CASE REPORT

Psammomatous melanotic schwannoma with cystic changes from old hemorrhages in the cervical spinal canal: a case report

Qing-hua Zhao MD, Shan Zhi MM, Zhen Wang MM, Ji-wei Tian MD

Department of Orthopedic Surgery, The First Affiliated Hospital of Shanghai Jiaotong University, Shanghai, China

Key words: Cervical vertebrae; Hemorrhage; Neurilemmoma

Introduction

Psammomatous melanotic schwannoma (PMS) represents a rare type of schwannoma; its occurrence in the spinal canal is even rarer¹. In the present study, we report a case of pathologically confirmed PMS with cystic changes from old hemorrhages in the cervical spinal canal. The patient underwent surgical resection of the tumor with postoperative radiotherapy, and had a smooth recovery. Follow-up X-rays and MRI at 16 months after the operation revealed no evidence of tumor recurrence.

Case report

A 46-year-old male patient presented on 18 August, 2007 complaining of pain and distension in his neck for 2 years and numbness of his left hand for 1 year; the symptoms had worsened 1 month before admission. Previous radiographic examination at a local hospital had suggested degenerative changes of the cervical vertebrae. Physical examination on admission revealed cervical curvature without obvious tenderness in the neck. The left hand numbness was especially obvious in the fourth and little fingers. No obvious sensory abnormalities were found in the right hand; neither muscular atrophy nor reduced muscle strength was detected in the upper limbs. The patient showed a positive Hoffmann reflex on the left and had normal bilateral patellar reflexes and Achilles tendon reflexes. The straight-leg raising test was negative, anal reflex positive and bilateral Babinski sign negative. Saddle area sensation was normal. No obvious abnormalities were found on routine blood or urine tests, or in chest X-ray films or ultrasonic cardiography.

Address for correspondence Ji-wei Tian, MD, Department of Orthopedic Surgery, The First Affiliated Hospital of Shanghai Jiaotong University, 100 Haining Road, Hongkou District, Shanghai, China 200080 Tel: 0086-21-37798566; Fax: 0086-21-63241377; Email: tjw609@163.com

Received: 30 September 2010; accepted 16 January 2011

DOI: 10.1111/j.1757-7861.2011.00133.x

Three-dimensional reconstruction of the spinal vertebrae revealed defects in the left intervertebral foramina at the C6 and C7 levels, the edges of the foramina had become rounded and blunt (Fig. 1). Cervical MRI displayed a $0.6 \times 1.1 \times 0.8$ cm mass in the spinal canal at the C7 level. Coronal images showed that the mass was located to the left of the spinal cord outside the dura mater, and was protruding through to the intervertebral foramina at the C6 and C7 levels, resulting in a dumbbell-shaped appearance. The tumor mass outside the spinal canal measured $1.7 \times 2.1 \times 1.5$ cm and was causing a right shift of the spinal cord and dura mater. On T1-weighted images (T1WI) the tumor showed hyperintense signals, with hypointense signals on T2WI with scattered hyperintense spots.

Surgical resection of the tumor was then scheduled. During surgical exploration, a soft dark blue tumor mass with hemorrhagic foci was identified outside the dura mater (Fig. 2). Macroscopically, the tumor showed clear boundaries and had a diameter of 1.5 cm. The cross section of the tumor was black and hard, and showed infiltration of some small compartments which were aligned in a fence-like or spiral fashion. Microscopy showed spindle-shaped tumor cells aligned in intersecting strands. Very little nuclear polymorphism was detected in the tumor cells, which exhibited normal mitosis and contained large quantities of melanin granules with occasional psammoma bodies and hemorrhages (Fig. 3). Immunohistochemistry showed S-100- and HMB-45-positive foci in the tumor. The tumor cells were positive for vimentin, and negative for creatine kinase, epithelial membrane antigen, glial fibrillary acidic protein, coagulation factor VIII, cluster of differentiation 34, smooth muscle actin, Syn, and neuron-specific enolase. A pathological diagnosis of PMS with cystic changes from old hemorrhages was established. The patient received radiotherapy to the total dose of 55 Gy after surgical tumor removal. Follow-up X-ray and MRI performed 16 months after the operation showed optimal position of the

Figure 1 Three-dimensional reconstruction of the cervical vertebrae showing defects of the intervertebral foramina on the left at C6 and C7 levels, the foramina having rounded edges. The arrowhead indicates the location and shape of the mass.

internal fixation without signs of tumor recurrence (Fig. 4).

Discussion

Psammomatous melanotic schwannoma occurs mostly in young patients of either sex, the median age at onset being 38 years. We compared and contrasted our case with

Figure 2 Intraoperative finding of the tumor outside the dura mater, it is soft, has a dark blue appearance and contains hemorrhagic foci.

Figure 3 Tracts of the proliferating spindle-shaped tumor cells containing melanin granules and psammoma bodies (hematoxylin and eosin staining, original magnification: $\times 100$).

several previous published cases of PMS (Table 1). The commonest locations of PMS are the nerve roots and the sympathetic ganglia, and the initial symptom is typically pain in the involved area. The tumor generally has a clear boundary without a capsule, hemorrhage, necrosis, cystic changes and melanin granules of varying quantities are often present. Besides melanin granules, the tumor cells often contain calcified psammoma bodies and express the

Figure 4 Lateral X-ray film performed 16 months after the operation showing optimal position of the internal fixation without evidence of tumor recurrence in the cervical spine.

Table 1 Comparison with previously reported cases of PMS

Case No.	Age	Sex	Location	Pathology	Recurrent	Carney Syndrome
1*	46	M	C7 level	Spindle-shaped tumor cells aligned in intersecting strands; little nuclear polymorphism; normal mitosis and containing large quantities of melanin granules with occasional psammoma bodies and hemorrhages	N	N
2 ⁴	53	F	C6 nerve root, primarily intradural and growing through the enlarged C5–6 neural foramen to focally infiltrate the dura mater.	Melanin-pigmented tumor with spindle-shaped and epithelioid cells and scattered psammoma bodies	N	Unclear
3 ⁵	53	M	T9–T10, extending into the right intrathoracic space (2.5 cm)	Melanotic schwannoma with mitotic index not more than one in ten fields on high magnification, indicating low risk of malignancy	N	N
4 ⁶	56	F	Spine	Composed of polygonal epithelioid or spindle-shaped cells, some of which contained melanin. Small calcifications were dispersed throughout the lesion.	N	—
5 ⁶	58	M	Spine	Composed of polygonal epithelioid or spindle-shaped cells, some of which contained melanin. Small calcifications were dispersed throughout the lesion.	Recurred locally	—
6 ⁷	36	M	Left C4 spinal root.	Most tumor cells were spindle-shaped. A few cells were large and epithelioid-like, with a prominent nucleolus. Mitoses were rarely seen. Many tumor cells contained coarse or fine granules of a brown pigment.	—	N
7 ⁸	32	M	Well-circumscribed, within the sacral canal	Heavily pigmented, consisted of numerous aggregates of coarse brown granules. Several purple-staining psammoma bodies; considerable necrosis, mild nuclear atypia.	N	Y

*, current case.

tumor cell markers S-100 and HMB-45. PMS with concurrent myxoma, skin pigmentation, and endocrine disorder (as in Cushing's syndrome) is known as the Carney complex². Research has suggested that about 55% of people with tumors containing psammoma bodies have the Carney complex³. Patients with PMS and concurrent Carney complex have a relatively young age of disease onset (around 23 years), and often present with multiple tumor foci. In these patients, the melanocytes are mostly benign, but occasionally may produce tissue invasion and metastasis. Therefore a definite diagnosis of PMS has to depend on a comprehensive evaluation of the clinical characteristics, histological and morphological findings of the tumor as well as the results of immunohistochemistry.

Histologically, PMS differs from common schwannoma in that PMS contains psammoma bodies, melanin and adipose tissue⁹. In general, melanotic schwannoma has a relatively long latency period before relapse (4–5 years), whereas malignant melanotic schwannoma is characterized by rapid recurrence and metastasis. Melanotic schwannoma involving the central nervous system and the

head/face is often associated with a poor prognosis, therefore long-term follow-up of these patients is essential¹⁰.

Psammomatous melanotic schwannoma may display characteristic signal patterns on MRI which assist clinical diagnosis when present. The melanin granules in PMS are responsible for the hyperintense signals on T1WI and isointense signals on T2WI. In our case, both surgical exploration and pathological examination detected hemorrhagic foci in the tumor; these were the cause of the hyperintense spots on T2WI. A hematoma in the sub-acute stage may display hyperintense signals on T1WI and hypointense signals on T2WI without enhancement of the hematoma, whereas an old hematoma presents hyperintense signals on both T1WI and T2WI. The differential diagnosis of PMS includes various benign or malignant pigmented lesions such as metastatic malignant melanoma without an identifiable primary focus, cell-rich blue nevus, melanotic neuroectodermal tumor of infancy, and pigmented neurofibroma. PMS in the central nervous system should be differentiated from

melanocytic meningioma, melanotic ependymoma, and melanotic medulloblastoma⁷.

Radical surgical resection is the primary treatment of choice for PMS. PMS involving a single vertebra can be easily removed, while PMS compromising multiple vertebrae can have extensive involvement with obscure boundaries and potential risk of massive bleeding, rendering total resection difficult. In the latter case, reduction of the tumor volume by partial resection is recommended before making an attempt to separate the tumor tissue from neural tissues. PMS is likely to recur postoperatively, and staged surgeries can be considered when necessary.

Disclosure

The manuscript submitted does not contain information about medical device(s)/drug(s). No benefits in any form have been, or will be, received from a commercial party related directly or indirectly to the subject of this manuscript.

References

1. Brat DJ, Giannini C, Scheithauer BW, *et al.* Primary melanocytic neoplasms of the central nervous systems. *Am J Surg Pathol*, 1999, 23: 745–754.
2. Carney JA. Psammomatous melanotic schwannoma. A distinctive, heritable tumor with special associations, including cardiac myxoma and Cushing's syndrome. *Am J Surg Pathol*, 1990, 14: 206–222.
3. Carney JA. The Carney complex (myxomas, spotty pigmentation, endocrine overactivity, and schwannomas). *Dermatol Clin*, 1995, 13: 19–26.
4. Kuchelmeister K, Lotz C, Schönmayr R, *et al.* April 2004: woman in her early fifties with a cervical intraspinal and extraspinal mass lesion. *Brain Pathol*, 2004, 14: 453–454, 458–459.
5. De Cerchio L, Contratti F, Fraioli MF. Dorsal dumb-bell melanotic schwannoma operated on by posterior and anterior approach: case report and a review of the literature. *Eur Spine J*, 2006, (Suppl.)5: S664–S669.
6. Ludvíková M, Michal M, Marek J, *et al.* Psammomatous melanotic schwannoma. *Cesk Patol*, 1997, 33: 141–145.
7. Léger F, Vital C, Rivel J, *et al.* Psammomatous melanotic schwannoma of a spinal nerve root. Relationship with the Carney complex. *Pathol Res Pract*, 1996, 192: 1142–1146.
8. Martin-Reay DG, Shattuck MC, Guthrie FW Jr. Psammomatous melanotic schwannoma: an additional component of Carney's complex. Report of a case. *Am J Clin Pathol*, 1991, 95: 484–489.
9. Dodd LG, Marom EM, Dash RC, Matthews MR, McLendon RE. Fine-needle aspiration cytology of "ancient" schwannoma. *Diagn Cytopathol*, 1999, 20: 307–311.
10. Prieto-Rodríguez M, Camañas-Sanz A, Bas T, *et al.* Psammomatous melanotic schwannoma localized in the mediastinum: diagnosis by fine-needle aspiration cytology. *Diagn Cytopathol*, 1998, 19: 298–302.