XANTHOMA OF THE SPHENOID SINUS:
A RARE PRESENTATION

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ABSTRACT

OBJECTIVES: 1) Present what is to the best of our knowledge the first reported case of a patient with a sphenoid sinus xanthoma. 2) Discuss the details of the clinical presentation, histopathological and radiological findings, and management of this patient.

METHODS: Medical charts and the scientific literature were reviewed.

RESULTS: A 36-year-old female patient with family history of hyperlipidemia presented with a 1-year history of sporadic epistaxis and an episode of severe headache 2 months prior to evaluation. The rest of her medical history and physical examination were unremarkable. CT-scan and MRI images revealed a soft tissue lesion occupying and infiltrating the posterolateral aspect of an expanded sphenoid sinus on the left side. Patient underwent endoscopic sinus surgery in December 2011 with intraoperative findings of a yellow-tan, friable soft tissue filling the left sphenoid sinus. Biopsy was taken for which histologic and immunohistochemical analysis revealed proliferating foamy histiocytes consistent with xanthoma cells. As of her last follow-up examination 16 months post-op, the patient has remained asymptomatic without evidence of disease progression on nasal endoscopy or CT scan.

CONCLUSION: Very few cases of xanthomas arising in the paranasal sinuses have been described in the literature. Those that have been described have only occurred in the maxillary sinus. We present the case of a 36-year-old woman with family history of hyperlipidemia with a left sphenoid sinus xanthoma. This is to our knowledge the first case of such a lesion reported in the English literature.

INTRODUCTION

Xanthomas are nonneoplastic granulomatous lesions composed primarily of lipid-laden macrophages accompanied by varying degrees of fibrosis, inflammatory reactions and crystalline cholesterol deposits. They are mostly associated with disorders of lipid metabolism, which result in abnormally high circulating levels of cholesterol and secondary to other metabolic disorders such as diabetes mellitus and hypothyroidism. The most common sites for xanthoma formation are along the Achilles, patellar and extensor tendons of the hands as well as exposed surfaces of the body susceptible to repetitive trauma. Solitary xanthomas have rarely been observed in the head and neck area and most occur along the temporal bone surface. A search of the literature revealed only 2 cases of maxillary sinus xanthomas. This is a case report of a 36-year-old female referred to our institution for evaluation of a sphenoid sinus tumor; later diagnosed as a xanthoma of the sphenoid sinus. To the best of our knowledge this is the first reported case of a patient with a sphenoid sinus xanthoma.

CASE REPORT

The patient is a 36-year-old Hispanic female with past medical history of Hashimoto’s hypothyroidism and family history of hyperlipidemia that presented with a 1-year history of sporadic left sided epistaxis and an episode of severe headache 2 months prior to evaluation. She denied facial pain, hypoesthesia, visual change, or symptoms of sinusitis, and also denied a history of facial trauma. The rest of her medical history, review of systems and physical examination, including in-office nasal endoscopy, were unremarkable. Computed tomography (Figure 1) and MRI images of the paranasal sinuses revealed a soft tissue lesion occupying and infiltrating the posterolateral aspect of an expanded sphenoid sinus on the left side. Patient underwent endoscopic sinus surgery in December 2011 with intraoperative findings of a yellow-tan, friable soft tissue mass filling the left sphenoid sinus. Biopsies were taken which revealed proliferating foamy histiocytes (xanthoma cells) and cholesterol clefts surrounded by foreign body giant cells (Figure 4). A diagnosis of primary xanthoma was made. As of her last follow-up examination 16 months post-op, the patient has remained asymptomatic without evidence of disease progression on nasal endoscopy (Figure 3) or CT scan (Figure 2).

DISCUSSION

The differential diagnosis of a sphenoid sinus soft tissue mass includes inflammatory polyps, mucocele, infiltrative lesions such as fibrous dysplasia, and various benign and malignant neoplasms including intra-sphenoidal pituitary tumors, inverted papilloma and squamous cell carcinoma. Solitary xanthomas are rare in the head and neck; they are more likely to occur at the elbows, buttocks, patellar and Achilles tendons. This is only the third report of such a lesion in the paranasal sinuses, and first one in the sphenoid sinus.

Xanthomas are benign lesions formed in areas with increased vascular permeability due to minor trauma or vasoactive compounds. This causes lipid-rich plasma leakage into tissues where it undergoes phagocytosis by macrophages, creating “foamy cells.” Cholesterol builds up in a crystalline form inside the macrophages leading to inflammation characterized by giant cells and fibrosis. The metabolic disorders associated with xanthomas typically include heritable hyperlipidemias, diabetes mellitus, renal disease, hypothyroidism, alcohol abuse, and liver disease.

Most patients with xanthoma are managed medically, without surgery. The underlying cause must be identified and treated accordingly. The mainstay of treatment is alteration in diet, with restriction of cholesterol and saturated fats. When cholesterol and triglycerides are not sufficiently lowered by diet alone, pharmacotherapy is indicated with lipid-lowering agents.

Since xanthomas are benign lesions, surgical treatment may be limited to a biopsy for histologic identification. The indications for excision are cosmetic deformity or local symptoms. Continued growth has been observed in a number of cases in which the xanthoma had been debulked. Considering the above, when surgery is performed, attempts should be made to completely excise the mass with minimal morbidity.

CONCLUSION

Very few cases of xanthomas arising in the paranasal sinuses have been described in the literature. Those that have been described have only occurred in the maxillary sinus. We present the case of a 36-year-old woman with family history of hyperlipidemia with a left sphenoid sinus xanthoma. This is to our knowledge the first case of such a lesion reported in the English literature.

REFERENCES


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