<table>
<thead>
<tr>
<th>Title</th>
<th>Cellular Neurothekeoma A with an Immunohistochemical Study</th>
</tr>
</thead>
<tbody>
<tr>
<td>Author(s)</td>
<td>Wen, Chunyang; Ito, Masahiro; Chen, Long-Dian; Nakayama, Toshiyuki; Nakashima, Masahiro; Tsuneda, Katsumi; Sekine, Ichiro</td>
</tr>
<tr>
<td>Citation</td>
<td>Acta medica Nagasakiensia. 2002, 47(1-2), p.71-73</td>
</tr>
<tr>
<td>Issue Date</td>
<td>2002-06-18</td>
</tr>
<tr>
<td>URL</td>
<td><a href="http://hdl.handle.net/10069/16213">http://hdl.handle.net/10069/16213</a></td>
</tr>
</tbody>
</table>

NAOSITE: Nagasaki University’s Academic Output SITE

http://naosite.lb.nagasaki-u.ac.jp
Cellular Neurothekeoma

A Case Report with an Immunohistochemical Study

Chun Yang Wen¹,², Masahiro Ito³, Long-Dian Chen², Toshiyuki Nakayama⁴, Masahiro Nakashima⁴, Katsumi Tsuneda⁵, Ichiro Sekine¹

¹) Department of Molecular Pathology, Atomic Bomb Disease Institute, Nagasaki University School of Medicine
²) Department of Digestive Disease, Nanjing Gu-Lou Hospital, Medical School of Nanjing University
³) Department of Pathology, National Nagasaki Medical Center
⁴) Tissue and Histopathology Section Division of Scientific Data Registry, Atomic Bomb Disease Institute, Nagasaki University School of Medicine
⁵) Tsuneda Medical Laser Institute and Plastic Surgery Clinic

Cellular neurothekeoma preferentially affects women and has a predilection for the head, neck and arms of children and young adults. Cellular neurothekeoma is a well-circumscribed tumor involving the reticular dermis and consisting of fascicles of polygonal or spindle cells with eosinophilic cytoplasm and neuroid characteristics. We describe an additional case of cellular neurothekeoma on the upper lip. Microscopically, the neoplasm had a multinodular or lobulated architecture that were partially or completely invested by a dense band of collagen and was composed of plump, spindled, and epithelioid cells arranged in solid, infiltrating nests and fascicles. Cytologically, the neoplastic cells were relatively large and contained lightly eosinophilic, finely vacuolated cytoplasm. The nuclei were round to oval with vesicular chromatin, with no atypical mitotic figures identified. Immunohistochemical examination showed that the neoplastic cells were diffusely positive for vimentin and positive for NSE, but were negative for S-100 protein. Our case was a representative example of a typical cellular neurothekeoma.

Key Words: neurothekeoma, cellular neurothekeoma, Vimentin, S-100 protein

ACTA MEDICA NAGASAKIENSIA 47 : 71−73, 2002

Case Report

Introduction

Neurothekeomas are benign neoplasms with probable neural differentiation. They were described originally in 1969 by Harkin and Reed⁷ who used the name nerve sheath myxoma. The term neurothekeoma was coined by Gallager and Helwig⁶ in 1980 based on their 53 patients. Names including pacinian neurofibroma, bizarre cutaneous neurofibroma, cutaneous lobular neuromyxoma, and perineurial myxoma have been used by other authors⁶, 7, 8. A similar lesion consisting predominantly of epithelioid cells with minimal mucin was termed cellular neurothekeoma by Rosati et al.⁷ in 1986. Several years later, Barnhill and Mihm⁸ described five additional cases of cellular neurothekeoma. In this report, we describe an additional case of cellular neurothekeoma on the upper lip.

Case report

The patient was a 59-year-old woman presenting with an asymptomatic nodule on the upper lip, which had been present for several years. The tumor had been gradually increasing in size. Clinically, the lesion was firm in consistency, measuring 0.7x0.7 cm in diameter, and was covered by normal-appearing skin. The clinical diagnosis considered was histiocytoma. The lesion was completely removed and submitted for microscopic examination. The specimens were fixed in 10% buffered formalin and embedded in paraffin. Sections of 4 μm thick were stained with hematoxylin and eosin, and alcian blue stain. Immunohistochemical staining was
performed using the avidin-biotin-peroxidase complex (ABC) method and antibodies for vimentin, neuron-specific enolase (NSE), S-100 protein, glial fibrillary acidic protein (GFAP), Leu 7 (CD57), neurofilament protein, epithelial membrane antigen (EMA), cytokeratin, smooth muscle actin (SM-actin), and desmin.

The excisional specimen revealed a rather well circumscribed tumor in the dermis. The cut surface was nodular, yellowish white. Microscopically, the neoplasm had a multinodular or lobulated architecture that were partially or completely invested by a dense band of collagen and was composed of plump, spindled, and epithelioid cells arranged in solid, infiltrating nests and fascicles (Fig 1). Cytologically, the neoplastic cells were relatively large and contained lightly eosinophilic, finely vacuolated cytoplasm. The nuclei were round to oval with vesicular chromatin, with no atypical mitotic figures identified (Fig 2). The stroma consisted of wispy collagen fibers that tended to condense in the more epithelioid areas, scattered capillary-sized vessels, and stromal mucosubstance. Focal myxomatous change, demonstrated by alcian blue staining, was present.

Immunohistochemical examination showed that the neoplastic cells were diffusely positive for vimentin (Fig 3) and positive for NSE (Fig 4), but were negative for S-100 protein. The immunohistochemical results for this case are given in Table 1.

Figure 1. Microscopically, the neoplasm had a multinodular or lobulated architecture that were partially or completely invested by a dense band of collagen and was composed of plump, spindled, and epithelioid cells arranged in solid, infiltrating nests and fascicles. Hematoxylin & eosin, x40

Figure 2. Cytologically, the neoplastic cells were relatively large and contained lightly eosinophilic, finely vacuolated cytoplasm. The nuclei were round to oval with vesicular chromatin, with no atypical mitotic figures identified. Hematoxylin & eosin, x200

Figure 3. Immunohistochemical examination showed that the neoplastic cells were diffusely positive for vimentin. Vimentin, x200

Figure 4. Immunohistochemical examination showed that the neoplastic cells were positive for NSE. NSE, x200
that tumor cells of cellular neurothekeoma were positive for vimentin and negative for S-100 protein\(^\text{1b}\), while the myxoid type of neurothekeoma shows consistent S-100 protein immunoreactivity\(^\text{1b}\), supporting a Schwannian differentiation. The cell of origin of the cellular type of neurothekeoma is less clear due to the lack of S-100 protein immunoreactivity. Our findings support the observation that cellular neurothekeoma differs from the myxoid type of neurothekeoma.

In a study of cellular neurothekeoma, Zelger et al\(^\text{14}\) noted additional similarities between cellular neurothekeoma and fibrous histiocytoma, including immunohistochemical expression of SM-actin. In our case, however, SM-actin and CD68 were not expressed. Although Laskin et al\(^\text{15}\) reported that cellular and mixed neurothekeoma variants lack neurosustentacular differentiation and may be potentially related to tumors included in the fibrohistiocytic category, their true origin is unclear.

**References**