Fibromyxomatous spindle cell neoplasm of the ethmoid sinus with extension into the optic cavity: report of a case and review of the literature

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ABSTRACT

Introduction: Fibromyxomatous spindle cell neoplasms represent a recently described form of fibrous dysplasia. Case Report: A 43 year-old woman presented to our clinic with chronic sinusitis and an infiltrative ethmoid mass consistent with a large mycetoma. Clinical manifestations of the tumor included right frontal and para-orbital pain with proptosis and chronic eye-drainage. Comparison of CT images to prior studies demonstrated a slowly growing mass with diffuse enhancement in the right ethmoid complex with obstruction of the frontal sinus and erosion through the lamina papyracea. The tumor was removed, with subsequent pathology demonstrating a fibromyxomatous spindle cell neoplasm. Unfortunately, the tumor recurred following surgery, and within three months had grown to its initial size.

Discussion: Myxomatous tumors are rarely found in the paranasal sinuses. Paranasal fibromyxomas have been reported in the literature. These rare tumors are characterized as aggressive lesions with a propensity for recurrence. In this manuscript, we present the first reported case of a fibromyxomatous spindle cell neoplasm occurring in the ethmoid sinuses. While immunohistochemistry is necessary to differentiate this lesion from paranasal myxomas, the clinical course of both entities appears similar.

Conclusion: Fibromyxomatous spindle cell neoplasm is a rare tumor of the ethmoid sinus. This entity must be included in the differential diagnosis for an aggressively growing lesion of the para-nasal sinuses.

INTRODUCTION

Fibromyxomatous spindle cell neoplasms, not further classified, represent a recently described form of dysplasia with histologic similarity to sinonasal and odontogenic myxomas. Differentiated by their histologic appearance and unique immunophenotype, this report represents the first known case of this unusual tumor, and first report of its occurrence within the sinonasal pathway.

CASE REPORT

A 43 year-old woman was referred to our clinic from an outside physician with complaints of chronic sinusitis and sneezing unrelied by two prior functional endoscopic sinus surgery (FESS) and septoplasty procedures. Review of surgical pathology revealed the identification of calcified material removed from her right frontal sinus. On initial exam the patient had vital signs consisting of blood pressure: 137/79, pulse: 75, respiratory rate: 20 and temperature: 97.6F. On nasal endoscopy a small section of the right, distal-inferior turbinate showed signs of previous excision, while a large mass of the entire right ethmoid complex was seen without signs of purulence or polyps. Comparison of previous computed tomographic studies demonstrated a slowly growing mass with diffuse enhancement of the right ethmoid complex with opacification of the frontal sinus and erosion through the lamina papyracea [Figure 1]. After extensive discussion the patient elected to proceed with surgical marsupialization of the ethmoid mass, with utilization of a Stealth CT protocol. The surgery was completed without complication, with excision of the right ethmoid mass as well as anterior and posterior ethmoidectomy. The excised mass was firm, tan-white in appearance and measured 3 x 2 x 0.5 cm in aggregate. Immunohistochemistry yielded a diagnosis of fibromyxomatous spindle cell neoplasm, not further classified [Figure 2], with an unusual immunophenotype containing vimentin, S-100 protein and desmin, and negative for MSA, SMA, CD31 and CD34. Proliferation rate by KI-67 staining was estimated at 5-7%.

Following surgery the patient suffered a dramatic recurrence of disease, with the tumor returning to its initial size within three months.

DISCUSSION

Myxomatous tumors represent a group of mesenchymally-derived benign neoplasms. These rare tumors have been described in various locations throughout the body, including the skin, subcutaneous tissue, heart and various sites involving the head and neck. Within this group of myxomatous tumors, sinonasal and odontogenic myxomas share the greatest amount of clinical and histologic similarity with the recently described fibromyxomatous spindle cell neoplasm, without further characterization, but differ in important ways. While sinonasal and odontogenic myxomas are both locally aggressive tumors with a propensity for recurrence, reports of sinonasal myxomas are limited to pediatric patients with maxillary masses. Odontogenic myxomas, on the other hand, frequently occur in young adults, display radiolucent imaging and a unique immunophenotype that differentiates it from fibromyxomatous spindle cell neoplasms.

Immunohistochemical studies have characterized this unique phenotype as positive for vimentin, S-100 protein and desmin, and negative for CD31, CD34 and the muscle markers MSA and SMA. It is the lack of CD34 presentation that differentiates fibromyxomatous spindle cell neoplasms from solitary fibrous tumors. In this manuscript, we present the first reported case of a fibromyxomatous spindle cell neoplasm, not otherwise specified, found in the ethmoid sinus. While immunohistochemistry is necessary to differentiate this lesion from various forms of myxomatous disease, the aggressive clinical course of these entities appears the same.

REFERENCES