Calcifying Fibrous Pseudotumor of the Neck

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Calcifying fibrous pseudotumor is a rare lesion of uncertain histogenesis that has a unique histologic appearance. We report herein a case of a 24-year-old woman with a mass on the right posterior side of the neck. Magnetic resonance imaging with contrast showed a well-circumscribed mass between the right splenius and semispinalis cervicis muscles; the study suggested high collagen content. Simple excision was performed. The histologic findings were diagnostic of calcifying fibrous pseudotumor. Our review of 19 previously reported cases suggests that a good outcome is expected when a diagnosis of calcifying fibrous pseudotumor is made.


Calcifying fibrous pseudotumor is a rare, benign neoplasm characterized histologically by a hyalinized fibrous stroma with a lymphoplasmacytic infiltrate and psammomatous or dystrophic calcifications. This lesion was first described by Rosenthal and Abdul-Karim1 in 1988 when they reported 2 cases. They called the lesion childhood fibrous tumor with psammoma bodies. In 1993, Fetsch et al2 reported 10 cases of the same lesion. They renamed the entity, citing 3 points: the lesion is not limited to the pediatric population, the calcification is not limited to psammoma bodies, and a neoplastic characterization is not favored. Subsequently, childhood fibrous tumor with psammoma bodies became calcifying fibrous pseudotumor. The Fetsch series included lesions in the extremities, trunk, scrotum, groin, neck, and axilla. Recently, the lesion has also been reported in the pleura,3 the chest wall,4 and the mediastinum.5

REPORT OF A CASE

The following is a case report of a calcifying fibrous pseudotumor presenting as a mass on the posterior side of the neck in a young woman and a review of 19 previously reported cases.

Clinical Presentation

A 24-year-old, previously healthy, Middle Eastern woman presented to her physician with an enlarging, painless mass on the right posterior side of the neck. She had no history of trauma or surgery to her neck and denied any systemic symptoms suggestive of malignancy. On physical examination, the mass was noted to be 4 × 6 cm, firm, mobile, nontender, and located on the right side of midline in the posterior cervical musculature. The patient underwent magnetic resonance imaging with contrast (Figure 1), which showed the mass to be well circumscribed and located between the right splenius and semispinalis cervicis muscles. The mass was not infiltrating the surrounding structures and demonstrated a signal intensity suggesting fibrosis and high collagen content.

Operation

The patient was taken to the operating room and underwent excision of the mass through a midline posterior approach. The mass was noted at surgery to be solitary, lobulated, tan in color, firm, and easily freed from the surrounding tissues. On frozen section at the time of surgery, the diagnosis was “hyalinized fibrous tissue with calcifications.”

Pathologic Findings

Grossly, the lesion was a tan, lobulated mass measuring 5.5 × 4.5 × 2.5 cm, with a firm, white, whorled cut surface. Microscopically, the lesion was well circumscribed but not encapsulated. It was composed of a fibroblast-poor sclerotic stroma with calcification that was predominantly psammomatous and focally dystrophic (Figure 2). There were lymphoid aggregates with admixed plasma cells (Figure 3). Neither atypia nor increased mitotic activity was a feature of the lesion. The lesion was uniformly hypocellular.

Figure 1. MRI of neck, right parasagittal view, showing solitary mass in posterior cervical musculature.

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Clinical Data

<table>
<thead>
<tr>
<th>Variable</th>
<th>Index Case</th>
<th>Rosenthal and Adbul-Karim, 1988¹</th>
<th>Fetsch et al, 1993²</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. of cases</td>
<td>1</td>
<td>2</td>
<td>10</td>
</tr>
<tr>
<td>Age, y</td>
<td>23</td>
<td>2 and 11</td>
<td>1±33 (mean = 16.2)</td>
</tr>
<tr>
<td>Sex</td>
<td>Female</td>
<td>2 Females</td>
<td>5 Male, 5 females</td>
</tr>
<tr>
<td>Size, cm</td>
<td>5.5</td>
<td>5 and 6</td>
<td>2.5–15</td>
</tr>
<tr>
<td>Site</td>
<td>Posterior side of the neck</td>
<td>Upper thigh and forearm</td>
<td>Extremities (3), trunk (2), scrotum (2), groin (1), neck (1), and axilla (1)</td>
</tr>
<tr>
<td>Follow-up</td>
<td>18 mo</td>
<td>6 mo and 6 y</td>
<td>2 mo–10 y (median, 41.5 mo)</td>
</tr>
</tbody>
</table>

Follow-up

Treatment was limited to surgical resection. The patient is free of disease without local recurrence at 21 months.

COMMENT

In the Table, the clinical data of our case and of the previously published cases are summarized. Calcifying fibrous pseudotumor is apparently an entity of young adults, with no particular sex predilection. The lesion has been previously reported in the neck. Malignant transformation has not been reported. In all reports of calcifying fibrous pseudotumor thus far, simple local excision has been curative. Recurrence was noted in only one instance in which the authors speculated incomplete primary excision.

The immunohistochemical staining of the lesion has been reported, but immunohistochemistry is not necessary for diagnosis. The spindle cells stain positively with vimentin and α-smooth muscle actin. Staining for cytokeratin CAM 5.2, epithelial membrane antigen, desmin, muscle-specific actin, factor VIII–related antigen, S100 protein, neurofilament, CD34, and CD31 was reported as negative in one case. The same case was reported to be diploid by flow cytometry.
The pathogenesis of this lesion is not known. In one case, there was a history of antecedent trauma. The microscopic features would suggest a reactive or inflammatory process; nonetheless, cytogenetic abnormalities have been reported in benign fibrous tumors. A recent study indicated that the inflammatory myofibroblastic tumor (also known as inflammatory pseudotumor) has chromosomal evidence of clonality.

There are no published reports of cytogenetic abnormalities in calcifying fibrous pseudotumors. Using the archival paraffin-embedded tissue that was available, we attempted fluorescent in situ hybridization to detect trisomy 7 and trisomy 8, because these karyotypes have been reported in other benign fibrous tumors. The signal intensity was too low to complete the study. Further fluorescent in situ hybridization studies were not attempted.

Other benign soft tissue lesions should be distinguished from calcifying fibrous pseudotumor. Fibromatosis is a lesion that, like calcifying fibrous pseudotumor, can be hypocellular. Psammomatous calcifications and lymphoplasmacytic aggregates are not characteristic of fibromatosis. Compared with inflammatory myofibroblastic tumor, the calcifying fibrous pseudotumor is uniformly hypocellular, whereas the inflammatory pseudotumor can vary in cellularity from field to field.

Although too few cases have been reported to reliably classify the biologic behavior of calcifying fibrous pseudotumor, a review of the literature suggests that when the diagnosis is made, recurrence is not expected. The pathogenesis of the lesion is still unknown. Future cytogenetic studies would be helpful to ascertain whether the lesion is better classified as a neoplasm or a reactive process. Another nomenclature revision may ensue.

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References