Pediatric Nodular Fasciitis in the Head and Neck
Evaluation and Management

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IMPORTANCE Nodular fasciitis is a rare benign tumor that can present in the head and neck in children. A better understanding of this rare condition is critical to optimize management.

OBJECTIVE To review the presentation, evaluation, diagnosis, and management of pediatric nodular fasciitis of the head and neck.

DESIGN, SETTING, AND PARTICIPANTS Retrospective review of all patients treated for nodular fasciitis of the head and neck over a 20-year period at a pediatric tertiary care center.

INTERVENTION Surgical excision.

MAIN OUTCOMES AND MEASURES Clinical data, including age, presenting symptoms, anatomical site(s), evaluation, treatment, and complications.

RESULTS Fifteen children with pathologically confirmed nodular fasciitis of the head and neck were identified, including 8 boys and 7 girls. The median (range) age at diagnosis was 9.3 years (2 months to 18 years). Patients most commonly presented with a firm, enlarging soft-tissue mass. Two patients reported pain, and 1 patient presented with erythema. The most common location was the maxillofacial region (5 patients). Other locations included the scalp (3 patients), forehead (2 patients), neck (2 patients), mandible (1 patient), postauricular region (1 patient), and nasal dorsum (1 patient). One patient reported a preceding trauma, and 1 patient, a preceding infection. Presurgical imaging varied; imaging modalities used included computed tomography, magnetic resonance imaging, radiography, ultrasound, and sialography. All patients underwent surgical excision, which focused on excising the mass while preserving surrounding normal tissues. Mean (range) follow-up was 7.69 (0-46) months. Two minor complications were reported: 1 patient who underwent a near-total excisional biopsy experienced residual firmness and tenderness at the site of the lesion and another patient was left with an unfavorable cosmetic scar that necessitated intralesional steroid injection. No patient demonstrated recurrence at follow-up.

CONCLUSIONS AND RELEVANCE Although an uncommon diagnosis, nodular fasciitis should be considered in the evaluation and treatment of head and neck soft-tissue masses in children. Preoperative imaging is nonspecific and variable. Pathological findings are necessary for diagnosis. Surgical excisional biopsy is curative, with no instances of recurrence in our series.
Nodular fasciitis is a benign, discrete proliferation of fibroblasts in the subcutaneous tissues often centered on the deep fascia. Its rapid growth can be deceptively similar to that of soft-tissue sarcomas. In addition, pathologic analysis may show high cellularity, high mitotic index, and infiltrative borders, further pointing to malignancy. In the past, this may have led to unnecessary radical surgery as treatment for this benign lesion.

Nodular fasciitis has been described as a reactive phenomenon with an etiology that may be injury related. It is most commonly diagnosed in adults aged 20 to 40 years and has been reported to occur predominantly in males.1 Its prevalence in children is low, with only 10% of reported cases presenting in the pediatric population.2-3 Within the pediatric population, tumor location varies, although it is most commonly reported in the head and neck.4

Published reports of children with nodular fasciitis in the head and neck are limited given the rarity of this condition.4-12 The purpose of this study was to describe our experience with head and neck nodular fasciitis in a pediatric population at Boston Children's Hospital and to delineate its clinical presentation, radiologic appearance, pathologic characteristics, and management. In addition, a literature review and analysis of previously reported cases of nodular fasciitis was undertaken.

Methods

The Boston Children's Hospital institutional review board approved this study, and its guidelines were followed. Informed consent was waived due to the retrospective nature of the study. A retrospective review of medical records was performed on all pediatric patients with a pathologic diagnosis of nodular fasciitis evaluated at Boston Children's Hospital between January 1993 and September 2013. Exclusion criteria included the diagnosis of nodular fasciitis outside the head and neck, age older than 18 years, and absence of clinical data. Data including age at diagnosis, sex, presenting symptoms, comorbidities, diagnostic procedures and imaging, surgical management, complications, and follow-up time were collected and analyzed. In each case, pathologic specimens were reviewed by a senior pathologist (A.P.-A.), and all available images were reviewed by a senior radiologist (V.M.S.).

Results

Nineteen pediatric patients received a diagnosis of nodular fasciitis of the head and neck during the study period. Four patients had confirmed pathologic diagnoses but were not evaluated at Boston Children's Hospital; therefore, no clinical patient data were available for review. These 4 patients were excluded from the study cohort, leaving 15 patients for analysis. Of these 15 patients, 8 were boys, and 7, girls. The median (range) age at diagnosis was 9.3 years (2 months to 18 years). Self-declared ethnicities included white (n = 5), Hispanic (n = 4), Asian Indian (n = 1), and African American (n = 1). Ethnicity data were not available for 4 patients.

The most common location for nodular fasciitis in the head and neck was the maxillofacial region (5 of 15 patients). Other locations included the forehead, scalp and skull, postauricular region, chin, and neck (Table). Seven lesions were on the patient's right side and 8 were on the left. There were no bilateral or multicentric lesions. The mean (range) diameter of the lesion at the time of diagnosis was 2.17 cm (8 mm to 4 cm). Nearly all (14 of 15) lesions were described as firm on presentation. Six lesions were noted to be immobile or fixed; 4 were mobile. The records of the other 5 patients had no documentation on mobility. One patient reported overlying skin erythema, and another had overlying ecchymosis. One patient had an overlying pit but no redness (Figure 1). No other skin changes were noted. None of the patients had concurrent pathological lymphadenopathy.

All patients presented with the complaint of an enlarging soft-tissue mass. One patient endorsed pain at the site of the mass, and another with a temporal mass reported generalized headache. One individual experienced preceding trauma at the site of the mass. In this case, swelling appeared 2 to 3 days after the trauma and increased in size in the following few weeks. Of the remaining patients, 8 denied trauma and trauma data were not available for 6. In regard to infection, 1 individual reported a preceding infection at the site of the lesion; 10 patients denied preceding infection, and 4 did not have recorded data. Time to evaluation ranged from 2 days to 9 months.

Imaging studies available for interpretation varied. Preoperative imaging examinations included 6 computed tomographic (CT) scans, 6 magnetic resonance images (MRIs), 5 ultrasound studies, 3 radiographs, and 1 sialogram. Three patients had no preoperative imaging.

All patients underwent surgical therapy. Eleven of the 15 patients were treated with excisional biopsies or complete resections if they had undergone prior biopsy. Two patients underwent subtotal resections. One patient was treated with an incisional biopsy followed by enucleation. Another patient, who was thought to have had a preceding infection, underwent an in-office incision and drainage with biopsy of what appeared to be a cyst wall. This patient received no additional surgical treatment after the biopsy. Surgical services involved with these patients included general pediatric surgery, plastic surgery, oral and maxillofacial surgery, neurosurgery, dermatology, and otolaryngology.

At follow-up, 2 minor complications were reported: 1 patient who underwent a near-total excisional biopsy experienced residual firmness and tenderness at the site of the lesion and another patient was left with an unfavorable cosmetic scar that necessitated intralesional steroid injection. Mean (range) follow-up was 7.69 (0-46) months. No recurrences were noted after surgical treatment at our institution, although 1 patient presented to us for treatment following 3 recurrences at another hospital.

Discussion

Nodular fasciitis was first described by Konwaler et al13 in 1955. They reported 8 cases of subcutaneous lesions that resembled fibrosarcoma. They termed these lesions pseudosarcomatous...
fibromatosis or fasciitis. Subsequently, in 1961, Stout2 coined the term nodular fasciitis, presuming that the lesions originated from the superficial and deep fascial layers. Interestingly, there have also been reports of this lesion occurring in areas devoid of fascia. Nodular fasciitis has also been referred to as infiltrative fasciitis or cranial fasciitis if located in the scalp region.

Few studies have reported on nodular fasciitis in children. Bemrich-Stolz et al12 reported 18 cases of nodular fasciitis in pediatric patients over a 12-year period. Seven of these lesions were located in the head and neck. Thirteen of the patients in this series were boys. Although a predilection in males has been suggested, our case series showed no sex predilection, with an approximately equal number of boys and girls. Tomita et al6 presented 25 cases of nodular fasciitis over a 10-year period. Of the 3 patients younger than 18 years in the series, 1 had a lesion located in the forehead and those of the others were in the chest wall and left flank. Pandian et al14 compiled one of the larger reviews of pediatric nodular fasciitis in a series of 26 patients. Interestingly, none of the patients in the study had lesions in the head and neck. DiNardo et al4 described 12 cases of nodular fasciitis in children over a 12-year period, and half were located in the head and neck.

In our study, all patients presented with a rapidly enlarging mass. All lesions were unilateral and solitary. Nearly all masses were described as firm. Six lesions were described as fixed or immobile. Similar to reports in the literature, there was no preference in laterality. Two patients had undergone prior evaluation for their masses before presentation at our institution. One patient was noted to have had 3 recurrences at another hospital before requiring a wide local excision with rotational flap. Another patient had been seen at another institution and evaluated after attempts to aspirate the mass had failed.

The most common presenting location was the maxillofacial region. The second most common location was the scalp, at the occiput in 2 patients and the parietal region in

<table>
<thead>
<tr>
<th>ID/Sex/Age, y</th>
<th>Laterality</th>
<th>Location</th>
<th>Diameter, cm</th>
<th>Symptoms</th>
<th>Imaging</th>
<th>Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/F/15</td>
<td>R</td>
<td>Lower occiput</td>
<td>2</td>
<td>Lesion growth</td>
<td>CT</td>
<td>Near-total excisional biopsy</td>
</tr>
<tr>
<td>2/M/15</td>
<td>L</td>
<td>Maxilla</td>
<td>2</td>
<td>Lesion growth</td>
<td>None</td>
<td>Excisional biopsy</td>
</tr>
<tr>
<td>3/F/4</td>
<td>L</td>
<td>Maxilla</td>
<td>2.5</td>
<td>Lesion growth</td>
<td>X-ray, ultrasound, MRI, CT</td>
<td>Incisional biopsy, subtotal resection</td>
</tr>
<tr>
<td>4/F/2</td>
<td>R</td>
<td>Maxilla</td>
<td>2.5</td>
<td>Lesion growth</td>
<td>MRI, CT</td>
<td>Incisional biopsy, enucleation of mass</td>
</tr>
<tr>
<td>5/F/11</td>
<td>L</td>
<td>Nasal dorsum</td>
<td>0.8</td>
<td>Lesion growth</td>
<td>CT</td>
<td>Excisional biopsy</td>
</tr>
<tr>
<td>6/M/2</td>
<td>R</td>
<td>Occiput</td>
<td>4</td>
<td>Lesion growth, skin erythema</td>
<td>Ultrasound, MRI</td>
<td>Incision and drainage, excisional biopsy</td>
</tr>
<tr>
<td>7/F/4</td>
<td>L</td>
<td>Postauricular</td>
<td>3</td>
<td>Recurrence after outside hospital excisions</td>
<td>CT</td>
<td>Wide local excision with rotational flap</td>
</tr>
<tr>
<td>8/F/0.2</td>
<td>L</td>
<td>Parietal skull</td>
<td>2.5</td>
<td>Lesion growth</td>
<td>X-ray</td>
<td>Excisional biopsy</td>
</tr>
<tr>
<td>9/M/18</td>
<td>L</td>
<td>Mandible</td>
<td>1.5</td>
<td>Lesion growth</td>
<td>Ultrasound</td>
<td>Excisional biopsy</td>
</tr>
<tr>
<td>10/F/6</td>
<td>L</td>
<td>Temporal forehead</td>
<td>1</td>
<td>Lesion growth, headaches</td>
<td>None</td>
<td>Incisional biopsy, complete resection</td>
</tr>
<tr>
<td>11/M/3</td>
<td>R</td>
<td>Level 2 neck</td>
<td>3</td>
<td>Lesion growth, overlying skin pit</td>
<td>MRI</td>
<td>Excisional biopsy</td>
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<tr>
<td>12/M/11</td>
<td>L</td>
<td>Inferior SCM</td>
<td>1.8</td>
<td>Lesion growth</td>
<td>X-ray, CT, ultrasound, MRI</td>
<td>Incisional biopsy, complete resection</td>
</tr>
<tr>
<td>13/M/9</td>
<td>R</td>
<td>Cheek</td>
<td>3</td>
<td>Lesion growth, pain</td>
<td>Ultrasound</td>
<td>Punch biopsy, excisional biopsy</td>
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<td>14/M/13</td>
<td>R</td>
<td>Brow, eyelid</td>
<td>1</td>
<td>Increase in size after incisional biopsy</td>
<td>None</td>
<td>Incisional biopsy, excision and local tissue rearrangement</td>
</tr>
<tr>
<td>15/M/17</td>
<td>R</td>
<td>Cheek</td>
<td>2</td>
<td>Lesion growth</td>
<td>X-ray, MRI, sialogram, CT</td>
<td>Excisional biopsy</td>
</tr>
</tbody>
</table>

Abbreviations: CT, computed tomography; L, left; MRI, magnetic resonance imaging; R, right; SCM, sternocleidomastoid muscle.
1. Only 2 patients presented with nodular fasciitis in the neck. One patient had a level 2 neck mass; the other mass was located at the inferior aspect of the sternocleidomastoid muscle. Neither of these patients reported pain or decreased neck range of motion. In fact, most patients did not endorse discomfort at the site of the swelling regardless of the location.

Although the etiology of nodular fasciitis has been suggested as a trauma-related phenomenon, only 1 patient in our series reported preceding trauma before diagnosis. Similarly, only 1 patient in our study group had a preceding infection. This is also the only patient who also presented with overlying skin erythema. This child was treated with an incision and drainage procedure after a trial of oral antibiotic therapy resolved the erythema but not the mass itself. No other patients in our study group identified a particular inciting factor or etiology to their swelling.

In our series, presurgical imaging varied and different imaging modalities such as CT, MRI, radiographs, ultrasound, and sialograms were used. The imaging features of nodular fasciitis are nonspecific and variable. Often a long list of radiologic differential diagnoses is provided in the presurgical evaluation of these patients. In the head and neck, nodular fasciitis may manifest as a discrete solid or cystic-appearing mass, depending on the predominant stromal elements. Lesions may be centered within the subcutaneous space or may be more deeply situated along deep fascia or embedded in muscle. In the head and neck, nodular fasciitis is usually round or ovoid, and well-defined margins are more common than poorly defined ones. Linear extension of the mass along the fascia (“fascial tail sign”) in deeply seated lesions may suggest the correct diagnosis.

On ultrasonography, lesions are hypoechoic or anechoic, homogeneous, and well defined without detectable Doppler signal. No distinctive enhancement pattern has been reported on CT or MRI, and lesional enhancement ranges from mild to marked, homogeneous to heterogeneous, and confluent to ringlike in appearance (Figure 2). Intense vascularity to no vascularity has been described in patients who have undergone catheter angiography. Aggressive features such as rapid growth and bone erosion or destruction can be seen on imaging, leading to preoperative concerns for high-grade malignant neoplasms such as rhabdomyosarcoma or other types of sarcoma (Figure 3). In cases with more benign imaging features, considerations of dermoid or epidermoid, minor salivary gland tumor, aggressive fibromatosis, or dermatofibroma are raised. When the lesion is rapidly growing and accompanied by fever and upper respiratory tract symptoms, these lesions may be mistaken for reactive or infectious adenitis on imaging. Given the rarity of nodular fasciitis and the nonspecific imaging findings, the diagnosis is rarely entertained before surgery. Regardless of this, we recommend preoperative imaging (either MRI or CT) to characterize the lesion, assess the extent of the mass, detect associated bone remodeling or destruction, and to exclude other pathologic entities that may require different management.

All patients in our series underwent surgical treatment, most commonly complete excision. Only 4 patients in our series were treated otherwise. One patient underwent subtotal resection for a maxillary lesion; there was no recurrence reported after 10 months of follow-up. A second patient underwent near-total resection, and his course was complicated by residual firmness and tenderness at follow-up 1 year later. Another patient with a maxillary mass was treated with enucleation, and there were no complications or recurrence noted 3 years postoperatively. The fourth patient presented with a preceding infection and underwent incision and drainage and incisional biopsy. His last follow-up was 2 weeks after the procedure, and no further information was available regarding possible recurrence. This patient also had a complex medical history including 16p11.2 duplication, seizure disorder, and respiratory failure occurring after cardiac arrest, rendering him a poor surgical candidate.

In all patients, diagnosis was made histopathologically. Grossly, the specimens were usually pink-tan to yellow-tan, lobulated, and smooth. In our series, the microscopic appearance showed discrete and nodular hypercellular growth of fi-
broblasts and myofibroblasts centered in the subcutaneous tissue (Figure 4). Prominent mitotic activity, areas rich in ground substance with myxoid and microcystic change, and extravasated erythrocytes and inflammatory cells were often present. This was consistent with the classically described histologic characteristics of nodular fasciitis.23(pp48-49)

Since its first description, the neoplastic nature of nodular fasciitis has been debated. Until more recently, no specific or consistent recurrent cytogenetical abnormality has been found. In 2011, however, Erickson-Johnson et al24 identified high expression levels of USP6 mRNA in 2 examples of nodular fasciitis. USP6 belongs to a subfamily of deubiquitinating enzymes that may serve as oncogenes. Subsequent investigation revealed genomic rearrangements of the USP6 locus in 44 of 48 nodular fasciitis lesions (92%). The authors suggest that USP6 transcriptional up-regulation may be the driving force behind the high proliferative activity and growth of nodular fasciitis. Therefore, in the future, cytogenetic testing for nodular fasciitis may serve as an important diagnostic tool in clinical practice.

Treatment options for nodular fasciitis include observation vs partial or complete resection. Given the typical history of a growing mass and the need for pathologic review for diagnosis, we do not recommend observation alone. In our patient group, no recurrences were reported. Although the majority of our patients did undergo complete surgical resections, the 3 patients in our series who underwent incomplete resection also reported no recurrence. Given the outcomes in this series, it is our recommendation that patients undergo complete local excision for treatment if possible. However, excision should be conservative with sparing of normal anatomy and function.

Limitations to our study are those inherent to a retrospective review. There was no control group of patients treated with observation alone without surgery. The number of patients in our study group is small, but this is expected given the rarity of nodular fasciitis in children. In addition, the mean follow-up time for our patients after treatment was short. Therefore, the incidence of local recurrence or complications in the long term is not known. In addition, patients may have sought follow-up treatment unbeknownst to us at another facility.

Conclusions
Nodular fasciitis is a benign lesion that can be found in the head and neck in children. Its presentation can mimic that of soft-tissue sarcomas given its rapid growth. Although an uncommon diagnosis, nodular fasciitis should be considered in the evaluation and treatment of head and neck soft-tissue masses in children. Patients should undergo preoperative imaging to rule out other potential diagnoses that could alter clinical management. Surgical excision seems to be curative and recurrence is uncommon; thus, more radical surgical procedures can be avoided.
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ARTICLE INFORMATION
Submitted for Publication: June 23, 2014; final revision received September 15, 2014; accepted September 25, 2014.

Author Contributions: Dr Hseu had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study concept and design: Hseu, Watters. Acquisition, analysis, or interpretation of data: All authors. Drafting of the manuscript: Hseu, Rahbar. Critical revision of the manuscript for important intellectual content: Hseu, Watters, Perez-Atayde, Silvera. Statistical analysis: Hseu, Rahbar. Study supervision: Watters, Perez-Atayde, Silvera.

Conflict of Interest Disclosures: None reported.

Previous Presentation: This study was presented as a poster at the American Society of Pediatric Otolaryngology annual meeting; May 16, 2014; Las Vegas, Nevada.

REFERENCES