Ameloblastic fibrosarcoma of the maxillary sinus in an infant: a case report with long-term follow-up

Nona Zabolinejad⁎, Mehran Hiradfarb, Kazem Anvari, Alale Shojae Razavidad

Department of Pathology, Dr Sheikh Children Hospital, 1358 Mashhad, Iran
Department of Pediatric Surgery, Dr. Sheikh Children Hospital, 1358 Mashhad, Iran
Department of Oncology, Omid Hospital, 1358 Mashhad, Iran
Research Development Unit, Dr Sheikh Children Hospital, 1358 Mashhad, Iran

Received 27 May 2007; revised 10 September 2007; accepted 14 September 2007

Index words:
Ameloblastic fibrosarcoma; Ameloblastoma; Maxillary sinus

Abstract
Ameloblastic fibrosarcoma (AFS) or ameloblastic sarcoma is an extremely rare odontogenic neoplasm. The authors report AFS in the maxillary sinus of a 4-month-old boy. The tumor was composed of odontogenic epithelium, resembling that of ameloblastoma, and a mesenchymal part exhibiting features of fibrosarcoma. We also found some areas with deposition of dentinoid material closely adjacent to the ameloblastic epithelium. Although AFS has occurred in a wide age range, this is the first report of this tumor in infancy with long-term follow-up. © 2008 Elsevier Inc. All rights reserved.

Ameloblastic fibrosarcoma (AFS), first described by Heath [1] in 1887, is an extremely rare malignant odontogenic tumor. It is composed of a benign odontogenic epithelium, resembling that of ameloblastoma, and a mesenchymal part exhibiting features of fibrosarcoma.

Until now, 64 cases have been reported in the English literature. Most of them have occurred in the mandible and in the third decade of life.

In this article, an unusual case of AFS that arose in the maxilla of a 4-month-old boy is described, and a brief review of its clinicopathologic features is given.

1. Case report

A 4-month-old boy with a tumoral lesion in left maxillary region was referred to our pediatric health center. His mother had noticed this rapidly growing mass about 1 week before admission. On admission, the patient had fever. Extraorally, the face was asymmetrical because of a large and firm swelling of the malar region. The intraoral examination revealed buccal and palatal cortical bone expansion affecting the left posterior region of the maxillary alveolar ridge extending over the hard and soft palates and the mucobuccal fold with no clinical evidence of perforation. The other findings from a general physical examination were normal, and laboratory values were in normal ranges. Water’s x-ray showed a radiopaque lesion caused cloudiness of the left maxillary sinus and destruction of medial and superior walls of sinus. Computed tomographic scan also revealed a soft tissue mass with a maximum diameter of about 12 cm in the left maxillary sinus with invasion to pterygopalatine space, protruding toward the orbital cavity, and destruction of peripheral bones (Zigoma, maxilla, and basal and medial walls of the orbit), as well as septal deviation (Fig. 1).

An incisional biopsy was performed. Microscopic examination showed a malignant neoplastic lesion composed of 2 typical components: (1) benign epithelial tissue

⁎ Corresponding author. Tel.: +98 511 7280217; fax: +98 511 7277470. E-mail address: nonazabolinejad@yahoo.com (N. Zabolinejad).
indistinguishable from ameloblastoma with a central network of interconnected stellate-shaped cells outlined by peripheral palisaded columnar cells with hyperchromatic nuclei polarized away from the basement membrane and clear vacuolated cytoplasm and (2) malignant mesenchymal tissue with rounded to spindle-shaped cells arranged in fascicular pattern (Fig. 2). These stromal cells exhibited moderate nuclear pleomorphism, hyperchromatism, and mitotic figures (up to 5 mitoses per high-power field). There were also wide areas of necrosis. Immunohistochemically, malignant spindle cells were positive for vimentin but negative for smooth muscle actin, MyoD1, and S-100 protein. Ameloblastic epithelium showed positive immunoreactivity for pancytokeratin. A diagnosis of AFS was made. The patient underwent Caldwell-Luc approach of the left maxillary sinus, and the tumor was completely excised with removal of the cortical bone of the maxillary sinus roof. The cancellous bone around the maxillary sinus was curetted. The orbital side of the bony cortex of the maxillary roof was retained.

In this specimen, we also found some areas with deposition of dentinoid material closely adjacent to the ameloblastic epithelium, which classified the diagnosis as ameloblastic fibrodentinosarcoma (Fig. 3). The patient received radiation therapy (irradiation of 50 Gy over 7 weeks, administered in 150-cGy fractions daily) and, at
4.5 years' follow-up, showed no evidence of radiographic and clinical recurrence (Fig. 4).

2. Comment

Ameloblastic fibrosarcoma or ameloblastic sarcoma is an extremely rare odontogenic neoplasm. As far as we know, 64 cases have been reported in the English literature up to 2006 [2]. Development of this lesion in the jaws either de novo or from preexisting ameloblastic fibroma has been well documented. Ameloblastic fibrosarcoma occurs in a wide age range, from 3 to 83 years (mean, 27.3 years), but no case of this tumor has been reported in infancy before. The male-to-female ratio is 1.6:1. The most commonly affected site within the jaw is the posterior part of the mandible. In our patient, the lesion was located in the maxillary sinus, which is a rare location according to the literature. Clinical appearance and symptoms vary among the reported cases, but swelling and pain are the most constant findings [3-10].

The histologic study of the present case revealed a highly cellular mesenchymal tissue with spindle-shaped cells with marked pleomorphism, arranged in a fascicular pattern. There were also small areas with deposition of dentinoid material closely adjacent to ameloblastic epithelium. The World Health Organization distinguishes odontogenic sarcoma devoid of dental hard tissue (AFS) from those displaying focal evidence of dentinoid (ameloblastic fibrodentinosarcoma) or dentinoid plus enamoloid (ameloblastic fibrodontosarcoma), but the World Health Organization panel acknowledges that presence or absence of dental hard tissue in an odontogenic sarcoma is of no prognostic significance [11].

The information available concerning the treatment, course, and prognosis of AFS is limited because of the paucity of cases reported. According to previous reports, 20% of patients have died within 3 months to 19 years, resulting from locally aggressive tumor growth [2].

In view of the local aggressiveness and its high tendency to recur (37% of the reported cases have had at least 1 recurrence [2]), the treatment of choice is wide surgical excision with long-term follow-up. Because documented metastasis has been reported only once in the literature, routine neck dissection for AFS seems to be unnecessary [4].

Our patient received radiation therapy after surgical removal of the tumor. During 4.5 years’ follow-up, no recurrence occurred. Adjuvant postoperative radiotherapy, as in our patient, was used successfully in a case with a very extensive maxillary lesion by De Nittis et al [5]. The use of radiation therapy in the management of this patient as in the other childhood cancers might be determined, in part, by a knowledge of the late effects on the normal tissues. Clinically, radiation effects on the growing bone might be hypoplasia of bones (such as the maxilla) and on the eye include tissue necrosis, decreased tear production, telangiectasia, scleral melting, cataract, corneal neovascularization, radiation retinopathy, and radiation-induced cancers [12]. Although retarded bone growth, to some extent, occurred in our patient, fortunately, it did not cause any significant facial asymmetry, and the last ophthalmologic examination did not define a remarkable side effect on his eye. Our patient received 50 Gy in divided doses (150 cGy daily), and this is most likely the reason for the lack of clinically significant ocular side effects. We recommend adding radiation to treatment of this tumor for prevention of recurrence, especially in cases of incomplete surgical removal.

References


