

Chondromyxoid Fibroma of the Ethmoid Sinus Complicated by a Brain Abscess

– A Case Report and Literature Review –

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Chondromyxoid fibroma (CMF) is a relatively rare bone tumor that was first described by Jaffe and Lichtenstein in 1948. CMF of the sinonasal tract is very rare. A 28-year-old male presented with long-standing, intermittent, pulsatile pain in the right temporal area. A computed tomography scan showed a 20 × 19 mm round, bony density in the right ethmoid sinus with fluid collection in the ethmoid and frontal sinuses. Additionally, a cystic lesion with surrounding edema was found in the right frontal lobe. The patient underwent a partial ethmoidectomy and frontostomy. A histological examination showed polygonal and stellate cells in a myxoid and chondroid background with a pattern of lobulation and plaque-like calcification. The bone lesion was revealed as a CMF of the ethmoidal sinus, and the frontal lobe cystic lesion was a brain abscess associated with the CMF. We present the case of a CMF of the ethmoid sinus complicated by a brain abscess.

Key Words : Fibroma; Ethmoid bone; Brain abscess

Chondromyxoid fibroma (CMF) is a relatively rare bone tumor that was first described by Jaffe and Lichtenstein¹ in 1948. This tumor makes up less than 1% of all bone tumors and often involves the proximal metaphyses of the long bones, especially the femur and tibia.² CMF of the craniofacial bones is very rare, with the mandible and maxilla being the most common site for CMF of the craniofacial bones.³ Involvement of the sinonasal tract is rare. Only ten cases of sinonasal CMF have been reported in the English literature.⁴ Among them, eight cases were ethmoidal CMF. It is important to distinguish ethmoidal CMF from chondrosarcoma and other malignant myxoid tumors. In this article, we present a rare case of an ethmoidal CMF complicated by a brain abscess. We also discuss the clinical and pathological features and review the findings of previous ethmoidal CMF reports.

CASE REPORT

A 28-year-old male presented with long-standing, intermittent, pulsatile pain of the right temporal area. He had no relevant medical or surgical histories. His pain had intensified 3 weeks prior in association with an upper respiratory infection. Initial laboratory test results were within normal limits. An initial brain computed tomography (CT) scan revealed a 20 × 19 mm round, bony density in the right ethmoid sinus with fluid collection in the ethmoid and frontal sinuses. Additionally, a cystic lesion with surrounding edema was found in the right frontal lobe (Fig. 1). The lesion was suspected to be a brain abscess. A later paranasal sinus CT revealed that the bony density was suggestive of a benign bone tumor such as an osteoma or osteoblastoma (Fig. 2). The patient underwent surgical curettage. Surgery revealed edematous ethmoid bullae with scanty discharge on the mucosal surface. Curettage of the ethmoidal

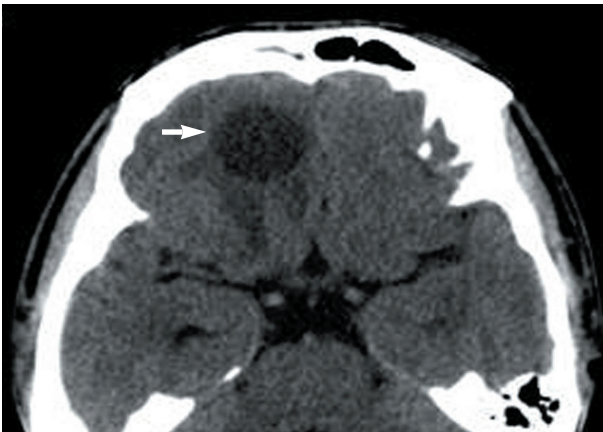


Fig. 1. Cystic mass-like lesion with surrounding edema of the right frontal lobe is seen on a brain computed tomography scan.



Fig. 2. Lobulated mass of the right ethmoid sinus is seen on a paranasal sinus computed tomography scan.

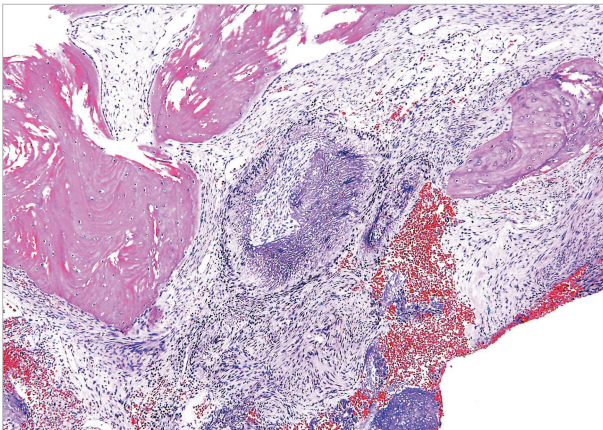


Fig. 3. Tumor lobules are composed of a chondroid or myxoid matrix with calcification in the central area and polyhedral or round or stellate shaped cells in the periphery.

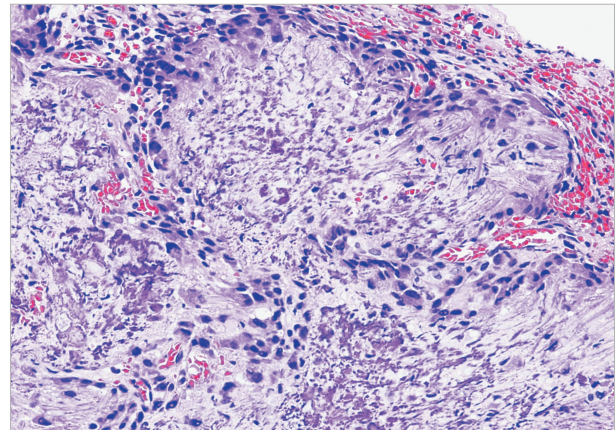


Fig. 4. Polyhedral, round, or stellate shaped cells in the periphery of the tumor lobules.

bone tumor was performed. Subsequently, a frontostomy was also performed, and a pus-like discharge was aspirated from the frontal sinus. The curettage specimen was fragmented, dark brownish, capsuleless, glassy, soft, and resembled fibro-chondral tissue. A histological examination showed a characteristic lobular configuration of the tumor. The lobules were separated by a fibrous band composed of spindle cells (Fig. 3). The lobules were composed of a chondroid or myxoid matrix with calcification in the central area and polyhedral, round, and stellate shaped cells in the periphery (Fig. 4). The tumor cells had occasional hyperchromatic nuclei with abundant pink cytoplasm, and the nucleoli were inconspicuous. One mitotic figure was found in the entire area. Necrotic areas were not found. The calcification pattern was a fine, granular or dense, plaque-like formation. Immunohistochemical staining for Sox9 was positive for the nuclei of about 20% neoplastic cells. A few neo-

plastic cells were positive for S-100. These findings were consistent with CMF. The patient's pain was relieved after surgery.

DISCUSSION

The World Health Organization defines CMF as "a benign tumor characterized by lobules of spindle-shaped or stellate cells with abundant myxoid or chondroid intercellular material."⁵ These tumors have a predilection for the long bones of children and adults in their 20s, while involvement of the craniofacial bones is found in only 5.4% of patients.⁶ CMF has been reported in many craniofacial region sites; however, the tumor is extremely rare in the ethmoid sinus, with only eight cases reported in the English literature (Table 1).^{3,7-13} This tumor type has been described in neonates and adults of both genders. There is no

Table 1. Nine cases of ethmoid CMF: a clinicopathological and radiographical literature review

Authors	Age (yr)/Sex	Size (cm)	Radiologic findings	Clinical presentation	Treatment
Koay <i>et al.</i> ⁷	57/F	2.5 × 2.0	Mass on the nasal bridge extending into the frontal sinus, eroding the right orbit	Slowly expanding, painless swelling over bridge of nose	Incomplete surgical excision
Baujat <i>et al.</i> ⁸	50/F	NA	Osteolytic lesion in the ethmoid bone invading the adjacent bone and dura mater	Frontal headache, pain, nasal obstruction	Surgical excision with dura removal
Nazeer <i>et al.</i> ³	20/M	NA	Mass with bone erosion, scalloping, extension into the nasal cavity	Respiratory difficulty since birth	Surgical resection
Hashimoto <i>et al.</i> ⁹	32/M	6.0 × 3.5	Well-defined, focally-destructive mass extending into the left nasal cavity and frontal sinus	Painless frontal swelling and progressive exophthalmos	Surgical resection
Isenberg ¹⁰	34/F	2.5 × 1.4	Calcified mass at the right middle turbinate	Difficulty in breathing	Endoscopic ethmoidectomy
Cruz <i>et al.</i> ¹¹	10/F	NA	Large mass in the ethmoidal sinus eroding the medial orbital wall	Progressive exophthalmos	En block resection
Mendoza <i>et al.</i> ¹²	0/M	NA	Solid, well-defined mass in the ethmoid sinus	Respiratory difficulty since birth	Block resection
Szmeja <i>et al.</i> ¹³	8/F	7 × 6	Suggestive benign tumor without destruction of the surrounding bone	Occlusion of the nasal cavity, dislocation of the left eyeball	Total enucleation
Our study	28/M	2 × 1.9	Round, bony density in the right ethmoid sinus with fluid collection in the ethmoid and frontal sinuses	Intermittent pulsatile pain in the right temporal area	Partial curettage

CMF, chondromyxoid fibroma; NA, not applicable.

established radiological pattern for CMF due to its rarity. In all cases, the diagnosis was confirmed only after an operation. After reviewing previous reports, a CMF of the ethmoid sinus complicated by a brain abscess has not been reported previously.

A differential diagnosis of a CMF is difficult histologically as well as clinically. It is very important to distinguish between a CMF and chondrosarcoma because the management of these two entities differs. Histologically, a CMF is usually well-demarcated, even when it erodes the bone. The lesions are nodular with myxoid lobules separated by thin, fibrous septa. Importantly, a CMF has a paucicellular center rather than the uniform cellular arrangement observed in chondrosarcoma, and mitotic figures are exceptional.⁵ When compared to a CMF, chondrosarcoma differs in that the tumor encases the preexisting bony trabeculae.⁸ Chondrosarcoma reveals well-differentiated hyaline cartilage and no fibrous component,^{4,11} with individual cells showing nuclear pleomorphism and atypia. Additionally, the differential diagnosis includes fibrous dysplasia and chordoma. Fibrous dysplasia lacks the epithelioid cells and lobulation pattern seen in a CMF.¹⁴ Chordoma, especially in its chondroid form, can be distinguished from a CMF by its infiltrative margin and physaliferous cells with an eosinophilic cytoplasm and absence of a fibrous component.⁸

Clinically, CMFs arising in the paranasal sinuses can present with pain, swelling, nasal obstruction, or an asymptomatic mass.⁴ Our patient presented with intermittent temporal pain that was intensified 3 weeks prior to his clinic visit; we suspected that

the brain abscess arose at this time. We believe that the CMF of the ethmoid sinus may have been one of the causes of the brain abscess, as the CT scan showed that the ethmoidal bone lesion extended into the frontal sinus. Pus from the narrowed frontal sinus opening was drained by a frontostomy. Blockage of mucus flow in the paranasal sinus was a possible source of infection. From these findings, it is reasonable to suggest that the CMF in the ethmoid sinus may have contributed to the development of the brain abscess.

Sox9 is a transcription factor that was recently described as a master regulator of chondrogenesis. It plays a role in the early phases of chondrocyte differentiation and is useful for distinguishing between undifferentiated tumors in the small, blue, round-cell category and tumors of true cartilaginous origin.¹⁵ Although Sox9 reactivity does not differentiate CMF from a low-grade chondrosarcoma because both are cartilaginous, it could assist in establishing a diagnosis of CMF when histological features are suggestive of a different cell lineage.⁴

Although CMFs are benign tumors, invasion into adjacent bone and the dura mater of the ethmoid sinus has been reported.^{3,8,9} Thus, complete surgical resection is considered to be the optimal means for treating a CMF.² However, a CMF of the craniofacial bones is best treated by curettage because of the functional and cosmetic deformities that result from complete surgical resection. Curettage is ordinarily successful but increases the risk of recurrence.² The recurrence rate from curettage is approximately 25%.³ Careful periodic surveillance for recurrence

is recommended for patients who have received curettage. Some authors have propose using radiotherapy for local relapses following surgical excision, particularly at the base of the skull.¹²

In conclusion, a CMF of the ethmoid sinus is very rare. Although a CMF is a benign bone tumor, serious medical conditions such as a brain abscess may be associated with a CMF, depending on the tumor location. Patients who receive curettage of the CMF should be checked periodically for recurrence.

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