INTRODUCTION

Successful surgical treatment of chronic frontal sinus disease has been, for a long time, an Achilles heel for rhinologists. With its rigid bony encapsulation, and relatively small portal into the nasal cavity, the frontal sinuses present a great challenge in rhinology.

Besides the difficulties encountered in the management of chronic inflammatory diseases of the frontal sinuses, the diagnosis and management of benign tumors in this region are also challenging.

Benign tumors of the fronto-ethmoid region are frequently locally aggressive, with a potential of spreading anteriorly to the skin, posteriorly into the anterior cranial fossa, or inferiorly into the ethmoid, nose, or orbit.

Several surgical procedures have been proposed and used over the years for the management of frontal sinus diseases. Some procedures aimed at the evacuation and obliteration of the frontal sinuses, whereas other procedures aimed at establishing a better drainage pathway for the frontal sinuses. Although traditionally open approaches remain widely used, many endoscopic techniques developed for the management of inflammatory sinus disease are now being applied to the management of tumors of the frontal sinus.

In this case report, the authors present a case of frontal sinus mucocele and fibro-osseous and respiratory epithelial hamartoma of the fronto-ethmoid region. The diagnosis and management of this case are presented, in addition to a detailed discussion of the tailor-made surgical approach applied in its management.

CASE REPORT

A 31-year-old Lebanese lady presented to the ENT clinic chiefly complaining of right eye protrusion of one month duration. She also complained of right neck and ear pain for the last three years, and of repetitive attacks of upper respiratory tract infections. She did not have any relevant past medical or surgical history, did not have any known allergies, and was not on any medication.

On examination, she was found to have exophtalmus, bilateral middle meati polyps, and bilateral septal deviation.

A CT scan of the paranasal sinuses was ordered, which revealed a soft tissue mass in the right frontal sinus, suggestive of a mucocele, eroding the roof of the right orbit with intact posterior wall of the frontal sinuses. There was
also opacification of the left frontal sinus. The CT scan also revealed ground glass opacification of the ethmoid sinuses, suggestive of fibrous dysplasia of the ethmoid sinuses.

The patient was taken to the operating room for surgical management of the fronto-ethmoidal sinus disease. The operative procedure was done under general anesthesia. A bicoronal incision was done, skin and frontalis muscle were elevated down to the level of the orbital rim, and the osteoplastic flap was fractured caudally. Intra-operatively, the frontal sinuses were found to be entirely filled with mucocele which seemed to be independent right and left. The right mucocele was eroding the roof of the right orbit and was tightly adherent to the periorbita. The posterior wall of the frontal sinuses was intact. The mid frontal sinus region was filled with fleshy fibrous tissue. Brittle bone was found to be occupying the central portion of the frontal sinus and frontal recess, and adhered to the lamina papyracea and the base of the skull. This bone was part of the tumor which included the middle turbinates, and the anterior and posterior ethmoid cells. Several small mucoceles were found in the tumor bulk. This tumor bulk, when grabbed from one end and moved, was found to be semi-mobile, and seemed to move as a block.

Using the operative microscope, the frontal sinus mucocele was dissected from the periorbita of the right orbital roof, and from the remainder of the frontal sinus and removed. The tumor mass, composed of fleshy tissue and brittle bone, was then removed in bits leading eventually to the removal of the entire interorbital frontal floor. Working through this exposure, the superior part of the septum was removed along with the anterior and posterior ethmoid cells and the middle turbinates. A large naso-frontal communication was obtained. Minimal CSF leak was noticed in the left cribiform plate which stopped spontaneously after covering it with mucosa, and later with surgicel and gelfoam. Two silastic tubes (1 cm in diameter) were placed through the frontal recesses reaching the inferior turbinates, and were sutured together and to the remaining part of the posterior septum.

The specimen was submitted to pathology in two parts, entitled “Frontal sinus” and “Ethmoid sinus.” Both parts consisted of multiple grayish polypoid fragments, associated with multiple bony fragments. The largest polypoid fragment was brownish and measured 1.8 x 1 x 0.6 cm.

Microscopically (Figure 1), the tumor consisted of polypoid shaped portions covered by respiratory epithelium, having edematous and inflamed lamina propria, and variably sized, sometimes dilated glands lined by columnar ciliated cells admixed with fibro-osseous nodules. These glands were intimately admixed with fibro-osseous nodules, composed of a mixture of woven and lamellar bone, prominently rimmed by osteoblasts and separated by intervening fibrous tissue. In addition, seromucinous glands were noted focally. The diagnosis of fibro-osseous and respiratory epithelial hamartoma was established.

DISCUSSION

This case report presents a rare pathology of the sinonasal tract, namely fibro-osseous and respiratory epithelial hamartoma. In addition, the operative approach employed in the management of fronto-ethmoidal disease in this case is explained and discussed.

Hamartomas of the nasal cavity are rare entities. These tumors have been called various names depending on their histologic components. Hamartomas of the mesenchyme are more common and have been named chondroid, chondromesenchymal, angiomatosus, or lipomatous, depending on the preponderant tissue. The epithelial hamartomas, on the other hand, comprise such lesions as respiratory epithelial adenomatoid hamartoma, seromucinous hamartoma, or salivary gland tumor of the nasopharynx.

Rare instances of respiratory epithelial hamartoma with additional chondro-osseous component have been described. The epithelial hamartomas of the respiratory tract have been reported as single cases. The largest series was described by Wenig et al. [1] where the authors describe the clinicopathologic features of 31 cases of epithelial adenomatoid hamartomas occurring in the nasal cavity, paranasal sinuses, and nasopharynx.

The paranasal sinuses, especially the ethmoid sinuses, are rarely involved. Presenting signs and symptoms include nasal obstruction and epistaxis. The histopathologic features are those of glandular proliferation that results in a polypoid formation in some areas. These glands are variably sized and shaped, and lined by ciliated respiratory type epithelium. Atrophic changes of the glands were noted in several cases. Other than the glan-
dular proliferation, the histologic appearance also includes inflammatory changes, such as stromal edema, seromucinous gland proliferation, and vascular and fibroblastic proliferation.

Within the nasal cavity the most common site of occurrence is the nasal septum, particularly along the posterior aspect [1]. The gross appearance of the mass lesion suggests a diagnosis of an inflammatory polyp, but because of subtle differences, including more indurated quality, these polyps are considered unusual for typical inflammatory polyps [1].

The differential diagnosis of these respiratory epithelial hamartomas includes schneiderian papillomas of the inverted type, and adenocarcinomas.

Limited but complete surgical resection is the treatment of choice. Weing et al. [1] document no instances of recurrent disease after such treatment modality. The surgical treatment of chronic frontal sinusitis usually takes one of two courses. Surgical intervention is directed towards either obliteration of the frontal sinus or enlarging the naturally small drainage system. The surgical procedures used for the management of frontal sinus disease employ an approach that is either open, endonasal, or a combination of both. Lothrop, in 1914, described an open approach procedure for dealing with fronto-ethmoidal disease. In the original Lothrop procedure, the interfrontal sinus septum was removed and a large opening was created connecting the frontal sinus cavity with the nasal cavity. This procedure, as it was originally described, is a frontal sinus mucosa preservation procedure that was done through a transorbital approach. Unfortunately, the original Lothrop procedure was associated with high failure rates because it requires removal of portions of the lamina papyracea bilaterally, which was associated with medial prolapse of orbital content and obstruction of frontal drainage.

Montgomery, in the 1960s, popularized another open approach, the osteoplastic flap with fat obliteration of the frontal sinus. This procedure has become the chosen method for the treatment of chronic frontal sinus diseases [2]. However, obliteration of the frontal sinus was found to be associated with complications. Mucoceles formation in the frontal sinus following osteoplastic obliteration was documented by several authors. Weber et al. recently reported mucoceles seen on magnetic resonance imaging in 9.4% of the patients, an average of two years after frontal sinus obliteration [3]. The mucosa of the frontal sinus tends to invaginate itself into dehiscences and foramina in the periosteum of the posterior and superior walls of the frontal sinus, namely the foramina of Breschet [4]. Failure to eradicate such mucosa from the sinuses may result in postoperative mucocele formation. Furthermore, the amount of implanted fat was found to be reduced to 20% of its original level within 15 months postoperatively [5]. This may prevent fibrous tissue replacement of the frontal sinus, and predispose to mucocele formation. In addition, the osteoplastic flap procedure was associated with significant postoperative pain [6].

The modified Endoscopic Lothrop procedure, a relatively new procedure, allows for the endonasal creation of a large common nasofrontal drainage pathway. This procedure is accomplished by endoscopic removal of the floors of the frontal sinuses bilaterally, and connection of the right and left frontal sinuses through excision of the frontal sinus septum and the postero-superior part of the nasal septum [7]. The main benefit of this procedure was noticed in cases of failed osteoplastic obliteration of the frontal sinus, where it is used as a salvage technique [8].

In the course of this study, we found that the modified Endoscopic Lothrop procedure shows a failure rate of 18%, associated with closure of the fronto-nasal drainage [9].

In the case presented in this report, the authors were faced with a combined pathology; mucoceles of the frontal sinuses, and a fibro-osseous tumor of the fronto-ethmoidal sinuses. The surgical intervention had to be tailor-made. The osteoplastic flap approach provided a wide exposure of the frontal sinus, and allowed ample maneuvering for resection of the ethmoid disease. Obliteration of the frontal sinus, however, lost in favor as the operation proceeded for two reasons. On one hand, the mucosa of the frontal sinus may not have been completely eradicated especially on the right side where the mucocele was intimately adherent to the orbital periosteum. On the other hand, there was a possibility of incomplete microscopic removal of the fronto-ethmoidal fibro-osseous tumor, with its interspersed epithelial elements. Leaving mucosa in the frontal sinus underneath fat obliteration is a high risk for recurrence of frontal sinus mucocele.

After removal of the ethmoid disease, along with the central part of the floor of the frontal sinuses, and the superior part of the nasal septum, a wide frontal nasal communication was created. This communication will allow for appropriate drainage of the frontal sinuses, and will prevent potential complications of incomplete mucosal eradication of the frontal sinus.

In other words, a tailor-made procedure was done to manage the presented sino-nasal pathology. This procedure establishes a Lothrop-type fronto-nasal drainage pathway through an osteoplastic flap approach.

REFERENCES


