Aggressive Fibromatosis Arising in Temporal Muscle

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Fibromatosis of the temporal region is extremely rare. It has also been referred to as extra-abdominal desmoid which originates from muscle or fascia and it is a highly recurrent tumor. This paper presents a case of temporal muscle fibromatosis and a brief review of the pertinent literature.

Key Words: Aggressive fibromatosis, Temporalis muscle.

Aggressive fibromatosis, or desmoid tumor, is known as a tumor which frequently arises from abdominal musculature. However, extra-abdominal desmoid tumors are relatively rare. Historically, Ben net (1849) reported fibrinoid carcinoid growth in the thigh, upper extremity and parotid region. Paget (1856) reported a patient with this tumor in the arm and Nicolas (1923) was the first to define extra-abdominal desmoid tumor. Since then, Musgrove (1948) defined the macroscopic and microscopic criteria necessary for diagnosis of this tumor and defined desmoid tumor as a fibrous tissue neoplasm arising from musculoaponeurotic structures in any area of the body.

Recently, Masson (1966) reported on the clinical and pathological findings of head and neck area fibromatosis. Among the previous reports on fibromatosis of the head and neck area, lesions involving the temporal muscle are extremely rare. This paper presents a case of temporal muscle fibromatosis and a brief review of the literature.

CASE HISTORY

A 48 year old female was first seen on Feb. 10, 1982 with a mass in the right temporal area but without specific subjective symptoms (Fig. 1). She had been aware of a flat, hard, firm and ill-defined, tumor mass for the past 9 months. Subsequently, the tumor has grown slowly and she could no longer open her mouth fully.

![Fig. 1. Protruding mass of the right temporal area. Pre-operative appearance.](image1)

![Fig. 2. Mandibular deviation to right is demonstrated when the mouth is opened.](image2)
Physical examination demonstrated a non-tender, fixed, and hard mass measuring about 5 cm in diameter. The lesion was located in right temporal area. Mouth opening was limited with shifting of the jaw to the involved side (Fig. 2). There was no bony invasion of the tumor into the temporal bone on radiologic examination. On C-T scan, the tumor mass encompassed the zygomatic fossa.

**Operation and Surgical Findings**

Incisional biopsy identified the tumor mass as an aggressive fibromatosis or a desmoid tumor. Through a hemiconary and preauricular incision, dissection was performed at the subgaleal plane in temporal region and subcutaneously around the zygomatic arch area in order to protect facial nerve. The zygomatic arch itself was resected for easy removal of the tumor below the zygomatic arch bone. The tumor had invaded the anterior half of the temporalis muscle and extended close to the coronoid process. Complete resection of the tumor was achieved (Fig. 3). The resected zygomatic arch bone was returned to its original position and secured with wiring. The incision was closed routinely. The post operative course was uneventful. Moderate depression in the right temporal fossa area persists and there has been no tumor recurrence for the past 3 years (Fig. 4).

**Pathological Findings**

**Gross:** The specimen consisted of an irregular mass measuring 8×4.5×4cm. It was relatively well demarcated from the posterior part of the specimen containing normal temporal muscle. The lesion was gray, white and pink in color without encapsulation. The consistency was rubbery to firm. The cut surface demonstrated a glistening, white, coarsely trabeculated or irregularly whorled fibrinoid surface. There was a necrotizing cystic cavity in the center of the tumor mass (Fig. 5).
Microscopic: The tumor was composed of extremely mature fibrocyte. There was however, some difference in cellularity in different areas of the lesion. In the hypocellular area, it was composed of mature fibrous tissue consisting of uniform spindle cells and abundant, broad band-like collagen fibers, resembling a hypertropic scar. In the hypercellular area, it was composed of fibroblasts and at the periphery of the tumor, it showed fibroblastic proliferation in a rather edematous matrix which insinuated between the muscle groups and fibers. A few giant cells derived from regenerating striated muscle were present. Scattered capillaries were seen and atypical mitotic figures were not present. There were a small number of inflammatory cells, predominantly lymphocytes, in parts of the tumor (Fig. 6, Fig. 7).

DISCUSSION

Aggressive fibromatosis is the highly recurrent condition. Batsakis (1979) called it sarcoma differentiated from keloid because its microscopic finding was so variable. Fibromatosis is a tumor which originates from muscle or fascia of any area of the body. Desmoid tumors arising in the anterior rectus sheath are microscopically the same as extra-abdominal desmoid tumor elsewhere (Masson, 1966; Batsakis, 1979). However, those arising in the head and neck region are more aggressive (Convery, 1960; Batsakis, 1979). Stout (1960) differentiated the lesions into two types (cellular desmoid tumors as fibrosarcoma are differentiated from hypocellular desmoid tumors as fibromatosis). Batsakis (1979) therefore postulates that this is not a true neoplasm, but that fibromatosis is a rational terminology for the entity.

From a clinical stand point recurrence after surgical excision is the problem. This is usually due to extensive local invasion. Prognosis, therefore, is variable according to the area of the tumor and how close it exists to the vital organs.

Age at onset of aggressive fibromatosis is variable and females are affected more often than males. No endocrine abnormalities however, have been identified in these tumor patients (Masson, 1966; Enzinger, 1967). Masson has reported that of lesions appearing in the head and neck region, the neck is involved in about 85% of the cases and while the lesions appeared in separate locations on the scalp and face in the remaining 15% (Masson, 1966). In our review of literature only one case of temporal fibromatosis was reported (Masson, 1966) and that lesion recurred three years after surgical excision. At that time additional excision and irradiation were done. In many reports the tumor appeared to arise in the operative sites or local trauma even though etiologic factors responsible for fibromatosis were difficult to assess (Enzinger, 1967; Das Gupta, 1969). Recurrence rates for the tumor were reported as 57% by Enzinger (1967), 70% by Masson (1966) and 25% by Das Gupta (1969). Enzinger (1967) also reported two instances of spontaneous regression of desmoid tumors or aggressive fibromatosis. Wide surgical excision, therefore, is the treatment of choice. And role of irradiation is unclear but may also be helpful.

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