A previously healthy 3-year-old boy presented to the emergency room with a sudden onset of enlargement of the right mandible. Evaluation included routine laboratory tests and radiographs of the chest and neck, all of which were within normal limits. There was no history of trauma, fever, chills, or night sweats. The patient was diagnosed with a "reactive lymph node" and started on a 5-day course of antibiotics, with no improvement in symptoms. The patient presented again to his primary care provider with continued enlargement of the right mandible, which was treated with 2 additional courses of antibiotics. The treatment was complicated by an allergic reaction, and resolution of symptoms did not occur.

Two months later, computed tomographic scans (Figure 1) revealed an aggressive-appearing soft tissue mass arising within the right anterior mandible. The mass measured approximately $3 \times 4$ cm and showed perforation of the lingual cortex with invasion of the soft tissues of the floor of the mouth. The mass was slightly lower in attenuation than the neighboring muscles, suggesting the presence of watery or myxoid material. No matrix or ossification was noted. The extraosseous portion appeared to be well defined, suggesting an intact periosteum. The radiographic differential diagnosis included odontogenic myxoma, desmoplastic fibroma, and myofibromatosis.

Incisional biopsies of the intrabony mass and its soft tissue extension were performed. Both biopsy specimens consisted of a moderately dense, spindle-shaped cellular proliferation within a variably myxomatous and collagenized stroma (Figure 2; hematoxylin-eosin, original magnification $\times 20$). The spindle cells were arranged in a predominantly fascicular pattern. Variably dense collagen deposition was noted within a myxomatous extracellular matrix. The vesicular nuclei varied from small and thin to plump and polygonal, with 1 to 2 nucleoli. The spindle-shaped cells displayed narrow to polygonal vesicular nuclei with 1 to 2 small eosinophilic nucleoli (Figure 3; hematoxylin-eosin, original magnification $\times 100$). A few scattered, normal-appearing mitotic figures were evident.

What is your diagnosis?
Desmoplastic fibroma (DF) was first described as a distinct entity and given its present name by Jaffe in 1958. It is generally reported that DF of bone represents the intraosseous component of soft tissue fibromatosis or desmoid tumor. In 1856, Paget correlated the histologic appearance of the abdominal DF with similar lesions in extra-abdominal sites, such as the shoulder girdle and upper arm. The first cases of this lesion reported in bone were primarily located in the metaphysis of long bones.

Desmoplastic fibroma is one of the rarest bone tumors. There is only 1 case among the 776 benign bone tumors in the Dutch bone tumor registry. Dahlin and Unni presented 9 cases of DF in a series of 8542 primary bone tumors (0.11%). Böhm et al reviewed 191 cases of DF reported in 80 publications. In their review, the age of patients ranged from 15 months to 75 years, with a reported mean age of 23 years. Approximately 16% of the patients were in their first decade, 35% were in their second decade, and 23% were in their third decade. Seventy-four percent of the patients were younger than 31 years, and only 6% were older than 50 years. According to published data on tumor location in 184 patients, DF most often involved the following bones: mandible (22%), femur (15%), pelvic bones (13%), radius (12%), and tibia (9%).

The cause of DF is unknown, although trauma and genetic factors have been suggested. The most likely precipitating factor is trauma to the affected region. In long bones, the most common presenting sign of disease is a pathologic fracture. In the facial skeleton, a gradual swelling, asymmetry, or both are the most common presenting symptoms to alert the patient or clinician to its presence. Pain and functional impairment are occasionally present. Incidental discovery of asymptomatic lesions has also been noted.

Grossly, desmoid tumors are firm, rubbery, white, non-encapsulated, fibrous-appearing lesions. The nonspecific radiologic features include a unilocular or multilocular, encapsulated, fibrous-appearing lesions. The nonspecific morphological profile includes positive reactivity for smooth muscle actin and S100. Desmoplastic fibroma of bone may resemble the desmoid tumor of soft tissue when the latter infiltrates bone. In these cases, only clinical and radiographic features are helpful in the differential diagnosis.

Desmoplastic fibroma is generally classified as a benign tumor and has not been shown to metastasize. Varying surgical procedures have been used in the management of DF, including local curettage, wide excision, resection, or mandibulectomy. Local curettage has the highest rate of recurrence, so wide surgical excision is recommended. Radiation therapy is not indicated as a primary treatment, but has been shown to slow growth of lesions that have been incompletely excised. Chemotherapy has been used as an adjunct to total resection; however, most clinicians favor surgical resection as the treatment of choice.

Because of the tendency for recurrence, which has been reported to be as high as 35%, close postoperative follow-up is essential. Correlation of clinical, radiographic, and histologic findings is imperative for determining a definitive diagnosis and treatment plan for this potentially aggressive and recurrent lesion.

References