Congenital, Self-regressing Tufted Angioma

John Browning, MD; Ilona Frieden, MD; Eulalia Baselga, MD; Annette Wagner, MD; Denise Metry, MD

Background: Tufted angioma (known in Japanese literature as angioblastoma of Nakagawa) is an uncommon, histologically benign, vascular tumor. Lesions typically present during infancy or early childhood and are most commonly reported to persist and/or expand over time. Congenital presentations are rare, as are reports of spontaneous regression.

Observations: We present a series of 5 histopathologically confirmed cases of congenital tufted angioma that spontaneously regressed during infancy or early childhood. We also review the literature, focusing on both congenital and early-onset cases in infants.

Conclusion: We recommend that observation for potential regression be considered for otherwise uncomplicated congenital or early infantile cases of tufted angioma.

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Tufted angioma (TA), also known in the Japanese literature as angioblastoma of Nakagawa, is an uncommon, histologically benign, vascular tumor that most often manifests during infancy or early childhood. It shows characteristic histopathologic findings, with lobules or tufts of capillaries in the dermis. Within the tufts are benign spindle cells, which appear to push against the adjacent vessels, giving them a slit-like appearance. On low-power microscopy, the tufts have the appearance of cannonballs in the dermis.

Most cases of TA are sporadic, although familial cases have been rarely reported. There is no sex predominance, and the clinical presentation is variable. Lesions most often present as solitary tumors or large, infiltrated plaques that are dusky red or violaceous in color. Other characteristic features include increased lanugo hair, an overlying port-wine–like stain, nodularity, or cobblestoning. Tufted angiomas are most commonly reported to persist, often slowly enlarging over years, and may be tender. Kasabach-Merritt phenomenon (KMP) seems to be a rare complication of TA, more common to the kaposiform hemangioendothelioma, although these 2 lesions are considered by many to be within the same spectrum.

Congenital cases of TA are unusual, as are reports of spontaneous regression. We present a series of 5 histopathologically confirmed cases of congenital TA, 4 of which completely regressed clinically during infancy and 1 that had partially regressed clinically by the time the child was 4 years old. We also review 10 prior reports of congenital or early-onset TA.

REPORT OF CASES

In Table 1 we present 5 new cases of congenital TA that spontaneously regressed, 4 during infancy and 1 by the time the child was 4 years old. Biopsy specimens were taken from all patients, and histopathologic findings from all specimens showed classic features of TA. Complete regression of the TA during infancy was observed in 4 patients, 3 of whom had residual skin changes. One patient exhibited partial regression by age 4 years. None of our cases was complicated by KMP. One patient received an intralesional corticosteroid injection whereas the other 4 were observed without intervention.

Ten reports of congenital or early-onset TA, all confirmed by histologic results, have been previously published in the literature; 4 of the lesions were congenital, 5 developed within the first 3 months of life, and 1 was noted by age 9 months. Tenderness or pain was noted in all. None were complicated by KMP. Eight of the 10 patients showed complete or partial regression. The 2 lesions that did not regress were noted to be less tender over time.

COMMENT

Tufted angioma is uncommon but not rare; there are more than 200 reports in the English and Japanese literature. However, presentation at birth is unusual, and reports
### Table 1. Case Series

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset</td>
<td>Birth</td>
<td>Birth</td>
<td>Birth</td>
<td>Birth</td>
<td>Birth</td>
</tr>
<tr>
<td>Age at presentation</td>
<td>5 wk</td>
<td>7 wk</td>
<td>Birth</td>
<td>Birth</td>
<td>3 d</td>
</tr>
<tr>
<td>Sex</td>
<td>Boy</td>
<td>Boy</td>
<td>Boy</td>
<td>Girl</td>
<td>Girl</td>
</tr>
<tr>
<td>Appearance</td>
<td>5 × 4-cm violaceous plaque</td>
<td>1.25 × 1.5-cm firm purple plaque</td>
<td>Firm blue nodule</td>
<td>5 × 4-cm soft dusky red plaque</td>
<td>9 × 7-cm firm violaceous plaque</td>
</tr>
<tr>
<td>Site</td>
<td>Left shoulder</td>
<td>Left cheek</td>
<td>Left forearm</td>
<td>Right temple</td>
<td>Right popliteal fossa</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Soft, nontender; vellus hair</td>
<td>Firm, nontender</td>
<td>Firm, nontender</td>
<td>Soft, nontender</td>
<td>Firm, nontender</td>
</tr>
<tr>
<td>Kasabach-Merritt phenomenon</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Diagnosis confirmed by biopsy findings</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes; staining for GLUT-1 negative</td>
<td>Yes</td>
</tr>
<tr>
<td>Course</td>
<td>Slow growth during first few months of life</td>
<td>Increased growth and firmness until age 4 mo; transient ipsilateral face flushing</td>
<td>Stable, no growth after birth</td>
<td>Rapid involution over 4 mo</td>
<td>Increased size and firmness until age 11 mo, followed by slow decrease in induration and color</td>
</tr>
<tr>
<td>Management</td>
<td>Observation</td>
<td>Intralesional triamcinolone observation</td>
<td>Observation</td>
<td>Observation</td>
<td>Observation</td>
</tr>
<tr>
<td>Outcome</td>
<td>Regression by age 4 mo</td>
<td>Regression by age 15 mo</td>
<td>Regression by age 1 y</td>
<td>Regression by age 5 mo</td>
<td>Partial regression by age 4 y</td>
</tr>
</tbody>
</table>

Abbreviation: GLUT-1, glucose transporter protein isoform 1.

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**Figure.** A, Tufted angioma (TA) involving the right temple at age 3 days; B, the same child at age 5 months following rapid involution of the TA with mild residual erythema, and C, at age 2 years with further resolution of the erythema, revealing cutaneous atrophy with prominent underlying vasculature.
of spontaneously regressing TA even more so; there are only 4 cases confirmed by histologic results reported prior to the series described herein. Regression may be a phenomenon more common to lesions that present at birth or early infancy because there are even fewer reports of spontaneous regression in patients with later presentation. In contrast, to our knowledge there is only 1 report of a congenital TA that failed to improve over time. Other than early onset, we found no other clinical or histopathologic features more common to spontaneously regressing vs persistent lesions.

In the cases reported herein and in prior cases, regression occurred over months to a few years. Most cases completely regressed, whereas in 3 cases partial regression was observed, with decreased induration and tenderness. Prior to regressing, slow growth often occurs (Table 1). Three of our patients had residual skin changes following complete involution, although this finding is not described in previous reports and is common to other spontaneously regressing vascular tumors such as infantile hemangioma and rapidly involuting congenital hemangioma. One child had cutaneous atrophy with prominent vasculature following involuting congenital hemangioma. One child had an overlying eczematous dermatitis after resolution. In conclusion, spontaneous regression of TA may occur and seems to be a phenomenon possibly more common to congenital or early infantile presentations. Whether such lesions represent the same entity as acquired, persistent cases or is a variant is unknown. We suggest that physicians consider observation for otherwise uncomplicated congenital or early infantile cases of TA before considering intervention, particularly surgical intervention.

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REFERENCES