Inflammatory pseudotumor of the paratesticular area is a rare entity, composed of spindle cells admixed with variable amounts of extracellular collagen, lymphocytes, and plasma cells. In the genitourinary tract, inflammatory pseudotumor most commonly occurs in the urinary bladder.\textsuperscript{1,2} Fibrous or inflammatory pseudotumors are within the spectrum of benign paratesticular lesions, a category which includes distinct inflammatory myofibroblastic tumors and calcifying fibrous tumor. In Korea, five such cases have been reported.\textsuperscript{3-5} Here, we report one additional case, showing typical clinicopathologic features of a paratesticular inflammatory pseudotumor, and we briefly review the previously reported cases.

**CASE REPORT**

A 34-year-old man, who had had a palpable scrotal mass for 10 years, visited our hospital, due to a recently enlarged non-tender scrotal lesion. Mass excision, including the epididymis, was done. After the operation, no additional chemotherapy or radiation was performed. During four months of follow-up, he had no recurrence. Grossly, the excised mass had a multinodular appearance (Fig. 1), and the cut surface was homogeneous brownish-tan. The texture was rubbery. Microscopically, the mass predominantly exhibited sclerosis and fibrosis, composed of hypocellular fibrous tissue with dense “keloid-like” fibrosis and sparse inflammatory cells (Fig. 2A). In a small focus of the slightly cellular portion, bland-looking cigar-shaped spindle cells were loosely arranged in a dense hyaline stroma (Fig. 2B). They had vesicular nuclei and no mitosis. The infiltrated cell population was composed of mature small lymphocytes, and plasma cells with some eosinophils. Serial sections revealed no necrosis or hemorrhages. The moderately cellular portion was rich in blood vessels, by which the endothelial and muscle walls were thickened. Most of them showed luminal obliteration (Fig. 2C). True vasculitis was not found. Immunohistochemistry was done by avidin-biotin-peroxidase complex method, using antibodies against antigens. This is shown in Table 1. Immunohistochemically, the spindle cells expressed vimentin (VIM3B4; 1:80, Dako, Glostrup, Denmark), focally smooth muscle actin (IA4; Zymed, San Francisco, CA, USA,
1:100 dilution, Fig. 3A) and CD68 (Zymed, 1:50 dilution), but not desmin (D33; Zymed, 1:50 dilution), cytokeratin (AE1/AE3; Zyomed, 1:50 dilution), p53 (BP53.12; Zyomed, 1:250 dilution), Ki-67 (polyclonal, Dako, 1:250 dilution) or ALK-1 (p80; DAKO, 1:40 dilution). The histological and immunohistochemical results are summarized in Table 1. Electron microscopy revealed abundant extracellular collagen fibrils, with rarely-found spindle cells (Fig. 3). The spindle cells had thin peripherally-located myofilaments, and abundant rough endoplasmic reticula with focal densities and were found to be mixed with inflammatory cells including plasma cells and mast cells.

**DISCUSSION**

Variable terms such as inflammatory pseudotumor, inflammatory myofibroblastic tumor, proliferative funiculitis (pseudo-sarcomatous myofibroblastic proliferation of spermatic cord) and fibrous pseudotumor (peri-orchitis) have been used to describe lesions like the reported lesion. Currently, the term inflammatory myofibroblastic tumor has been generally accepted, encompassing all such lesions. This is essentially a tumor which exhibits cellular, fascicular fibroblastic/myofibroblastic proliferations, accompanied by a prominent infiltrate of chronic inflammatory cells, particularly plasma cells and mast cells. The spindle cell component typically has plump, variably atypical nuclei. The mitotic rate is variable. Immunohistochemically, histiocytes or bizarre spindle cells are generally smooth muscle actin-positive, and overexpress ALK-1 protein (p80), which stains positively in 40% of inflammatory myofibroblastic tumors.10

The present case is considered to be located at the extreme pole of more cellular inflammatory pseudotumors, i.e. typical histology of inflammatory myofibroblastic tumor. Because the present case showed sparse cellularity, we think that more cellular cases of inflammatory myofibroblastic tumors should be distinguished from inflammatory leiomyosarcoma, and even malignant lymphoma. Cigar-shaped, centrally-located blunt nuclei are regarded as a characteristic of leiomyosarcoma, but this is frequently obscured by infiltrating inflammatory cells, especially in inflammatory leiomyosarcoma, which leads to the misdiagnosis of inflammatory pseudotumors, as the above malignant tumors. In such circumstances, immunohistochemistry is often helpful in ascertaining the nature of the spindle cells.

**Table 1. Review of the previously reported Korean cases including the present case**

<table>
<thead>
<tr>
<th>Author &amp; year</th>
<th>Cases</th>
<th>Age (yr)</th>
<th>Presenting symptom</th>
<th>Symptom duration</th>
<th>Location</th>
<th>Gross finding</th>
<th>Calcification on H-E</th>
<th>Immunohistochemistry</th>
<th>Electron microscopy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yim, et al. (1994)</td>
<td>1</td>
<td>59</td>
<td>Slow growing, nontender mass</td>
<td>Incidentally found mass</td>
<td>Testicular tunics, proximal spermatic cord</td>
<td>Two separate nodules</td>
<td>Absent</td>
<td>+: vimentin, actin</td>
<td>Not done</td>
</tr>
<tr>
<td>Paik, et al. (1995)</td>
<td>2</td>
<td>28</td>
<td>Slow growing, nontender mass</td>
<td>6 months</td>
<td>Testicular tunics</td>
<td>Multinodular appearance</td>
<td>Absent</td>
<td>+: vimentin, actin, S-100 protein (weak)</td>
<td>Not done</td>
</tr>
<tr>
<td></td>
<td>3</td>
<td>30</td>
<td>Slow growing, nontender mass</td>
<td>5-6 yr</td>
<td>Testicular tunics</td>
<td>Multinodular appearance</td>
<td>Present</td>
<td>+: vimentin, actin</td>
<td>Not done</td>
</tr>
<tr>
<td>Yoo, et al. (2000)</td>
<td>4</td>
<td>52</td>
<td>Slow growing, nontender mass</td>
<td>10 yr</td>
<td>Testicular tunics</td>
<td>Multinodular appearance</td>
<td>Absent</td>
<td>+: vimentin -: actin, desmin, S-100 protein</td>
<td>Not done</td>
</tr>
<tr>
<td></td>
<td>5</td>
<td>38</td>
<td>Slow growing, nontender mass</td>
<td>14 months</td>
<td>Testicular tunics</td>
<td>Multinodular appearance</td>
<td>Absent</td>
<td>+: vimentin -: actin, desmin, S-100 protein</td>
<td>Not done</td>
</tr>
<tr>
<td>Kim, et al.* (2004)</td>
<td>6</td>
<td>32</td>
<td>Slow growing, nontender mass</td>
<td>10 yr</td>
<td>Periepididymal area</td>
<td>Multinodular appearance</td>
<td>Absent</td>
<td>+: vimentin, actin, desmin, S-100</td>
<td>Fibroblasts (stage III), abundant Collagen fibrils</td>
</tr>
</tbody>
</table>

yr, years; +, positive immunoreactivity; -, negative immunoreactivity; H-E, hematoxylin and eosin stain; *, the present case.
for correct diagnosis. In most inflammatory leiomyosarcomas, the muscle markers such as smooth muscle actin or desmin, are positive in the fascicles, whereas ALK-1 (p80) does not stain in the spindle cells. However, the present case differs in that the spindle cells in the sparsely cellular area showed immunonegativity for smooth muscle actin and ALK-1, but minimal desmin immunoreactivity. Ultrastructurally, the presence of the fibronexus junction is critical for diagnosing myofibroblastic differentiation in inflammatory myofibroblastic tumor including fibrous pseudotumors. The fibronexus junction is a cell-to-matrix specialization composed of myofilaments, and fibronectin fibrils. It is a shelf-like region of the cell surface, where convergence of intracellular smooth muscle myofilaments and extracellular fibronectin filaments are present. It is regarded as an important, but not entirely specific, ultrastructural marker for fully differentiated stage 4 myofibroblasts. The present case showed abundant rough endoplasmic reticulum, and peripheral myofilaments, indicating stage 3 fibroblasts (“myoid” fibroblasts). This observation supports the conclusion that the extremely fibrous inflammatory pseudotumor is located at the extreme pole of inflammatory myofibroblastic tumors.

After reviewing the previous reported Korean cases of parates-

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**Fig. 2.** (A) Low-power view shows loosely organized spindle cells admixed with chronic inflammatory cells and lymphoid follicle formation. (B) Spindle-shaped cells arranged in a storiform pattern in dense fibrous stroma. (C) Characteristically, thickened vascular walls and luminal obliteration are found.
paratesticular inflammatory pseudotumors, all the cases typically presented with a slowly growing, non-tender lesion, occurring first in middle to late adulthood. The mean age was 40 years, and the mean duration of the symptomatic period was 4.6 years. All the cases yielded good outcomes without recurrences. These clinicopathologic findings are summarized in Table 1.

In summary, the authors emphasize slowly growing inflammatory pseudotumor in the paratesticular area, which seems to be located at the extreme pole of the more cellular type of inflammatory myofibroblastic tumors.

REFERENCES