Fibromatosis presenting as acute mastoiditis: A case report

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Abstract
We describe the case of a middle-aged man who presented with manifestations of acute mastoiditis caused by fibromatosis of the mastoid region. A lesion of the right mastoid bone had eroded its wall and extended toward the middle and posterior cranial fossae. The macroscopic and microscopic appearance of an excised portion of the lesion established the diagnosis of mastoid fibromatosis. After a more detailed work-up, a second procedure involving extensive removal of the tumor was performed, and the diagnosis was confirmed. The patient's postoperative period was uneventful, and he showed no evidence of recurrence during 3 years of follow-up.

Introduction
The term fibromatosis of the head and neck refers to a controversial, poorly defined group of fibrous proliferative lesions. This designation encompasses a wide histologic range of lesions that exhibit a deceptively harmless microscopic appearance. Clinically, they tend to grow, to infiltrate neighboring tissues, and to recur. The management of such lesions demands an accurate microscopic interpretation and expert surgical management.1

Fibromatosis of the head and neck has not been documented very often in the literature, and many issues concerning its identity and biologic behavior are not yet fully defined. The histologic complexity of fibrous tumors has defied efforts to establish a unifying concept of pathology, and this situation continues to hinder prognosis by pathologists and treatment by surgeons.

In this article, we describe a rare case of fibromatosis of the mastoid bone, and we discuss the unique characteristics of this interesting clinical entity.

Case report
A 56-year-old man was referred to the Emergency Department of our hospital for evaluation of purulent otorrhea.

The patient was schizophrenic and permanently housed in a psychiatric hospital.

Clinical examination revealed edema and redness in the right mastoid area and protrusion of the right auricle. On palpation of the skin, fluctuation was observed. Otoscopy detected swelling of the skin of the posterior canal wall and a tiny amount of purulent bloody fluid. The tympanic membrane was only partially visible, but it appeared to be intact and without any sign of infection. Findings on the remainder of the clinical examination were unremarkable.

Findings on laboratory testing were normal except for a high level of polymorphonuclear leukocytes and chronic anemia. Cultures of the draining ear fluid were obtained immediately upon admission, and combined antibiotic therapy with ticarcillin/clavulanic acid, metronidazole, and netilmicin was started. Computed tomography (CT) of the brain and temporal bone demonstrated an extensive opacification of the right mastoid process and indications of erosion of the mastoid surface that created an opening to the posterior cranial fossa at the level of the sigmoid sinus (figure 1).

Surgical exploration of the lesion was scheduled, and the patient underwent a mastoidectomy. Intraoperatively, a whitish, soft erosion of the right mastoid wall was found; the erosion involved the entire mastoid area and extended into the posterior and middle cranial fossae without distinct limits. The sigmoid sinus was thrombosed. Segments of the mass were excised and sent for microscopic evaluation.

In light of the unexpected degree of tumor extension and following a neurosurgical consultation, we terminated the operation in order to plan an appropriate procedure based on the biopsy results and the findings of imaging studies ordered to accurately estimate the tumor's margins.
The patient’s postoperative period was uneventful, and he exhibited only a low fever. Cultures grew both aerobic (*Staphylococcus aureus*) and anaerobic (*Bacteroides fragilis*) bacteria; as both are sensitive to ticarcillin/clavulanic acid and metronidazole, these agents were continued intravenously.

According to the histology report, the surgical specimen was made up of three small fragments of soft, whitish tissue; the largest of these fragments was 1 cm in its greatest dimension. Microscopically, the lesion contained elongated spindle-shaped cells that were uniform in appearance and separated from each other by abundant collagen. Some of these cells had atypical or hyperchromatic nuclei. Microhemorrhages and focal aggregates of lymphocytes and plasma cells were also seen (figure 2). At the periphery of one of the fragments, spicules of normal bone were identified. On immunohistochemical staining, the tumor cells were positive for vimentin and actin and negative for desmin and S-100 protein. Their overall appearance was consistent with fibromatosis.

Magnetic resonance imaging (MRI) of the brain revealed involvement of the right petrous wall (soft-tissue edema was present in this area) and further extension into the periphery of the right cerebellar hemisphere (figure 3). The image of the right sigmoid sinus was not clear. Arteriography identified mild hyperemia from small arterial branches at the area of the right petrous pyramid; these branches seemed to originate in the precavernous portion of the right internal carotid artery. Color Doppler ultrasonography of the neck vessels detected an old thrombosis in the right internal jugular vein. Revascularization, however, was adequate. The right external and internal carotid arteries were normal.

A second operation was performed 1 week after the first, this time with neurosurgical assistance. By using the same incision, we were able to attain wider exposure of the lesion in the middle and the posterior cranial fossae. Although we again terminated the operation without finding the distinct margins of the tumor (because of the broad extension of the mass and the infiltration of the underlying dura mater), we were able to achieve a near-total removal of the tumor. The specimen was sent for histologic study, and the results were the same, thereby confirming the diagnosis.

Again, the patient’s postoperative period was uneventful. Clinical findings and laboratory results were normal. On postoperative day 9, antibiotic therapy was stopped and the patient was discharged in good general health and without any medication or specific instructions. He showed no evidence of recurrence during 3 years of follow-up.

**Discussion**

Fibromatosis, often called *extraabdominal desmoid*, refers to the formation of locally aggressive but histologically benign fibroblastic lesions that sometimes arise in the head and neck region. These lesions are confined to the musculature and the overlying aponeuroses. They occasionally extend along the fascial plane or infiltrate the overlying subcutaneous tissues. In their most aggressive forms, these lesions involve the periosteum; aggressive lesions have a propensity to invade and erode adjacent bone and to encase vital structures in the head and neck. Fibromatosis of the head and neck accounts for 11 to 12% of all cases of extraabdominal desmoid; of these cases, 85% involve the neck (especially the supraclavicular fossa) and 15% involve the face and scalp. Fibromatosis is most common in Caucasians, and there appears to be a high female preponderance.

Fibromatotic lesions are usually more aggressive than most other benign neoplasms. Trauma and inflammation may play a role in the pathogenesis of these lesions, although most reports of large series have failed to prove such a relationship. In our patient, the pathogenic factor might...
have been the chronic middle ear inflammation. The clinical presentation and course are not unlike those of desmoid tumors found in other locations. Patients with fibromatosis generally present with a painless, poorly defined, enlarging, gray-white mass that has usually been present for less than 1 year. The mass is often fixed to underlying deep muscles or to bony structures. These lesions tend to grow much faster in the head and neck than they do in other extraabdominal sites; infection may play some role in the rapid growth. A high rate of recurrence is one of the most common characteristics of the various types of fibromatotic lesions, especially those in the head and neck.

The differential diagnosis is quite difficult. Very often, an inflammatory or malignant process is suspected in head and neck lesions. A high index of suspicion is important, but other types of fibromatosis and other types of lesions must also be considered, including pseudosarcomatous fasciitis, fibrous dysplasia, ossifying fibroma, or the highly malignant fibrosarcoma. In our patient, we initially suspected a malignancy because of the invasive nature of the tumor and the erosion of the adjacent bone, through which the extension to the middle cranial fossa occurred.

The diagnosis is usually made by incisional biopsy; radical excision is usually not warranted until the diagnosis has been firmly established and adequate treatment has been planned. On macroscopic examination, the cut section is firm, rubbery, and gray-white. The border is ill defined because of the lesion’s tendency to infiltrate surrounding structures; some lesions encase neural and vascular structures and erode bone. Distinguishing among the various types of fibromatosis is quite difficult, and we must often rely on the anatomic location and clinical behavior of the disease to make the diagnosis.

On microscopic examination of a purely fibrous tissue lesion, a full range of escalating stages can be seen, beginning with the normal reparative processes and scar formation and progressing to fibroma, keloid, pseudosarcomatous fasciitis, fibromatosis, differentiated fibrosarcoma, and finally to undifferentiated fibrosarcoma. Fibromatosis occupies an intermediary position, and microscopic sections contain markedly cellular lesions of mature proliferative fibroblastic tissue, with infrequent mitoses and occasional atypia, but no frank anaplasia. The morphologic pattern is quite uniform throughout, but the degree of cellularity may vary from area to area and from tumor to tumor. The collagenous component is abundant but mature. The presence of inflammatory cells may indicate a reactive process rather than a tumor.

The propensity of some lesions to invade adjacent muscle, adipose tissue, and bone may lead to an erroneous diagnosis of fibrosarcoma. The reliability of a purely histologic diagnosis of a fibrous tissue tumor is questionable because of the subtle differences between benign and malignant tumors. The patient’s clinical history therefore plays a crucial role in establishing the correct diagnosis. In our patient, the typical histologic features of fibromatosis allowed us to easily arrive at the diagnosis. However, our case was unique; our literature surveillance turned up reports of various mastoid lesions, but we did not find a case similar to ours. In one of these cases, the lesion extended through the sphenoid sinus into both pterygoid recesses, invaded the cavernous sinus, and involved the floor of the sella, the clivus, the petrous temporal bone, and the mastoid. However, the origin of this tumor could not be defined because it had been diagnosed at an advanced stage.

The treatment of choice for fibromatosis is wide surgical excision with an adequate margin. However, because the vital and complex anatomy of the head and neck poses technical difficulties, many head and neck surgeons hesitate to perform a radical procedure to treat a benign disease. Fortunately, inoperable and recurrent fibromatosis can be successfully treated with radiation therapy, which has been shown to result in a complete and long-term regression. Radiation therapy was not considered necessary for our patient because his tumor was successfully removed, but it was held in abeyance pending a recurrence.

References

Figure 3. MRI shows the lesion (arrow) in the right mastoid.