SPANDIDOS PUBLICATIONS

MOLECULAR AND CLINICAL ONCOLOGY 2: 156-158, 2014

Neurothekeomas of the thoracic and lumbar area in an adult man: A case report

JUN LI[1], ZHANJUN SHI[2] and JUEHUA JING[1]

[1]Department of Orthopaedics, The Second Hospital of Anhui Medical University, Hefei, Anhui 230601;
[2]Department of Orthopaedics, Nanfang Hospital, Southern Medical University, Guangzhou, Guangdong, P.R. China

Received August 8, 2013; Accepted October 17, 2013

DOI: 10.3892/mco.2013.211

Abstract. Neurothekeomas are unusual benign soft tissue tumors of the peripheral nervous system, which commonly occur on the head, neck and upper extremities of young females in the second and third decades of life. This is the first case report of neurothekeoma developing simultaneously in the thoracic and lumbar area in a 51-year-old male. The two tumors were completely excised along the capsule under local anaesthesia and the incisions were closed in layers following adequate hemostasis. Histopathologically, the examination of the microscopic slides from the thoracic and lumbar masses revealed the presence of spindle and similar epithelioid cells arranged in fascicles, often in a geographical pattern, in a background of myxoid stroma. The immunohistochemical analysis demonstrated that the tumor cells were positive for vimentin, CD57 and actin and negative for S-100 protein, HMB-45 and CD34. The tumors were diagnosed as neurothekeomas. The patient remains alive and well on follow-up at 7 years, without evidence of recurrence.

Introduction

Neurothekeomas, or nerve sheath myxomas, are benign peripheral nerve sheath-derived neoplasms that are continuously subjected to conceptual changes in classification. The origin of these tumors has not been elucidated and their etiology has been debated upon (1,2). Their reproducible clinical, histopathological and immunohistochemical differences were demonstrated in a large series of studies (2-4). Clinically, neurothekeoma is a slowly growing, usually asymptomatic, dermal or, less frequently, mucosal or submucosal tumor, occurring 2-4 times more frequently in women compared to men (5,6). Neurothekeoma commonly develops on the head, neck and upper extremities of young females in the second and third decades of life. Other studies have documented different locations, including the tongue, eyelids and oral mucosa (4,7,8). To the best of our knowledge, this is the first case report of neurothekeoma developing simultaneously in the thoracic and lumbar area in an adult male.

Correspondence to: Dr Juehua Jing, Department of Orthopaedics, The Second Hospital of Anhui Medical University, 678 Furong Avenue, Hefei, Anhui 230601, P.R. China
E-mail: jingjuehuapaper@163.com

Key words: neurothekeoma, thoracic, lumbar

Case report

A 51-year-old Chinese male patient presented with a 5-year history of a right lumbar mass and 1-year history of a right thoracic wall mass. The right lumbar mass enlarged slowly, unlike the right thoracic wall mass, which enlarged quickly, particularly over the last 6 months. There was no reported pain, fever, palpitations, irritability, dysphagia, dyspnea, weight loss, or other significant medical of familial history. The patient did not have a history of smoking and did not report any history of trauma. Examination of the lesions revealed a solid mass in the right thoracic wall and an elastic mass in the right lumbar area, which were painless, non-tender, elliptical, firm, without local erythema or edema of the overlying skin. Physical examination did not reveal signs of lymphadenopathy or hepatosplenomegaly. Neurological examination was normal.

Ultrasonography of the thoracic and lumbar masses showed a solid echo group, with clear boundaries and heterogeneous internal echo (Fig. 1). Color doppler flow imaging demonstrated a limited number of blood flow signals in the mass. The thoracic X-rays, electrocardiogram and serum electrolyte levels were normal.

The two masses were completely excised along the capsule under local anaesthesia and the incisions were closed in layers following adequate hemostasis. The surgical time was 2 h, with an intraoperative blood loss of 30 ml. The dimensions of the thoracic mass were ~7.0x7.0x3.0 cm and those of the lumbar mass ~4.0x7.0x3.0 cm. The tumors were encapsulated, with a solid and gelatinous sectional plane.

Histopathologically, the examination of the microscopic slides of the two tumors revealed spindle and similar epithelioid cells, arranged in fascicles, often in a geographical pattern, in a background of myxoid stroma (Fig. 2). The immunohistochemical analysis demonstrated that the tumor cells were positive for vimentin, CD57 and actin and negative for S-100 protein, HMB-45 and CD34. The tumors were

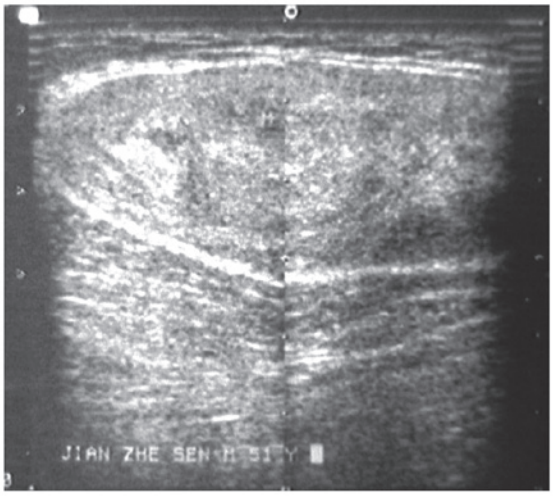

Figure 1. Ultrasound demonstrated regular solid group, clear boundaries and heterogeneous internal echo.

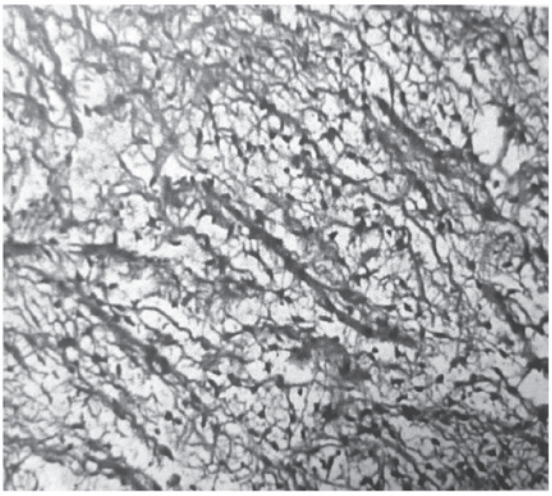

Figure 2. The mass contained spindle and similar epithelioid cells, arranged in fascicles, often in a geographical pattern, in a background of myxoid stroma (hematoxylin and eosin staining, magnification, x200).

diagnosed as neurothekeomas. The patient remains alive at the 7 year follow-up, without evidence of recurrence.

Discussion

Neurothekeoma was first described in 1969 under the term 'nerve sheath myxoma'; the name 'neurothekeoma' was adopted by Gallager and Helwig (9) in 1980. Neurothekeoma is an unusual benign soft tissue tumor of the peripheral nervous system, which commonly occurs on the head, neck and upper extremities of young females in the second and third decades of life (10-12). To the best of our knowledge, this is the first case report of neurothekeoma developing simultaneously in the thoracic and lumbar area in an adult male.

Historically, neurothekeoma has been subclassified as myxoid, cellular or mixed type, based on the amount of the myxoid component; in our case, the neurothekeomas were of the cellular subtype (13). Typically, cellular neurothekeomas display a characteristic fascicular pattern of spindle and epithelioid cells, with variable cytologic atypia and mitotic activity (14). Immunostaining demonstrated that the cells were negative for S-100 protein and HMB-45 and positive for vimentin, neuron-specific enolase and NKI/C3 (14).

Neurothekeomas are difficult to diagnose prior to performing a biopsy, due to the lack of specific clinical manifestations or imaging characteristics. The diagnosis of the thoracic mass was oriented towards lipoma prior to surgical excision. Clinically, neurothekeoma is a commonly asymptomatic, dermal, mucosal or submucosal tumor. On computed tomography scans, it is identified as a hypoattenuated-to-isoattenuated mass, with variable enhancement and vascularity patterns. On magnetic resonance imaging scans, it exhibits well-defined margins, intermediate signal on T1-weighted images, high signal on T2-weighted images and heterogeneous mild-to-moderate gadolinium contrast enhancement (15). The differential diagnosis of neurothekeoma should include other neural entities, such as schwannoma, true neuroma and myxoid neurofibroma. The effective treatment of choice is complete surgical excision with clear margins. No malignant transformation or metastasis have been reported and local recurrence is extremely uncommon when there are clear surgical margins.

In conclusion, neurothekeomas are uncommon, usually small tumours, which are treated by simple excision. To the best of our knowledge, this is the first case report of neurothekeomas developing simultaneously in the thoracic and lumbar area in an adult male. Neurothekeomas are difficult to diagnose prior to biopsy. Although this is a rare type of tumor, the clinician should consider this entity in differential diagnosis, as it is imperative to distinguish it from malignant lesions, in order to avoid unnecessary aggressive treatment.

Acknowledgements

This study was supported by the Anhui Provincial Natural Science Foundation (1308085MH156).

References

1. Barnhill RL and Mihm MC Jr: Cellular neurothekeoma. A distinctive variant of neurothekeoma mimicking nevomelanocytic tumors. Am J Surg Pathol 14: 113-120, 1990.
2. Hornick JL and Fletcher CD: Cellular neurothekeoma: detailed characterization in a series of 133 cases. Am J Surg Pathol 31: 329-340, 2007.
3. Fetsch JF, Laskin WB and Miettinen M: Nerve sheath myxoma: a clinicopathologic and immunohistochemical analysis of 57 morphologically distinctive, S-100 protein- and GFAP-positive, myxoid peripheral nerve sheath tumors with a predilection for the extremities and a high local recurrence rate. Am J Surg Pathol 29: 1615-1624, 2005.
4. Papalas JA, Proia AD, Hitchcock M, Gandhi P and Cummings TJ: Neurothekeoma palpebrae: a report of 3 cases. Am J Dermatopathol 32: 374-379, 2010.
5. Papadopoulos EJ, Cohen PR and Hebert AA: Neurothekeoma: report of a case in an infant and review of the literature. J Am Acad Dermatol 50: 129-134, 2004.
6. Marocchio LS, Oliveira DT and Consolaro A: Myxoid neurothekeoma of the oral mucosa: an unusual benign tumor. Oral Dis 10: 408-409, 2004.
7. Peñarrocha M, Bonet J, Minguez JM and Vera F: Nerve sheath myxoma (neurothekeoma) in the tongue of a newborn. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 90: 74-77, 2000.
8. Pepine M, Flowers F and Ramos-Caro FA: Neurothekeoma in a 15-year-old boy: case report. Pediatr Dermatol 9: 272-274, 1992.

9. Gallager RL and Helwig EB: Neurothekeoma - a benign cutaneous tumor of neural origin. Am J Clin Pathol 74: 759-764, 1980.
10. Roholt NS, Guitart J and Eramo LR: A gradually enlarging asymptomatic nasal mass. Pediatr Dermatol 12: 191-194, 1995.
11. Busam KJ, Mentzel T, Colpaert C, Barnhill RL and Fletcher CD: Atypical or worrisome features in cellular neurothekeoma: a study of 10 cases. Am J Surg Pathol 22: 1067-1072, 1998.
12. Strumia R, Lombardi AR and Cavazzini L: Cellular neurothekeoma. Acta Derm Venereol 79: 162-163, 1999.
13. Argenyi ZB, LeBoit PE, Santa Cruz D, Swanson PE and Kutzner H: Nerve sheath myxoma (neurothekeoma) of the skin: light microscopic and immunohistochemical reappraisal of the cellular variant. J Cutan Pathol 20: 294-303, 1993.
14. Barnhill RL: Nerve sheath myxoma (neurothekeoma). J Cutan Pathol 21: 91-93, 1994.
15. O'Rourke H, Meyers SP and Katzman PJ: Neurothekeoma in the upper extremity: magnetic resonance imaging and computed tomography findings. J Comput Assist Tomogr 29: 847-850, 2005.