Aggressive Fibromatosis of the Mandible in Childhood

David J. Sartoris, M.D. 1, Robert S. Arkoff, M.D. 2, and Bruce R. Parker, M.D. 1

1 Departments of Radiology, Stanford University Medical Center, Stanford, California, USA, and
2 The Children’s Hospital and Adult Medical Center, San Francisco, California, USA

Abstract. Aggressive fibromatosis, or infantile fibrosarcoma, is an uncommon form of juvenile fibromatosis which rarely involves the head and neck. Skeletal involvement is infrequently demonstrated by radiography in this condition. Two unusual cases with similar radiographic changes in the mandible are presented, a situation not previously described. Clinical, pathologic, and radiographic features of aggressive fibromatosis are discussed.

Key words: Fibromatosis – Aggressive fibromatosis – Fibrosarcoma – Mandible – Neoplasm – Fibrous tumor

Aggressive fibromatosis, also known as infantile fibrosarcoma, is an uncommon subtype of the juvenile fibromatoses which rarely involves the mandible. Two cases with strikingly similar radiographic changes in the mandible are presented. These features have not, to our knowledge, been heretofore described in this condition.

Case Reports

Patient 1 is an 8-month-old male who presented with progressive jaw enlargement over the preceding 6 months. History was otherwise unremarkable. Physical examination revealed a firm, non-tender mass which appeared contiguous with the mandible. Radiographic evaluation demonstrated a large soft tissue mass, predominantly on the left, with erosive changes and reactive bone formation in the mandible (Fig. 1). Biopsy revealed aggressive fibromatosis without evidence of sarcoma. Reactive bone was present in the biopsy specimen. The patient was placed on chemotherapy including vincristine, actinomycin-D, and cyclophosphamide with resolution of the mass [16]. When chemotherapy was stopped after one year, the tumor mass recurred. Reinstitution of chemotherapy resulted in further tumor regression. The patient was subsequently lost to follow-up.

Address reprint requests to: Bruce R. Parker, MD., Division of Diagnostic Radiology, Stanford University Medical Center, Stanford, CA 94305, USA

Fig. 1. Patient 1. Oblique view of the left side of the mandible reveals a large soft tissue mass with pressure erosion of the inferior cortex of the mandible and thick periosteal reaction. The bone does not appear infiltrated

Fig. 2. Patient 2. Oblique view of the right side of the mandible reveals a 2 1/2 x 3 cm well defined soft tissue mass with reactive bone formation and periosteal reaction. The bone is not infiltrated.

Patient 2 is a 2 1/2-year-old male with a history of a mass in the left jaw for 2 months. Physical examination revealed a 2 x 3 cm localized hard mass attached to the inferior aspect of the left side of the mandible. Radiographic evaluation demonstrated a soft tissue mass with erosive and periosteal changes in the mandible (Fig. 2). Excisional biopsy was performed. The initial histologic interpretation was neurofibroma, but review of the slides revealed aggressive fibromatosis. The patient had no other stigmata of neurofibromatosis. There has been no recurrence of disease.

Discussion

Aggressive fibromatosis, or infantile fibrosarcoma, is a rare form of the juvenile fibromatoses [3, 17] which include fibromatosis colli, fibrous hamartoma of infancy, recurrent digital fibrous tumor, juvenile aponeurotic fibroma, and congenital gen-
Fig. 2. Patient 2. Oblique view of the mandible demonstrates external pressure erosion with thick periosteal reaction. The radiographs suggest involvement of an undeveloped tooth bud, but no mandibular invasion was identified at the time of excisional biopsy.

eralized fibromatosis [5, 6, 10, 11, 13]. The tumor usually presents during the first two years of life and demonstrates a slight predilection for males [3, 6, 7]. Although any site may be involved, the extremities are favored [2, 3, 7]. Rapid growth is the rule, particularly in younger children, and there is a high propensity for local recurrence following therapy (17–43%) [2, 3, 5–7, 15]. Since only about 8% of tumors metastasize to lung, liver, and lymph nodes, the prognosis is more favorable than that of adult fibrosarcoma [3, 5, 14, 15, 17]. Five-year survival as high as 85–90% has been reported [3, 7, 15].

Grossly, the tumor is firm, shiny, and grayish-white, with variable areas of collagenization, myxomatous degeneration, hemorrhage, and necrosis. An enclosing pseudocapsule formed by compressed surrounding tissue may be present [7]. Infiltration of muscle, fascial structures, tendons, and subcutaneous fat is seen [6]. Deep tumors may involve the periosteum [5, 17]. Microscopically, interlacing fascicles of fusiform or spindle-shaped cells with reticulum fibers and collagen intermix with round cell aggregates and cleft-like or cavernous vascular spaces. Variable numbers of mitotic figures are present, but cellular pleomorphism and giant cells are rare [3, 6, 15, 17].

Subdivision into well- or poorly-differentiated and medullary or desmoid types has been made [4, 7, 15, 18]. Variability may exist within a given tumor, and histology does not correlate with clinical behavior or prognosis [2, 3, 5, 8, 15]. Pathologic distinction between “fibromatosis” and “fibrosarcoma” is thus no longer attempted [3, 7, 15, 18].

The etiology of aggressive fibromatosis is unknown. Trauma and prior therapeutic radiation have been implicated [18]. Wide field excision is the accepted mode of therapy, with amputation usually reserved for large or recurrent cases [2, 3, 15, 18]. Although few advocate local irradiation, definite tumor responses and several cures have been reported [5, 12, 14, 19]. Chemotherapy has been advised for surgically inaccessible cases, rapidly invasive lesions, recurrence, or metastasis [3, 14]. Results have generally been poor [7, 18]. As both late local recurrences and metastases have been documented, long-term clinical follow-up is advised [3, 7, 8].

Both the tumor location and extent of mandibular involvement seen in our patients are unusual. Swain et al. described three tumors which presented clinically as mandibular masses. Histologically these were felt to have arisen from the periosteum or connective tissue overlying the bone [18]. Radiographic findings were not reported. In various series, the incidence of head and neck involvement has ranged from 0 to 43%, and averages about 13% [2, 3, 6, 7, 15, 17].

A small number of reported infantile fibrosarcomas have demonstrated skeletal changes radiographically [1, 2, 9]. Bowing, cortical thickening, and other manifestations of chronic extrinsic pressure deformity, and/or cortical invasion and erosion, with or without extension to the medullary cavity, have been observed. Reactive changes have been reported in the humerus, ulna, radius, tibia, and fibula [2, 3, 5, 6]. Destructive changes have been described in the ulna, radius, tibia, fibula, and proximal phalanx of the thumb [1–3, 5, 15]. Pathologic fracture may occur [2]. Calcification of the soft tissue mass has been seen radiographically in several instances [5, 6].

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References