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Case report

A rare case of aggressive fibromatosis in the maxillary sinus

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ABSTRACT

Aggressive fibromatosis (AF) is a locally aggressive benign soft tissue tumor. It is rarely observed in the head and neck region and is particularly uncommon in the maxillary sinus. We report the case of a 40-year-old man who presented with left nasal purulent discharge and dull pain in the left maxillary region. Computed tomography of the head and neck region revealed an infiltrative mass in the left sinus and bone resorption of the posterior wall of the maxillary sinus and alveolar bone. Examination of biopsy samples, obtained twice, did not lead to a definite diagnosis. The tumor was removed by radical sinusotomy under general anesthesia. Histopathological analysis of the excised tumor revealed AF. No recurrence or metastasis was found 2.5 years after tumor excision. As AF of the oral and maxillofacial region frequently recurs, we recommend long-term follow-up of the patient.

1. Introduction

Aggressive fibromatosis (AF), also known as a desmoid tumor or desmoid-type fibromatosis, is a rare benign connective tissue process originating from the fascia and musculoaponeurotic tissues and shows locally aggressive behavior. Although characterized by local invasion and associated with a high rate of local recurrence, it is not known to metastasize. Recent studies have shown that AF is derived from mesenchymal stem cells [1]. The most common sites of these lesions are the mesentery, retroperitoneal space, and abdominal wall or extra-abdominal sites such as the trunk and extremities [2], but AF of the maxillary sinus has been rarely reported [3]. In this report, we describe a rare case of AF in the maxillary sinus of an adult man.

2. Case report

A 40-year-old man was referred to and examined at the Clinic of Oral and Maxillofacial surgery of our hospital in June 2010. He presented with left nasal purulent discharge and dull pain in the left maxillary region. The ENT department had diagnosed odontogenic sinusitis before the patient was referred to our department. He was 148 cm tall and weighed 49 kg; his family medical history was unremarkable. Intraoral findings included a half-impacted left third molar tooth. A slight swelling and redness was observed around the tooth. Computed tomography revealed a large expansile mass, 40 mm × 32 mm × 30 mm in size, that had expanded and eroded the posterior walls of the maxillary sinus and the alveolar bone (Fig. 1A and B). Biopsy samples of the tumor, obtained twice using the intraoral approach, were examined in June 2010. No diagnosis was reached as pathology only revealed hyperplasia of the fibrous tissue and inflammatory cells (Fig. 2). Therefore, we excised the tumor under general anesthesia in July 2010 using the Caldwell-Luc approach for the left maxillary sinus. Since the tumor was large and hard, like scar tissue, it was difficult to extract en bloc. No malignancy was found when a part of the lesion was submitted to the pathology department for rapid diagnosis using the fresh-frozen section procedure. We excised the tumor, which had extended to the ethmoid and sphenoid sinuses, to the maximum possible extent. The resected tumor was a white, fibrotic mass surrounded by a poorly defined capsule (Fig. 3). Histological analysis revealed heavily collagenized stroma with spindle cells with bland nuclei and elongated vessels, suggestive of AF (Fig. 4A and B). Immunohistochemical examination of the excised tumor showed that it was positive for vimentin and smooth muscle actin (SMA) and negative for S-100 protein and CD34 (Fig. 5A and B). Two and a half years after surgery, no evidence of recurrence or local metastasis was observed (Fig. 6A and B).

3. Discussion

Fibromatosis is an aggressive fibrous tumor, which is highly locally invasive and affects skeletal muscles without distant

☆ AsianAOMS: Asian Association of Oral and Maxillofacial Surgeons; ASOMP: Asian Society of Oral and Maxillofacial Pathology; JSOP: Japanese Society of Oral Pathology; JSOMS: Japanese Society of Oral and Maxillofacial Surgeons; JSOM: Japanese Society of Oral Medicine; JAMI: Japanese Academy of Maxillofacial Implants.

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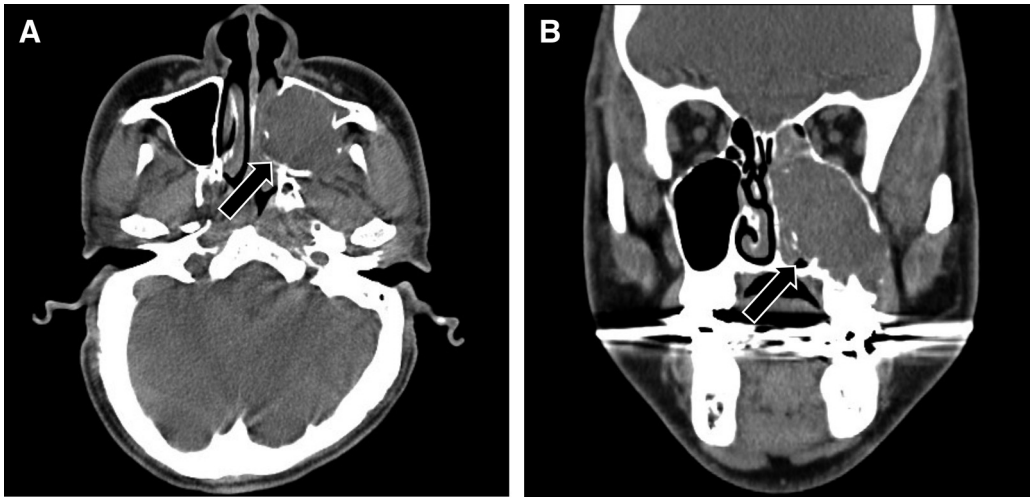

Fig. 1. (A and B) Axial and frontal computed tomography reveals an irregularly shaped, protruding mass, measuring 40 mm × 32 mm × 30 mm, in the left maxillary sinus. It expanded and eroded the posterior walls of the maxillary sinus and the alveolar bone (arrow).

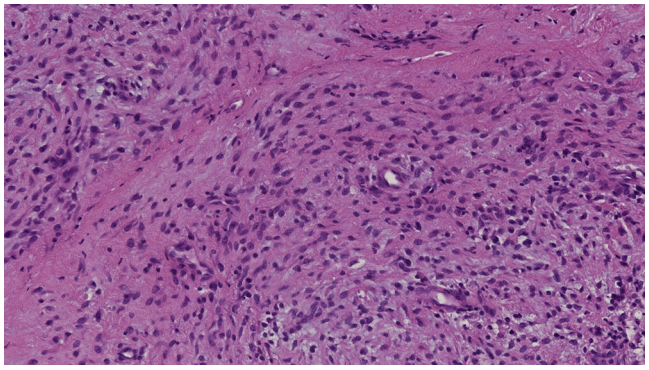

Fig. 2. Pathological examination of the biopsy specimen revealed hyperplasia of the fibrous tissue and the presence of inflammatory cells (hematoxylin–eosin stain, original magnification 200×).

metastasis. It can grow into and destroy adjacent normal tissues, including the vital structures or organs [4]. In terms of clinical symptoms, AF is painless in most cases and is often discovered incidentally on radiography or due to facial swelling. Radiography shows unilocular or multilocular radiolucent lesions with relatively clear borders, although diffuse bone resorption with unclear borders can also be observed [5].

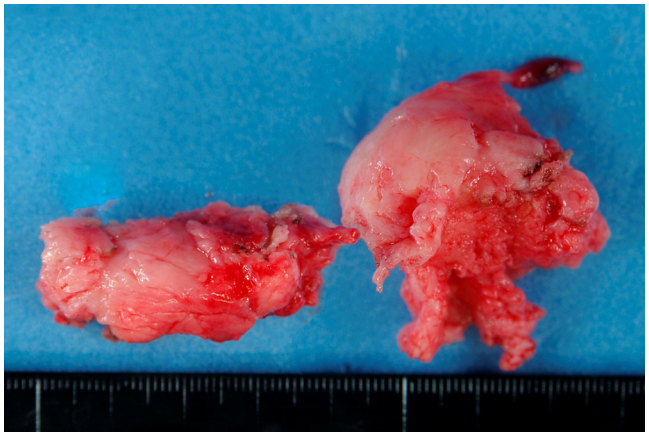

Fig. 3. Macroscopically, the tumor was a white, fibrotic mass with poorly defined margins.

AF is most common in patients between puberty and the age of 40 years, with a peak incidence between the ages of 25 and 35 years [1]. AF often presents in the upper arm, forearm, thigh, hip, shoulder, head, or neck. In the head and neck region, the tumor often occurs in the neck, mandible, or mastoid process, accounting for approximately 10–20% of all AF cases [6]. Between 1968 and 2008, 179 cases of AF of the head and neck were published. The male to female ratio was 91:82, with a mean age of 16.87 years, and 57.32% of cases were found in patients less than 11 years of age. The mandible and the neck were the most common tumor sites. AF rarely occurs in the maxillary sinus, accounting for approximately 8% of all AF in the head and neck region [3].

Histopathologically, the tumor process consists of elongated, slender, spindle-shaped cells of uniform appearance, surrounded and separated from one another by abundant collagen, with little or no cell-to-cell contact. The cells lack hyperchromasia or atypia, and cellularity varies within the lesion. The constituent nuclei are small, pale staining, and sharply defined, with 1–2 min nucleoli [1]. AF most closely resembles fibrosarcoma and reactive fibrosis [1]; therefore, it is very important to distinguish between these conditions. In contrast to fibromatosis, fibrosarcoma usually displays greater cellular uniformity and the cells exhibit a more consistent sweeping fascicular growth pattern. The nuclei are more hyperchromatic and atypical and have more prominent nucleoli than those found in fibromatosis [1]. It can be difficult to distinguish AF from reactive fibroblastic proliferations, following injuries such as trauma, minor muscle tear, or intramuscular injection, by examination of an incisional biopsy sample. In this case, the representative tumor tissue was only obtained after excision of the entire tumor; 2 biopsy specimens obtained before tumor excision revealed only proliferation of spindle-shaped fibroblastic cells with inflammatory granulation tissue. Thus, excision of the tumor was necessary to confirm the AF diagnosis.

According to reports, immunohistochemical analysis shows positive staining with vimentin in 100% of AF patients, with SMA in 78% of patients, with actin in 30% of patients, with S-100 protein in 8% of patients, and with desmin in 0% of patients [7]. Although immunohistochemistry is not necessary to confirm an AF diagnosis, it can aid in differentiating AF from other diseases with similar histological features.

The standard treatment for AF is wide surgical resection because AF grows relatively quickly, is invasive to surrounding tissues, has unclear borders, and a high frequency (20–70%) of local recurrence [8]. However, in the stomatognathic region, the clinical course is