumor cells were positive for S-100, HMB-45, and synaptophysin. The tumor showed no evidence of mitoses or cytological atypia. A thorough investigation revealed that the patient did not have any of the clinical manifestations of CNC or an af-

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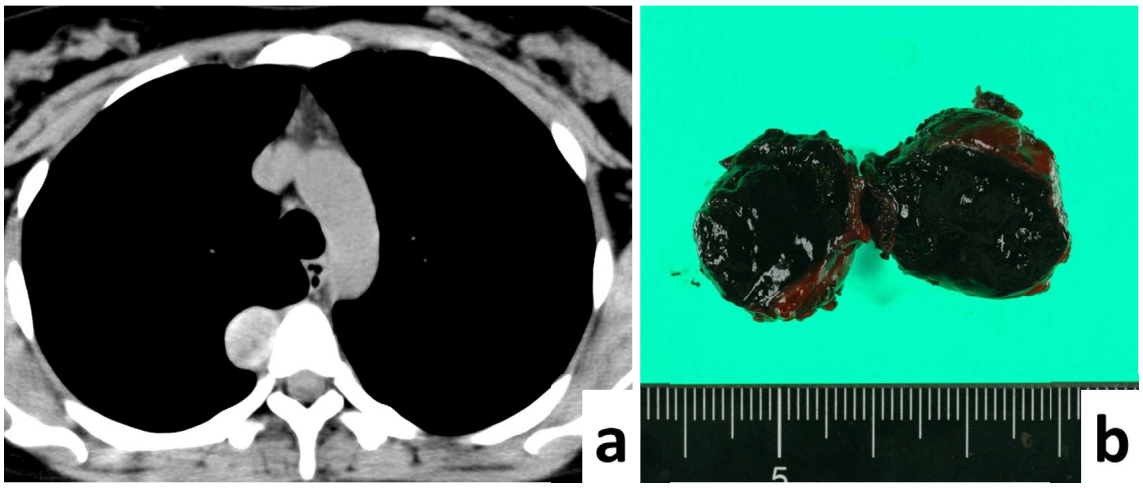

Figure 1. **a.** Chest computed tomography on admission showed a round mass (2.5 cm in size) in contact with the right thoracic vertebra. **b.** A gross examination of psammomatous melanotic schwannoma revealed an encapsulated black tumor with a smooth surface.

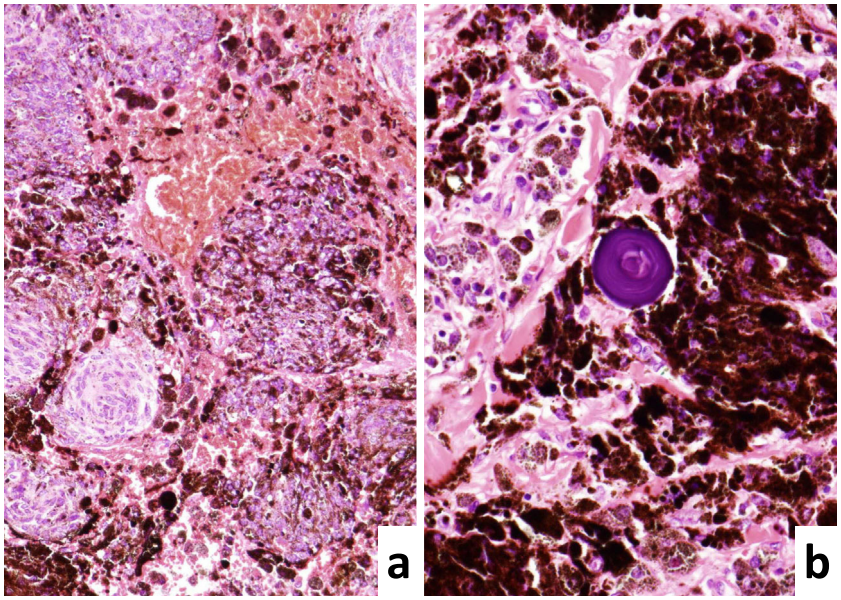

Figure 2. **a.** A histopathologic examination showed the lesion to be composed of epithelioid cells and occasional spindle cells arranged in whorl-like nests with abundant intracytoplasmic melanin pigment (hematoxylin and eosin stain, ×100). **b.** The characteristic feature of the tumor was the presence of psammomatous calcification (hematoxylin and eosin stain, ×400).

fected parent.

Ten years after curative resection, the patient remains alive without any evidence of recurrence.

DISCUSSION

MS is a rare variant of schwannoma that accounts for less than 1% of all nerve sheath tumors. It affects both males and females equally and generally occurs in young adults (mean age at the diagnosis: 38 years).[5] The tumor can be black, brown, blue, or purple in color and encapsulated, and it has a solid, cystic, or spongy appearance.[6] Generally, it is found in midline structures, such as the spinal roots or sympathetic ganglia in the chest and paraspinal area, but it is also occasionally

found in the gastrointestinal tract, heart, liver, bronchus, skin, soft tissues, or bone.[3,5] Both immunohistochemistry (e.g. HMB-45, melan A, vimentin, cytokeratin, Ki-67, laminin, and collagen IV) and morphology must be assessed in order to distinguish MS from malignant melanoma.[7] In addition, it is useful to test for BRAF V600E, which is typically present in most melanomas but is absent in MS.[8] In addition to malignant melanoma, pigmented meningioma, neurofibroma, rhabdomyosarcoma, clear-cell sarcoma of soft tissue, melanotic medulloblastoma, ganglioneuroblastoma, ectomesenchymoma (triton tumor), neurotrophic melanoma, and melanotic neuroendocrine carcinomas and carcinoids should also be distinguished from MS.[9] The present case was diagnosed as MS because of the morphologic features, absence of atypia, and immunohistochemical findings.

Total surgical excision of PMS is the most effective treatment. Data on the efficacy of radiotherapy or chemotherapy are not yet available.[2] Overall, 10-15% of MS cases are clinically malignant,[3] and 15-33% of these cases involve local recurrence or metastasis.[4,5] Although PMS is considered benign, it may recur several years after resection.[10] In the present case, we converted VATS to open thoracotomy to achieve total surgical excision of the tumor with tumor-free margins, which was critical given that the intraoperative rapid diagnosis indicated the possibility of malignancy. Benign posterior mediastinal tumors are ordinarily resected via VATS; however, tumors that extend into the intraspinal and mediastinal areas are contraindicated for VATS.[11]

PMS must be marked because more than half of patients with this tumor are known to have CNC, a multiple neoplasia syndrome characterized by spotty skin pigmentation (lentigines and blue nevi), myxomas (heart, skin, and breast), endocrine tumors (adrenal cortex, pituitary, testis, and thyroid), and schwannomas.[3,5,6] Approximately 20% of MSs are multiple and have an increased probability of associated CNC. CNC is an inherited autosomal dominant condition. Linkage analyses in CNC families have identified two main loci: 2p16 and 17q22-24. *PRKAR1A*, a gene encoding protein kinase A regulatory subunit 1A, was recently identified at 17q22-24 as a candidate gene for CNC.[12] CNC is a rare disease affecting approximately 1 in every 15,000 people, and it affects males and females equally. The median age at the diagnosis is 20 years. Approximately 70% of CNC cases are familial, whereas 30% are sporadic.[13] PMS occurs in an estimated 10% of affected individuals with CNC, and approximately 10% of schwannomas are associated with multisystem disorders, such as neurofibromatosis, schwannomatosis, multiple meningiomas, and CNC.[13] Among patients with CNC, 13% of those with PMS reportedly died as a result of the tumor.[6]

CONCLUSION

In summary, we presented a case of PMS not associated with CNC that exhibited a good clinical course after complete resection. Appropriate long-term follow-up is required for all cases of MS, given that it can recur or metastasize more than 5 years after resection, even in the absence of overt malignant histological features. The present case of PMS highlights the importance of being aware of the probability of associated CNC and differentiating the lesion from malignant melanoma, which significantly affects both the pre- and postoperative management.

本論文内容に関連する著者の利益相反：なし

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