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

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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.

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 or 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

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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.

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