Fibromatosis of the Submandibular Region

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Fibromatoses are benign tumors that can occur anywhere in the body. They are locally aggressive and tend to recur; they also cause considerable morbidity, particularly when they arise in the head and neck. Fibromatosis in the submandibular region is rare. Here, we present a case of fibromatosis of the submandibular region in a 42-year-old female who presented with a right submandibular mass. The patient underwent complete excision of the fibromatosis, showed negative margins and had no adjuvant therapy. Her recovery was uneventful, and there was no recurrence or neurological deficit 1 year after the operation. [*J Chin Med Assoc* 2009;72(9):492–494]

Key Words: fibromatosis, head and neck neoplasm, submandibular region

Introduction

Fibromatoses are benign fibrous tumors that arise from the connective tissue of muscle, overlying fascia, periosteum or aponeurosis.[1–5] Although there is no malignant or metastatic potential, fibromatoses are locally invasive, with a high propensity for recurrence, especially when they occur in the head and neck region, due to the proximity of vital structures.[1,6] Only 12–15% of all fibromatoses occur in the head and neck region, 26% of which originate in the submandibular area.[1,6,7] Here, we present a case of fibromatosis of the submandibular region and discuss the etiology, clinical presentation, pathology and various treatment modalities.

Case Report

A 42-year-old woman complaining of a right submandibular mass that had been growing for 2 years was referred to our department. One year earlier, she had undergone a biopsy of the mass at another hospital, and the pathology report showed only fibrosis. However, the mass had continued to gradually increase in size. Physical examination revealed a very firm, fixed, nontender mass in the right submandibular area; there were no other otolaryngological findings or neurological abnormalities. T1-weighted gadolinium-enhanced magnetic resonance imaging (MRI) revealed an enhanced mass measuring approximately $5.1 \times 3.3 \times 5.1$ cm in size in the right submandibular area, with invasion of the right mylohyoid muscle causing elevation of the floor of the mouth (Figure 1). In addition, the right submandibular gland was partially encased and was displaced posteriorly. Computed tomography (CT) showed a lobulated tumor in this area without a clear fat plane to the surrounding tissues. Blood test results were within normal limits.

The patient underwent wide excision of the mass under general anesthesia. The mass, which was cystic and solid with yellowish fluid content, was dissected from the surrounding structures and removed completely. The surgical specimen, which weighed 86 g and measured $9.5 \times 5.0 \times 4.0$ cm, consisted of a single tumorous mass plus the submandibular gland (Figure 2). Histopathological examination revealed fibromatosis characterized by proliferating fibroblasts with positive immunoreactivity for vimentin; it was negative for CD34, S-100 and smooth muscle actin (Figure 3). Collagen deposition and focal ossification were also identified. There was no pleomorphism or mitosis in the cells. The surgical margin was tumor-free, and the patient had no adjuvant therapy. Her recovery was uneventful, and there were no neurological deficits. Follow-up at 1 year showed no recurrence, and the patient remains under careful follow-up.

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E-mail: yaohshiang@gmail.com • Received: January 23, 2009 • Accepted: June 15, 2009

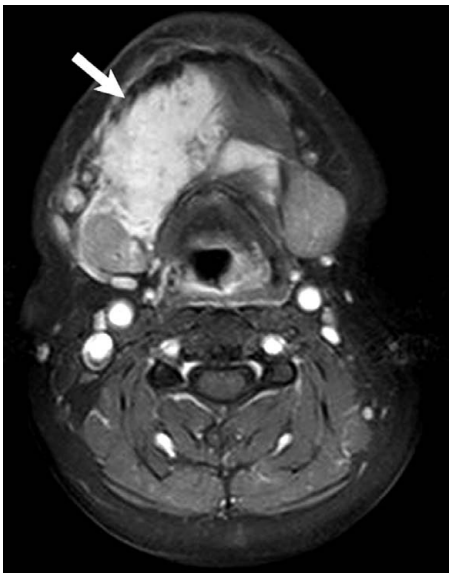

Figure 1. T1-weighted gadolinium-enhanced magnetic resonance imaging of the patient's neck shows an enhanced mass in the right submandibular region (arrow). The right submandibular gland is partially encased and posteriorly displaced.

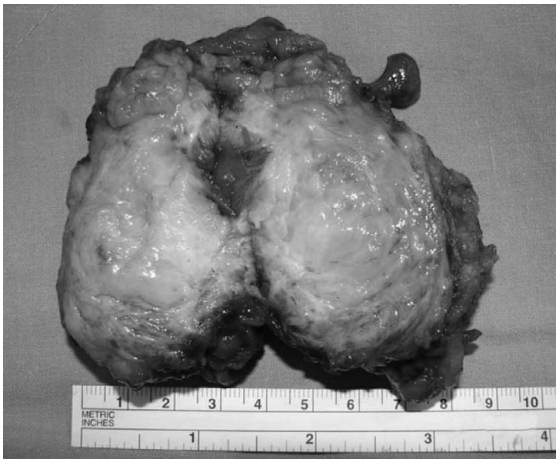

Figure 2. The surgical specimen after excision of the mass and right submandibular gland. The incised surface is trabecular in appearance.

Discussion

Fibromatoses, also called desmoid tumors, are benign fibrous tumors that arise from the musculoaponeurotic structures in the body.[1,2] They are locally invasive, with a propensity for recurrence after resection, but they are not prone to metastasis nor do they have malignant potential.[1,7] Clinically, these tumors are intermediate-grade neoplasms with biological behavior that falls somewhere between that of benign fibromas and well-differentiated fibrosarcomas.[3,4] Owing to their complex characteristics, they have been referred to as

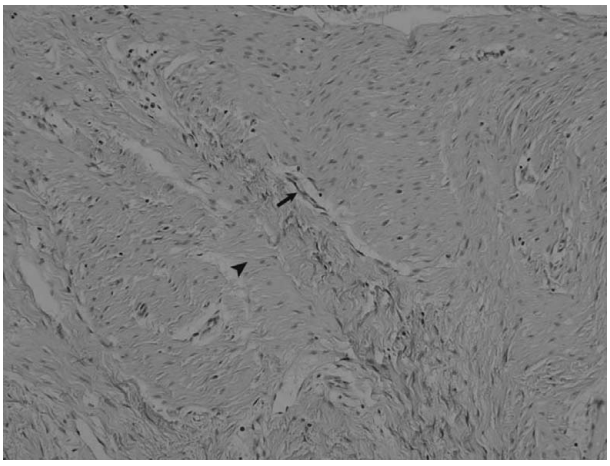

Figure 3. Photomicrograph shows the interlacing bundles of fibroblasts (arrow) separated by variable amounts of collagen (arrowhead) (hematoxylin & eosin, 200×).

desmoplastic fibromas, desmomas, aggressive fibromatoses and desmoid tumors.[1,3] Researchers have proposed several systems for categorizing fibromatoses, the most widely accepted being that described by Enzinger and Weiss.[1,7] Their system subdivides fibromatoses into 2 types, superficial and deep. Superficial fibromatoses encompass palmar, plantar, penile and knuckle pad lesions. Deep fibromatoses include the extra-abdominal, abdominal and intra-abdominal subtypes. Head and neck fibromatoses belong to the extra-abdominal subtype of deep fibromatoses.

Fibromatoses are relatively uncommon tumors, with an estimated incidence of approximately 2–4 cases per million patients per year.[2,8] Only 12–15% of all fibromatoses occur in the head and neck area, with 40–85% of these cases arising in the neck.[1,6] The cervical and supraclavicular regions are the most common sites for head and neck fibromatoses, followed by the face, oral cavity, scalp, paranasal sinuses, and orbital region.[1,3,9] This tumor usually manifests as a painless, firm, slow-growing, deep-seated mass that is fixed to the underlying soft tissues.[3] It can spread through fascial planes and invade surrounding structures, causing pain, dysphagia, respiratory distress, proptosis and epistaxis. When airway or major vessels are involved, there may be life-threatening consequences.[3,4,6]

The etiology of fibromatosis remains unclear. Physical, endocrine and genetic factors have all been hypothesized to play a role in the origin of fibromatosis; however, no compelling evidence supports any single factor.[1–5,8] Many physicians have reported patients with a history of local trauma preceding the development of fibromatosis, such as previous surgery or blunt force injury. This has led to the hypothesis that immature fibroblasts engaged in healing begin uncontrolled

proliferation that leads to tumor formation. However, there have not been enough patients with head and neck fibromatoses after local trauma to confirm this theory.[1,3] In particular, the patient reported here had no history of local trauma. Endocrine disturbance may play a part in the formation of fibromatoses, because some lesions have been found to grow during puberty or pregnancy and then regress following menopause or radiation-induced castration; some tumors have shown low levels of estrogen receptors.[1,3,4,9] In addition, an inherited pattern, inappropriate gene expression and chromosome abnormalities have been reported in some patients with fibromatoses.[1,3]

Grossly, fibromatoses are firm, rubbery, grayish-white lesions that tend to infiltrate surrounding tissues so that the border of the tumor is ill-defined.[1–4] Histologically, the tumors are composed of uniform, spindle-shaped fibroblasts arranged in fascicles with an abundant collagenous component.[1–3] Mitotic figures are rare, and there are no atypical cells or hyperchromatic nuclei.[1,3] The degree of cellularity can vary from region to region, however, and the tumor often invades adjacent structures.[1] Owing to these clinical and pathological characteristics, a fibromatosis may be confused with a reactive fibrosis, myxoma, nodular fasciitis, keloid, fibrous hamartoma, neurofibroma, neuroma, rhabdosarcoma or fibrosarcoma.[1–4,9]

At present, fibromatoses are primarily treated by complete excision with a clear margin.[1,5] Several studies have reported a correlation between a positive tumor margin and the incidence of local recurrence.[1,7,8] When the tumor occurs in the head and neck region, the proximity of vital structures, the complexity of the fascial planes, and the tumor's aggressive behavior often make it difficult to achieve negative margins. Thus, recurrence rates in this region are higher than in other locations.[3,7,9] Preoperative CT or MRI is essential to identify tumor borders and for planning the surgical approach to ensure extirpation of the tumor.[3,6] On CT, the lesion is usually hyperattenuated and mildly enhanced after the administration of contrast medium. MRI will show fibromatosis as a multilobulated lesion of heterogeneous signal intensity. The tumor is slightly hypointense on T1-weighted imaging and hypointense to hyperintense on T2-weighted imaging, with enhancement after the administration of gadolinium.[2] In patients who are not good surgical candidates due to medical or technical concerns, radiation or pharmacologic therapy should be considered. Although the efficacy of radiotherapy remains controversial, there are some reports of good local control rates using radiotherapy as either adjuvant or primary therapy.[1,5,6] Radiotherapy should thus be considered if there is gross residual tumor or positive margins after resection, if a tumor recurs, or if primary excision is not feasible.[1,3,5] Other treatments, such as anti-estrogen therapy, chemotherapy, steroids, nonsteroidal anti-inflammatory drugs, ascorbic acid or castration, have been attempted with variable success.[1–3,7,9] These alternative modalities may be tried if surgery and radiation fail.

In conclusion, although fibromatosis is a histologically benign neoplasm, treatment is challenging due to its aggressive clinical behavior and propensity for local infiltration with a tendency for recurrence. When the tumor occurs in the head and neck region, it can cause significant morbidity and is extremely difficult to address. The most effective treatment is complete excision; other therapies, including radiation and drug treatment, may be considered if surgery is abortive or not feasible. Due to the high incidence of recurrence, close follow-up is essential.

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