Self-healing juvenile cutaneous mucinosis: Cases highlighting subcutaneous/fascial involvement

Lavanya V. Nagaraj, MD, a William Fangman, MD, c Wain L. White, MD, a,c,f John T. Woosley, MD, b Neil Prose, MD, c M. Angelica Selim, MD, c,d and Dean S. Morrell, MD a

Chapel Hill, Durham, Winston-Salem, and Greensboro, North Carolina

Background: Self-healing juvenile cutaneous mucinosis is a rare disease affecting young people characterized by transient cutaneous lesions and sometimes mild inflammatory symptoms. The deep dermal and subcutaneous features of this disorder have not yet been well described.

Objective: The purpose of our study was to present 3 cases of self-healing juvenile cutaneous mucinosis in which the histopathologic features caused diagnostic confusion between this disorder and proliferative fasciitis.

Methods: The study includes clinical and histologic findings of 3 patients, complemented by a literature review.

Results: The histologic descriptions of nodular lesions in self-healing juvenile cutaneous mucinosis reveal features of proliferative fasciitis, including a myxoid stroma and gangliocyte-like giant cells.

Limitations: Self-healing juvenile cutaneous mucinosis is a rare condition and has not been frequently reported in medical literature. Our findings are based on the pathologic features of 3 patients.

Conclusions: Our findings further elucidate the histologic features of self-healing juvenile cutaneous mucinosis and expand the differential diagnosis for entities in which gangliocyte-like giant cells are noted. (J Am Acad Dermatol 2006;55:1036-43.)

Pediatric cutaneous mucinoses represent diagnostic challenges for clinicians and pathologists. Case reports and small series reporting the clinical and histologic features of self-healing juvenile cutaneous mucinosis (SHJCM) have been described since 1973. 1-11 Reports of subcutaneous histologic features were first mentioned by Wadee, Roode, and Schulz 8 in 1994 and then again by Cowen, Scott, and Mercurio 2 in 2004. Three cases of SHJCM are presented to highlight subcutaneous histopathologic features which are a potential cause of diagnostic confusion.

CASE REPORTS
Case 1

Patient 1 is an 18-month-old African American girl who presented with numerous nodules on her forehead and extremities that began 2 weeks before her initial presentation. Lesions were increasing in number. Nodules were initially asymptomatic but later were noted to be slightly tender to palpation. The patient was a product of an uncomplicated pregnancy and delivery and had no other medical problems. Her growth and development were appropriate. She did not have systemic symptoms, such as fevers, anorexia, weight loss, or arthralgias.

Physical examination revealed multiple, slightly erythematous, subcutaneous nodules up to 2 cm in diameter, which were most prominent on the forehead but also present on the upper and lower extremities (Fig 1). No periarticular papules were noted. No overlying epidermal change was noted. A deep biopsy of the left leg was performed and results
revealed a bottom-heavy infiltrate, which effaced the panniculus in a diffuse septolobular panniculitis pattern. A proliferation of delicate spindle cells with scattered lymphocytes was present in an abundant myxoid stroma. Scattered large epithelioid ganglion-like mononuclear cells were scattered through the infiltrate (Fig 2). No adjacent fascia was present to evaluate. The dermis contained only a sparse perivascular lymphocytic infiltrate with slightly increased connective tissue mucin. No pools of mucin were seen and no vasculitis was present. A presumptive diagnosis suspicious for proliferative fasciitis was made. An outside soft tissue pathologist was consulted and concurred with the diagnosis.

After presentation of the case and discussion at a regional meeting, the suggestion was made that the case showed features similar to SHJCM. Thus further investigation was prompted, with a re-examination of the biopsy specimen. The review resulted in a diagnosis of “myxocellular subcutaneous nodules with multiple ganglion cell–like and rhabdoid-like fibrohistiocytes, consistent with self-healing juvenile cutaneous mucinosis.”

At 3-month follow-up, the patient was noted to have resolution of some lesions and development of some new lesions, which were also undergoing resolution. She has subsequently been lost to follow-up.

**Case 2**

Patient 2 is a 7-year-old African American boy who presented in May 2003 for further evaluation of multiple subcutaneous skin nodules on the scalp, trunk, and extremities. The patient’s mother reported the initial onset of nodules when the patient was 18 months of age. The initial lesion was described as a “bump” on the posterior scalp that was thought to have occurred after the patient had experienced minor trauma. The lesion subsequently enlarged and was accompanied by the onset of similar lesions on the extremities.

Histopathologic features from excisional biopsy in July 2000 were described as a “fibrovascular hamartomatous process with vascular, myxoid and fibrous components.” New lesions continued to develop, which included involvement of the trunk. In August 2000, biopsy specimens were obtained from nodules on the chest and right knee. In addition, a bone marrow biopsy specimen was obtained; the findings were thought to be unremarkable with the exception of slight hypocellularity for the patient’s age. The cutaneous biopsy specimens were sent to leading soft tissue pathologists for consultation. The overall conclusion was that the biopsy results suggested a “reactive process with vascular proliferation in a myxoid background and variable inflammation.” No specific diagnosis was made, although it was believed that the findings were not suggestive of infantile fibromatosis, common juvenile aponeurotic fibromas, or infantile myofibromatosis. The sections from the excisional biopsy of the chest were reviewed. The specimen consisted entirely of subcutis and a small portion of underlying fascia (Fig 3). There was a prominent diffuse septolobular panniculitis pattern with mucin deposition, focally forming large pools of mucin. A lymphocytic infiltrate was present in septa and in myxoid areas. Large epithelioid ganglion-like mononuclear cells were numerous, especially in myxoid...
areas, often appearing to float within pools of mucin. The lymphocytic infiltrate barely extended into the underlying fascia. The diagnosis was thought to be SHJCM with features similar to proliferative fasciitis.

Throughout the duration of the patient’s course, the lesions have remained asymptomatic. He has not received any topical or systemic therapies for the nodules. His mother denies any exacerbating or alleviating factors. There is no family history of a similar condition. His medical history is remarkable only for mild asthma treated with occasional antihistamines. Findings and results of routine laboratory studies, including complete blood cell count, chemistry panel, liver function tests, rheumatoid factor, and antinuclear antibodies, were unremarkable. *Bartonella* titer was 1:256. A computed tomographic scan of the chest in August 2000 did not show any abnormality.

The lesions spontaneously regressed in late 2001. However, in November 2002, nodules returned to similar locations. His physical examination showed multiple subcutaneous nodules ranging in size from 5 mm to 2 cm on the scalp, forehead, right side of the chin and neck, left side of the abdomen, extensor elbows, extensor knees, distal forearms, and anterior lower legs. Lesions on the hands and significant enlargement of the thumbs were also noted (Figs 4 and 5).

With the characteristic scalp, forehead, and acral lesions and review of the prior histologic findings, a diagnosis of SHJCM was made. Recent correspondence reveals that the patient continues to have subcutaneous nodules that wax and wane. The patient is otherwise healthy and has followed normal growth and developmental curves.

### Case 3

Patient 3 is a 6-year-old Caucasian healthy girl who presented with developing pruritic, nontender skin lesions over a 4-week period, initially on the scalp but with later development in other locations. Prior allergy testing had been performed and the patient took cetirizine nightly. Previous treatment included topical tacrolimus and medium-potency topical corticosteroids, which somewhat alleviated symptoms. Pruritus and swelling of the lesions appeared to be markedly worse after exercise. The patient had no fever or other systemic symptoms.

Physical examination was remarkable for numerous firm, nontender, mobile, subcutaneous nodules, with the largest on the forehead and temples. Nodules were also noted on the nose, upper back, and left lower extremity. A total of 15 to 20 lesions were found. At the time of consultation, *Bartonella* titer was found to be 1:256 and results of complete blood cell count and sedimentation rate were unremarkable. A skull film was within normal limits. A skin biopsy of the lesion on the scalp was performed by the referring physician, which revealed a bottom-heavy mixed inflammatory cell infiltrate that primarily involved the subcutis and subjacent fascia. There was diffuse septolobular involvement with delicate spindle cells in a prominent myxoid stroma admixed with numerous lymphocytes and scattered neutrophils. Throughout the myxoid areas, scattered epithelioid mononuclear cells with prominent nuclei and nucleoli, producing a ganglion cell (gangliocyte) or rhabdoid appearance, accompanied the inflammatory infiltrate. The
inflammation extended into the fascia where edema and lymphocytes separated the adjacent skeletal muscles, producing a “checkerboard” pattern. The dermis contained a sparse perivascular lymphocytic infiltrate with a slight increase in interstitial mucin. No vasculitis was present (Fig 6). Despite the unusual clinical picture with multiple nodules, the initial pathologic diagnosis was thought most likely to be nodular fasciitis with proliferative fasciitis features.

After the biopsy, new lesions continued to develop on the nose, upper back, and distal lower extremities. Over a month, many of the lesions began to resolve with eventual complete resolution. Because of the atypical clinical presentation for nodular fasciitis with multiple lesions, a review of the histologic features and reconsideration of other diagnoses yielded an eventual diagnosis of SHJCM.

DISCUSSION

SHJCM is a rare disease first described in the French literature by Colomb, Racouchot, and Vittori and Bonerandi et al and was first described in the English-language literature in 1984 by Pucejovich et al. This disorder of young persons is characterized by rapid onset of asymptomatic papules and nodules on the face, periaricular regions, abdomen, and thighs. The ages of patients previously described in the literature range from 13 months to 15 years, with one case described in a 26-year-old patient. Lesions are associated with absent to mild inflammatory symptoms and lack extracutaneous involvement. In many reports, SHJCM shows spontaneous complete resolution in weeks to months; however, in several other previously reported cases and in our second case, the course may be more protracted. Painful polyarthritis may be present, as well as swelling of the knees, elbows, and hands.

In contrast to mucinoses affecting adults, SHJCM has not been associated with systemic disorders such as paraproteinemia, bone marrow plasmacytosis, or thyroid disease. One case has been reported coincident with nephroblastoma, whereas another case was associated with bilateral carpal tunnel syndrome. Two prior reports have noted increased aldolase levels in patients with SHJCM, and Cowen, Scott, and Mercurio and Pucejovich et al have proposed that all patients suspected of having SHJCM should undergo creatine kinase and aldolase testing. Carder et al suggested that antinuclear antibodies and complete blood cell count testing may be warranted in patients with signs and symptoms of connective tissue disease, although prior reports have not found any association between SHJCM and autoimmune diseases.

As noted above, two of our patients had positive Bartonella titers. The significance of this is unclear. A prior case report by Al-Matar et al described a 26-month-old boy with nontender, mobile cutaneous nodules on the knees, forearms, shins, elbows, and scalp as well as arthritis and positive titers for both B quintana and B henselae. However, neither of our patients had overt arthritis, and the biopsy performed in the previously published case showed proliferative angiomatosis with a final diagnosis of bacillary angiomatosis.

Classically, 3 types of skin lesions have been described in SHJCM: nontender ivory white papules on the head, neck, trunk, and periaricular regions, which may be grouped in a linear array on the chest and abdomen; deep nodules on the face and periaricular regions; and hard edema of the periorbital and zygomatic areas. The majority of reported cases have included descriptions of biopsy specimens taken from papular lesions rather than deep nodules. The histopathologic findings have classically described edema of the papillary and reticular dermis with separation of the collagen bundles, slightly increased number of fibroblasts, a mild perivascular infiltrate, and Alcian blue staining at pH 2.5, but not at pH 0.5.

Deep dermal and subcutaneous features of SHJCM have only been described in two reports.
Wadee, Roode, and Schulz included histopathologic data from nodular lesions, which exhibited mucin in the mid and deep reticular dermis, arborizing thin-walled vessels, and prominent plump to spindle-shaped fibroblasts. Cowen, Scott, and Mercurio described a case of a 3-year-old Caucasian boy with subcutaneous nodules located on the extremities, scalp, forehead, and axillae. The patient was also noted to have a deep-seated mass on the lower back as well as flesh-colored papules on the extensor knees and overlying the joints on the dorsal aspect of the hands. Results of numerous deep biopsies revealed the majority of histopathologic abnormalities to be in the subcutaneous tissue, with only mildly increased mucin in the dermis. The authors described stellate, spindled, and some rhabdoid or straplike cells embedded in a mucinous stroma. Dense fibrous bands containing fibroblast-like spindled cells were seen. Such significant subcutaneous involvement led consulted experts to also mention nodular fasciitis, proliferative fasciitis, as well as erythema nodosum in the differential diagnosis.

With relative sparing of the reticular dermis and significant subcutaneous involvement, the initial diagnosis entertained in our 3 patients included proliferative fasciitis/nodular fasciitis. Nodular fasciitis, first described in 1955 by Konwaler, Keasby, and Kaplan, is a benign reactive fibroblastic proliferation with a histopathologic appearance that may resemble sarcoma. Soule first used the term “proliferative fasciitis” synonymously with nodular fasciitis in 1962. Chung and Enzinger separated proliferative fasciitis from nodular fasciitis in 1975 based on the presence of ganglion cell-like giant cells in proliferative fasciitis. This entity has a similar appearance to proliferative myositis, which is a pseudosarcomatous proliferation of basophilic giant cells and fibroblasts that infiltrates skeletal muscle along fascial planes. An infiltrative growth pattern is present, and the component cells are plump spindle cells arranged in sheets within a matrix of mucoid material and collagen with a tissue culture appearance. Cellularity varies within lesions, with some fields exhibiting increased cellular density with closely packed spindle cells, collagen formation, and vascular proliferation, alternating with areas with tissue culture appearance. Normal-appearing mitotic figures may be present. The histologic findings are contained within the fascia and interlobular fibrous septa of the adipose tissue. Confusion with sarcoma may arise secondary to the presence of pleomorphism, mitoses, large nuclei, and inclusion-like nucleoli. Cases have been reported in which the lesions were initially interpreted pathologically as rhabdomyosarcoma and malignant fibrohistiocytic neoplasm and treated aggressively with combinations of wide excision, chemotherapy, lymph node dissection, and radiotherapy.

Proliferative fasciitis presents clinically as an asymptomatic, firm palpable subcutaneous nodule. The nodules commonly range from 1 to 5 cm in diameter. Two thirds of lesions occur on the extremities, and a history of trauma can be elicited in one third of cases. Proliferative fasciitis shows an equal sex distribution, and the majority of reported cases have been in adults. No racial predilection has been noted. Multifocal proliferative myositis and nodular fasciitis have been previously reported in the literature, but multifocal proliferative fasciitis has not been previously described.

As in proliferative fasciitis, the initial stimulus for inciting fibroblast proliferation and mucin production in SHJCM is not understood. Abnormal fibroblast proliferation and mucin production are postulated to be secondary to a reactive or reparative response due to chronic antigenic stimulation, such as viral infection or inflammation. In proliferative fasciitis, studies have debated the fibroblastic or pericytic origins of characteristic gangliocyte-like giant cells. Meis and Enzinger found that gangliocyte-like cells stained positively for vimentin, actin, KP-1 (CD 68), and α1-antitrypsin, which suggests a myofibroblastic and histiocytic origin. In a later case report, Kiryu, Takeshita, and Hori reported positive staining in fibroblasts for vimentin and actin, implying a myofibroblastic origin, whereas gangliocyte-like giant cells stained positively for vimentin and negatively for all other markers, including α-actin, lysozyme, cytokeratin, and S-100 protein; thus a fibroblastic origin was asserted. In contrast, Honda et al reported a case of proliferative fasciitis in an adult occurring in the abdominal region in which both gangliocyte-like giant cells and fibroblasts showed positive vimentin staining but negative smooth muscle actin staining, consistent with a fibroblastic origin.

Table I summarizes the findings of our cases as well as those of other reported cases of SHJCM, and Table II shows the main clinical and histologic features of other diseases that are included in the differential diagnosis of SHJCM.

As is evident from our case histories, we experienced difficulty in making the correct diagnosis. Our aim is to assist other clinicians and pathologists in similar cases, especially when potentially misleading histologic features are present in a relatively rare condition. In each of our cases, resolving and recurrent deep dermal and subcutaneous nodules were present in multiple locations including the scalp, torso, and extremities. The subcutaneous histologic
features lead multiple dermatopathologists and soft tissue pathologists away from the eventual correct diagnosis. The diagnostic confusion in our cases resulted from the pattern of deep-seated subcutaneous and fascial involvement and the presence of ganglion-cell like fibrohistiocytes. The report by Cowen, Scott, and Mercurio was pivotal in our establishing the diagnosis of SHJCM, for it clearly demonstrated the linkage in the same patient between the more superficial dermal lesion with pools of connective tissue mucin and the deep-seated proliferative fasciitis picture. As reflected in that report and in the experience with our cases, the subcutaneous and fascial histopathology in SHJCM, in any given field, is essentially indistinguishable from proliferative fasciitis. Connective tissue mucin is prominent in typical fasciitis, but in SHJCM sheets and pools of mucin often predominate over the cellular infiltrate throughout much of the subcutis. In patients with SHJCM, who either do not have separate superficial lesions or do not have biopsy specimens taken from the superficial lesions, the dominant fasciitis changes in the deep lesions detract the pathologist from the less dramatic collections of the dermal mucin (Fig 6, A). This dermal mucin is the common denominator histologically in both types of clinical lesions and, along with the extensive subcutaneous mucin deposition, is the only other histologic clue that the patient may have a disorder distinct from more conventional fasciitis. After further review of the histopathologic data, clinical evolution, and the literature, our view is that the correct diagnosis in these 3 cases is indeed SHJCM, in which biopsy results of the nodular lesions show deep-seated changes similar to nodular fasciitis/proliferative fasciitis.

Table I. Summary of reported cases of self-healing juvenile cutaneous mucinosis

<table>
<thead>
<tr>
<th>Reference No.</th>
<th>Age</th>
<th>Sex</th>
<th>Location of papules/nodules</th>
<th>Associated findings</th>
<th>Duration of lesions</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>13 y</td>
<td>F</td>
<td>Scalp, hands, knees, elbows, sacrum</td>
<td>None</td>
<td>2.5 y</td>
</tr>
<tr>
<td>4</td>
<td>14 y</td>
<td>M</td>
<td>Face, neck, scalp, trunk, thighs, hands</td>
<td>Hypertension, arthralgias</td>
<td>Few weeks</td>
</tr>
<tr>
<td>5</td>
<td>13 y</td>
<td>M</td>
<td>Face, trunk, knees, hands, elbows</td>
<td>Carpal tunnel syndrome, arthralgias, myalgias, weakness, anorexia, hoarseness, lymphocytosis</td>
<td>2.5 y</td>
</tr>
<tr>
<td>8</td>
<td>8 y</td>
<td>M</td>
<td>Scalp, face, arms, hands, sacrum</td>
<td>Nephroblastoma</td>
<td>2.5 y</td>
</tr>
<tr>
<td>11</td>
<td>6 y</td>
<td>M</td>
<td>Thighs, arms</td>
<td>None</td>
<td>3 mo</td>
</tr>
<tr>
<td>7</td>
<td>5 y</td>
<td>M</td>
<td>Face, knees, hands</td>
<td>Scleredema (face), arthralgias, fever, myalgias, weakness, elevated ESR and WBC</td>
<td>2 mo</td>
</tr>
<tr>
<td>26</td>
<td>15 y</td>
<td>M</td>
<td>Trunk, face, scalp, forearms, hands, knees</td>
<td>None</td>
<td>7 mo</td>
</tr>
<tr>
<td>13</td>
<td>14 mo</td>
<td>F</td>
<td>Scalp, face, hands, trunk, legs, arms</td>
<td>Lymphocytosis</td>
<td>Began resolution at 5 mo</td>
</tr>
<tr>
<td>9</td>
<td>21 mo</td>
<td>F</td>
<td>Face, trunk, periarticular regions</td>
<td>None</td>
<td>3 mo</td>
</tr>
<tr>
<td>2</td>
<td>3 y</td>
<td>M</td>
<td>Pretibial legs, axillae, scalp, forehead, knees, hands, trunk, periorbital</td>
<td>Refusal to walk, transient hypertension, elevated aldolase, MRI with evidence of myositis and subcutaneous edema</td>
<td>8 mo</td>
</tr>
<tr>
<td>Current case 1</td>
<td>18 mo</td>
<td>F</td>
<td>Head, upper and lower extremities</td>
<td>None</td>
<td>Resolution initiated at 3 mo</td>
</tr>
<tr>
<td>Current case 2</td>
<td>7 y</td>
<td>M</td>
<td>Scalp, chin, trunk, extremities</td>
<td>Bartonella titer 1:256</td>
<td>Continued development and resolution of nodules</td>
</tr>
<tr>
<td>Current case 3</td>
<td>6 y</td>
<td>F</td>
<td>Scalp, face, trunk, leg</td>
<td>Bartonella titer 1:256</td>
<td>Resolution at 1 mo</td>
</tr>
</tbody>
</table>

ESR, Erythrocyte sedimentation rate; MRI, magnetic resonance imagine; WBC, white blood cell (count).
Data from Carder et al.11
The gangliocyte-like giant cells initially thought to be characteristic of proliferative fasciitis appear to be the same cells previously described as rhabdoid or straplike cells by Cowen, Scott, and Mercurio. In fact, a MEDLINE query using the terms “gangliocyte-like cell,” “gangliocyte-like giant cell,” “gangliocyte-like fibrohistiocytes,” and “gangliocyte-like fibrous histiocyte” results in citations regarding literature about gangliocytic paraganglioma of the duodenum, polyoid nonchromaffin paraganglioma, and proliferative fasciitis. When these cells are noted on a histopathologic specimen, the differential diagnosis, in the correct clinical setting, should be expanded to include SHJCM.

These cases illustrate that when the diagnosis of proliferative/nodular fasciitis is rendered histopathologically in pediatric patients with multiple lesions, SHJCM should be a major diagnostic clinical consideration. Attention to the histologic detail of increased mucin in the papillary and superficial reticular dermis and predominating throughout much of the fasciitis, couched together with the clinical context,

Table II. Clinical differential diagnosis of self-healing juvenile cutaneous mucinosis

<table>
<thead>
<tr>
<th>Disease</th>
<th>Main clinical features</th>
<th>Main histologic features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Papular mucinosis of infancy</td>
<td>Congenital or perinatal, not self-healing, no polyarthritis</td>
<td>Papillary dermal mucin, usually no significant increase in fibroblasts</td>
</tr>
<tr>
<td>Mucinosis due to ingestion of toxic oil</td>
<td>Epidemic, interstitial pneumopathy and neuromuscular disorder</td>
<td>Dermal deposits of nonsulfated acid mucopolysaccharides with increased mast cells</td>
</tr>
<tr>
<td>Scleromyxedema</td>
<td>Slow-onset, urticarial plaques, no resolution, paraproteinemia</td>
<td>Dermal mucin, increased collagen deposition, proliferation of fibroblasts</td>
</tr>
<tr>
<td>Lipoid proteinosis</td>
<td>Autosomal recessive inheritance, hoarseness, “moniliform blepharosis,” husky voice</td>
<td>Extensive deposits of amorphous, eosinophilic, periodic acid—Schiff positive material</td>
</tr>
<tr>
<td>Juvenile hyaline fibromatosis</td>
<td>Progressive and disabling, gingival hypertrophy, flexion contractures of joints, osteolytic defects, stunted growth</td>
<td>Thickened dermis with chondroid appearance, fibroblast-like cells in amorphous eosinophilic ground substance</td>
</tr>
<tr>
<td>Fibroblastic rheumatism</td>
<td>Sudden onset, symmetric polyarthritis, sclerodactyly with contracture of palmar aponeurosis, Raynaud’s phenomenon, lung fibrosis</td>
<td>Plump myofibroblasts and fibroblasts in background of thickened and whorled collagen bundles</td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
<td>Deforming arthritis, cutaneous manifestations include rheumatoid nodules or rheumatoid vasculitis; typical serologic features</td>
<td>Nodules characterized by fibrinoid necrosis and palisaded histiocytes in the subcutis or deeper</td>
</tr>
<tr>
<td>Francois syndrome</td>
<td>Xanthoma-like lesions, hyperplasia of gingival mucosa, osteochondrodystrophy, bilateral corneal dystrophy</td>
<td>Early lesions contain bulky cells with light cytoplasm and eccentric nuclei (spongiocytes); late lesions contain compact fibrous tissue with few cells</td>
</tr>
<tr>
<td>Winchester syndrome</td>
<td>Dwarfism, osteolysis, corneal opacities, rheumatoid-like joint destruction, hypertrichosis, thickening of skin, widespread nodular lesions</td>
<td>Thickened dermis, marked increase in fibroblasts, no increase in dermal mucopolysaccharides</td>
</tr>
<tr>
<td>Multicentric reticulohistiocytosis</td>
<td>Mucocutaneous lesions and severe polyarthritis</td>
<td>Histiocytes, multinucleate cells, ground glass—like appearance</td>
</tr>
<tr>
<td>Scleredema</td>
<td>After streptococcal illness or associated with diabetes or monoclonal gammopathy, diffuse induration of the skin, no polyarthritis</td>
<td>Swollen, thickened dermal collagen fibers with mucopolysaccharide deposition</td>
</tr>
<tr>
<td>Proliferative fasciitis</td>
<td>Asymptomatic, firm palpable subcutaneous nodule; 1 to 5 cm in diameter; two thirds of lesions on extremities, may be associated with history of trauma</td>
<td>Plump spindle cells, gangliocyte-like giant cells, tissue culture appearance, infiltrative growth pattern</td>
</tr>
</tbody>
</table>

Data from references 1, 7, 14, 15, 27, 28.
should lead to the correct diagnosis of SHJCM, even in patients who do not have separate characteristic superficial lesions with pools of dermal mucin.

We thank Dr Ilona Frieden for assisting with the diagnosis of patient 1. We also thank Drs Sharon Weiss and Antonio Nascimento for their consultation on the biopsies of patient 2, as well as Drs William T. Sumner and William D. Hoover for their clinical observations of patient 3.

REFERENCES