

## Neurooncological Observation

# Benign fibrous histiocytoma of the pterygopalatine fossa with intracranial extension

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## Summary

A very rare case of fibrous histiocytoma arising in the pterygopalatine fossa with intracranial extension is described. Despite the histologic absence of nuclear pleomorphism, the tumor rapidly recurred after complete surgical resection. The patient, a 45 year old male, died shortly thereafter. The aggressive nature of our patient's tumor confirms previous observations that an aggressive radiographic appearance has prognostic value when dealing with skeletal and soft tissue tumors. The benefit of multimodal therapy has not been established in these rare head and neck lesions. In the subset of fibrous histiocytomas that invade bone, however adjunctive treatment with radiation and or chemotherapy may be appropriate.

**Keywords:** Benign; fibrous; histiocytoma; intracranial; skull base.

## Introduction

Fibrous histiocytic tumors comprise a group of lesions with certain overlapping morphologic features but with variable origin and biologic behavior [12]. Within this group, benign fibrous histiocytomas usually arise in superficial soft tissue, but can originate in deep soft tissue, parenchymal organs, or bone. Cellular variants of benign fibrous histiocytoma are rare, and are thought to be more locally aggressive than conventional benign fibrous histiocytomas, but do not metastasize. Malignant fibrous histiocytomas are characterized by marked nuclear pleomorphism, a high mitotic rate, necrosis, and fully malignant biologic behavior.

Benign and malignant fibrous histiocytomas are rare in the head and neck, but cases of fibrous histio-

cytoma with malignant or benign types arising from the calvaria and pericranium with extension into the brain [2, 5, 13] or isolated intracerebral occurrence [9] have been described. To the best of our knowledge, skull base origin, pterygopalatine fossa in particular with intracranial extension has not been described previously.

## Case report

A 45-year-old male presented with a two months history of progressive pain over the right maxillary tuberosity and right sided facial numbness along with impaired hearing on the same side. His clinical exam demonstrated V2 and V3 hypesthesia, serous otitis media, and decreased mandibular excursion on the right. Fiberoptic evaluation revealed mucoid discharge from the right sphenoid ostium. Computed tomography and magnetic resonance imaging demonstrated a large (5.3 × 4 × 3 cm) mass lesion of the infratemporal fossa, eroding the posterior and lateral walls of the maxillary sinus and the floor of the temporal fossa. Intracranial extension of tumor had elevated the right temporal lobe without evidence of brain invasion. The lesion also extended into the right sphenoid sinus (Fig. 1).

The lesion was biopsied via a transantral approach to the pterygopalatine fossa. Histologic findings were consistent with a benign fibrous histiocytoma.

The patient subsequently underwent a right frontotemporal craniotomy and skull base approach to the middle fossa, cavernous sinus and infratemporal fossa. Concurrently, right partial maxillectomy with transantral ethmoidectomy and sphenoidotomy were performed. After complete surgical exposure, the lesion appeared to originate in the pterygopalatine fossa, filling the infratemporal fossa, and entering the middle cranial fossa via the foramen rotundum. The lesion was resected extradurally and negative histopathologic margins

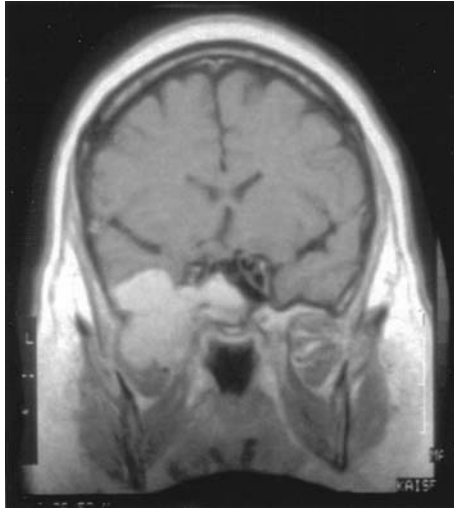


Fig. 1. Preoperative T1-weighted post-gadolinium magnetic resonance image demonstrating the extent of the original tumor. Bony destruction includes posterior/lateral maxillary sinus and floor of the temporal fossa

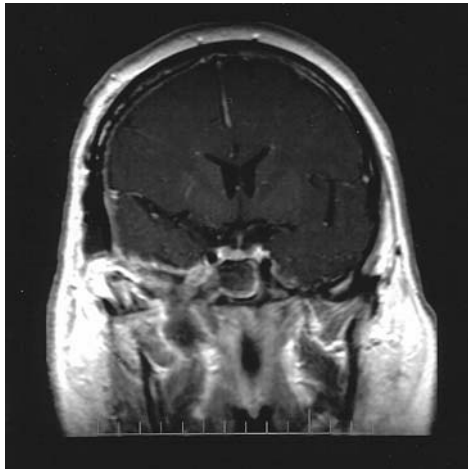


Fig. 2. Postoperative T1-weighted post-gadolinium magnetic resonance image showing gross total resection of the tumor that reveals complete resection of the tumor. A small enhancing residuum is visible inferolateral to the cavernous sinus

were obtained. Postoperative MRI evaluation confirmed a near-total removal with a small residual enhancement inferolateral to the cavernous sinus (Fig. 2).

Final histopathologic examination of the tumor demonstrated a proliferation of fibrohistiocytic cells with a regular vascular pattern (Fig. 3). Numerous benign-appearing giant cells were scattered through the lesion (Fig. 4). Areas of the tumor were somewhat more cellular with spindled cells arranged in short fascicles. Mitoses were variable, but averaged fewer than 1 per 10 high power fields, and there was minimal nuclear pleomorphism (Fig. 4). The tumor showed focal positive immunohistochemical staining for a marker of macrophages (CD68), but was negative for markers of endothelial, epithelial, and smooth muscle origin (CD34, epithelial membrane antigen (EMA), cytokeratin, smooth

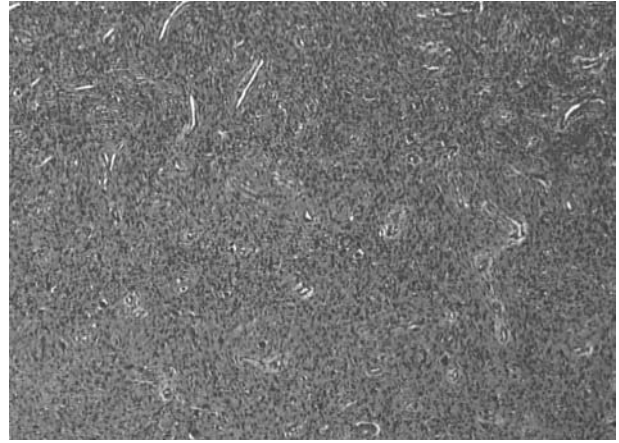


Fig. 3. Relatively low magnification of the tumor, demonstrating spindled, fibrohistiocytic cells and a regular vascular pattern without necrosis (Hematoxylin and eosin, 25 $\times$ )

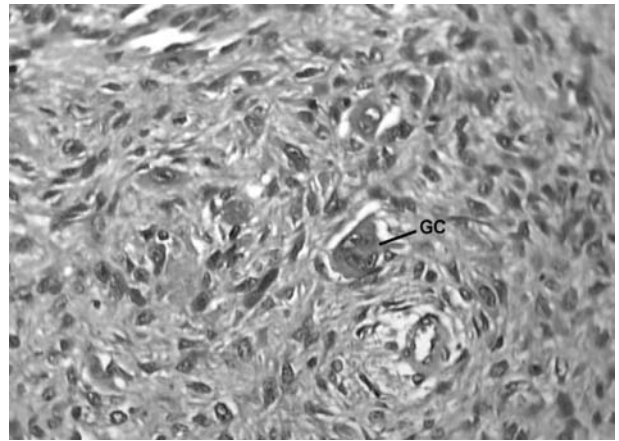


Fig. 4. Higher magnification showing a benign giant cell (GC) surrounded by fibrohistiocytic cells with minimal nuclear pleomorphism. Although not shown in this field, occasional mitoses were present (hematoxylin and eosin, 100 $\times$ )

muscle actin, factor XIII A and factor VIII). These findings were thought to be most consistent with a benign fibrous histiocytoma, although a lesion of low-grade malignancy could not be excluded. Areas of increased cellularity resembled the so-called cellular variant of fibrous histiocytoma.

The early postoperative period was uneventful with significant resolution of preoperative hypesthesia and complete resolution of preoperative pain. However, three months after surgery, the patient developed a right-sided visual loss with concomitant ipsilateral nasal obstruction along with an enlarging mass in the area of the right zygomatic arch. Repeat MRI revealed a larger mass extending from the right middle fossa to the infratemporal fossa, right maxillary and sphenoid sinuses. The lesion had also invaded the right cavernous sinus, encircling the intracavernous internal carotid artery; posteriorly and inferiorly and filling the nasopharynx (Fig. 5).

Despite radiation therapy with a total dose of 50 Gray, the lesion progressed and the patient died before attempts at palliative chemotherapy were instituted.

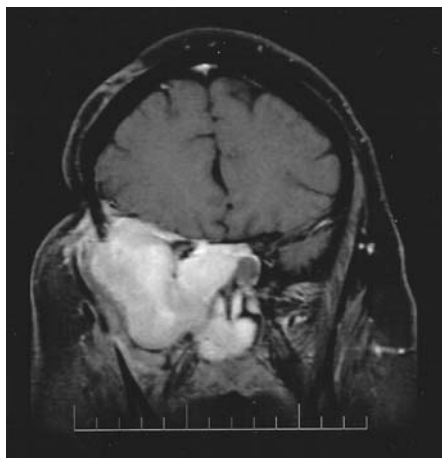


Fig. 5. T1-weighted post-gadolinium magnetic resonance image three months after resection, demonstrating massive recurrence involving the infratemporal fossa and cavernous sinus with intracranial extension into the middle fossa

## Discussion

Fibrohistiocytic lesions are a relatively ill-defined family of mass lesions of largely unknown pathogenesis and diverse biologic behavior. While some are thought to be reactive (e.g. xanthofibroma), lesions generally called “benign fibrous histiocytoma” or “malignant fibrous histiocytoma” are true neoplasms. The term “fibrous” or “fibrohistiocytic” is now used in a descriptive sense, rather than suggesting a cell of origin. Fibrohistiocytic tumors of the head and neck are rare. Lesions that do occur in this region usually originate in the maxilla or mandible [1, 8, 11].

Fibrohistiocytic tumors are usually classified as benign or malignant, with the distinction based largely on nuclear pleomorphism, overall cellularity, mitotic rate, and the presence or absence of necrosis [4, 11, 12]. Borderline lesions have been described, however, are often difficult to classify [10, 11]. Despite histologic distinction, both “benign” and “malignant” lesions can exhibit aggressive clinical behavior, so, some authors feel that the term “benign” should not be applied to any fibrohistiocytic tumor located in the deep soft tissue [3]. This may be especially true in the deep tissues of the head and neck. For example, Thompson and Shear [11], reviewed a series of 70 oral and maxillofacial fibrous histiocytoma cases, and pointed out that 91% of histologically “malignant” (pleomorphic) lesions and 36% of “benign” lesions exhibited aggressive local recurrence or distant metastases. As is true with most tumors involving bone, lesions with radiographic features of aggressive growth (e.g. bone infiltration and erosion,

poorly defined margins, cortical breakthrough with soft tissue extension), regardless of histology, were locally aggressive or metastasized [11]. Similarly, in a retrospective review of 150 cases of benign fibrous histiocytomas of the orbit, Font and Hidayat reported a local recurrence rate of 57% for tumors that had infiltrative margins or areas of increased cellularity [6].

The primary management of fibrous histiocytomas is complete surgical resection. Radiation therapy following surgery may provide local control in up to two thirds of the patients [14]. However, even in institutions with relatively larger experience, the number of patients with head and neck lesions are of limited number and therefore, a solid conclusion regarding the management protocol for this location, may not be possible. Chemotherapy, particularly with adriamycin or vinblastine, has been reported to achieve partial tumor response in some cases [2, 7].

## Conclusion

To our knowledge, this is the first documented case of fibrous histiocytoma arising in the pterygopalatine fossa with intracranial extension. The rapid local tumor recurrence and invasion of adjacent tissues supports the general principle of skeletal tumors, that radiographic features can be helpful when trying to anticipate the biologic behavior of a tumor with benign or borderline histology. Although the benefit of multimodal therapy in the treatment of fibrous histiocytomas has not been established, aggressive postoperative management with radiation and/or chemotherapy may be indicated for selected cases of tumors that invade bone.

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### Comment

The presented case offers interesting information for neurosurgical, ENT, and maxillo-facial readers, that an almost complete re-

section of the tumor, declared (patohystological findings) as benign, is malignant in reality and despite “successful” surgical treatment, the disease runs its deadly course with unchanged speed till the very end.

This report should be a stimulus to all the colleagues dealing with skull base pathologies to collect more information about such cases, and to try to find new solution for stopping the killer. While trying to find an appropriate answer – it is evident from this pathology – that an accurate patohystological diagnosis (obtained on the basis of needle biopsy) would be of some help in decision making for the combination and order of modalities of treatment. And, last but not least important is the fact that the clinical course of the disease is equally important as the patohystological finding and/or MRI findings.

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