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Journal of Ear, Nose and Throat Disorders

Case Report

Polymorphous Low-Grade Adenocarcinoma: A Rare Cause of Long-Term Epistaxis

Teng-chin Wang[1]*, Yi-Tsung Yang[2] and Ming-Tsun Tsai[3]

[1]*Department of Otolaryngology, Tainan Municipal Hospital, Taiwan*
[2]*Department of Internal Medicine, National Taiwan University Hospital Hsin-Chu Branch, Taiwan*
[3]*Institute of Clinical Medicine, National Yang-Ming University, Taiwan*

***Corresponding author**
Teng-chin Wang, Department of Otolaryngology, Tainan Municipal Hospital, Tainan City, No.670, Chongde Rd, East Dist, Tainan City 701, Taiwan, Tel: 886-6-2609926; Fax: 886-6-2606351; Email: tengchin27@hotmail.com

Submitted: 29 March 2016
Accepted: 16 May 2016
Published: 18 May 2016

Keywords
- Epistaxis
- Nasal polyp
- Polymorphous low-grade adenocarcinoma

Abstract

Epistaxis is a common complaint encountered in clinical practice and usually ceases spontaneously. However, certain recurrent intractable cases caused by several rare diseases can cause considerable morbidity. A systematic approach is required to establish the diagnosis in such conditions. We report a case of a 52-year-old man who presented with chronic recurrent Epistaxis due to polymorphous low-grade Adenocarcinoma (PLGA). This is the rare case of chronic Epistaxis caused by PLGA. Complete examination and timely surgery are the key components of a successful treatment strategy for such rare disorders.

ABBREVIATIONS

PLGA: Polymorphous Low-Grade Adenocarcinoma

INTRODUCTION

Epistaxis is a common problem that affects approximately 60% of all people in their lifetime [1] and is usually a benign self-limiting illness. However, it can occasionally be intractable and is caused by several rare diseases. Refractory epistaxisis attributable to hypertension, anti platelet medications, and alcohol abuse [2]; however, most cases are typically idiopathic. A systematic approach is required to establish the diagnosis of Epistaxis in such conditions. In this report, we present a case in which polymorphous low-gradeadenocarcinoma (PLGA), a rare disorder of the sinonasal tract, caused chronic recurrent epistaxis in an adult male and discuss management strategies for these lesions.

CASE PRESENTATION

A 52-year-old man presented with a 30-month history of intermittent Epistaxis and purulent discharge from the left naris. He was under regular aspirin medication for preventing coronary artery disease. In addition, he had undergone bilateral Caldwell–Luc operation for benign maxillary sinus disease 15 years previously. He visited different clinics several times for the aforementioned symptoms, which were subsequently diagnosed as chronic paranasal sinusitis and the adverse effects of antiplatelet agents. Aspirin was discontinued; however, it did not reduce the frequency and severity of the nose bleeds. Therefore, he visited our department to seek medical help. On physical examination, no obvious lesions or bleeders were observed over the common meatus, septum, or vestibule. Further evaluation through fiber optic endoscopy revealed scant crust accumulation in the left middle meatus, and an easily bleeding and nonfriable lobulated mass with neovascularization was identified after removal of the crust (Figure 1a). The extent of the mass was evaluated through contrast-enhanced computed tomography, which revealed soft tissue enhancement in the left ethmoid sinus and nasal cavity. No intra orbital destruction or skull base invasion was observed (Figure 1b). The left lamina papyracea was unremarkable.

Biopsy reported basal cell adenoma, but malignant carcinoma cannot be ruled out. After discussing with patient, modified

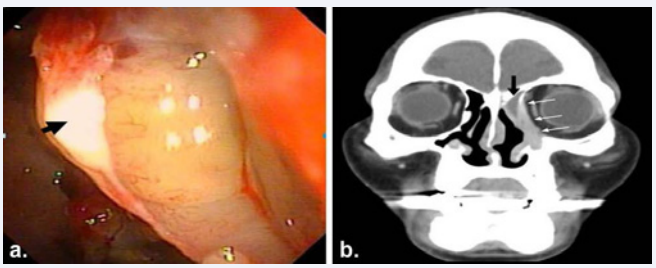

Figure 1 (a) Polyp-like lesion with neovascularization in the left middle meatus was observed (arrow: left middle turbinate); (b) Enhancement was observed in the adjacent soft tissues of the left ethmoid sinus and nasal cavity. The skull base (thick black arrow) was intact, and the periorbital contour was smooth (thin white arrow). In addition, absence of bony density in the left lamina papyracea was noted.

endoscopic medial maxillectomies were performed 2 weeks later under general anesthesia. The dissection plane was directed toward the skull base and lateral to the periorbita. The structures above this region were intact, and no gross residual tumor was observed. The excised tumor specimen measured 3.5 cm × 1.2 cm × 0.7 cm. The microscopy of the sections revealed PLGA comprising irregular infiltrating solid nests and cuboidal cells growing in an adenoid cystic pattern arranged in ducts, trabeculae, and tubules (Figure 2a). Under high magnification, most tumor cells were bland and relatively uniform. The round and ovoid nuclei were normal in size or slightly enlarged, and lymph vascular invasion was observed (Figure 2b). Mitoses were infrequent. Perineural, perivascular, and bony invasion, involving nearly all portions of the obtained fragments, were observed.

The pathologic staging was pT2Nx, and the patient received radiation therapy with a daily dose of 200 cGY and a total dose of up to 7000 cGY. In addition, tegafur/uracil at 300 mg/day in divided doses was administered as a post operative adjuvant therapy for 6 months. Epistaxis did not recur after surgery. Follow-up magnetic resonance imaging and fiberoptic endoscopy 25 months after treatment revealed tumor-free nasosinal cavities with mild scarring (Figure 3).

DISCUSSION

Epistaxis in Sino-nasal tract tumor can be fatal due to the massive size of the tumor leading to outstripping its blood supply. But in PLGA, the bleeding is often scanty rather than massive by its less aggressive entity. As in this case, the histopathology finding doesn't exhibit obvious hemorrhagic infarction and necrosis of the tumor. These characters may explain why epistaxis is intermittent and doesn't lead to severe constitutional symptoms for such a long time.

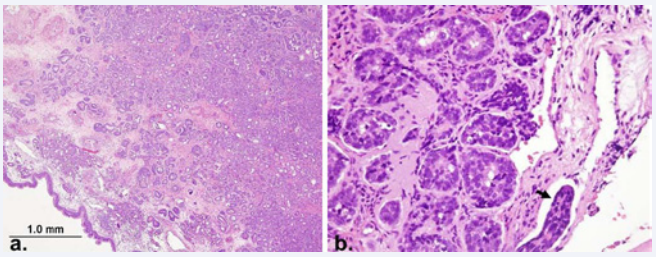

Figure 2 (a) Image of polymorphous low-grade Adenocarcinoma comprising irregular.

In filtrating solid nests and cells growing in an adenoid cystic pattern arranged in ducts, trabeculae, and tubules (40×, hematoxylin and eosin stain); (b) Nuclei are round and fairly uniform. Tumor cells are apparent in the lymphatic duct (black arrow) (400×, hematoxylin and eosin stain).

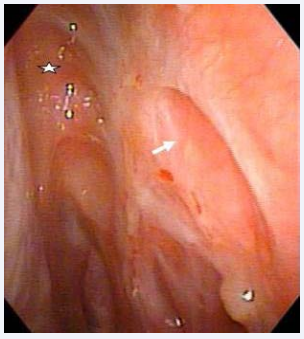

Figure 3 The nasal cavity is free of tumors with Mild scarring 25 months after treatment.
(White asterisk: the skull base; white arrow: the orbital wall)

PLGA is a minor salivary gland neoplasm that arises mostly in the oral cavity, particularly in the mucosa of the soft and hard palates [3]. PLGA presenting as a primary tumor of the extra oral region is rare, and only a few cases of PLGA in the sinonasal location have been reported [4,5]. These tumors tend to present as sinonasal polyps with or without nasal obstruction. Less common symptoms include pain, bleeding, and ulceration.

PLGA may exhibit a combination of several distinct growth patterns: tubular, trabecular, cribriform, and solid. In contrast to the morphological diversity, the nuclei are typically uniform with inconspicuous mitotic activity [6]. PLGA can usually be diagnosed from the typical cytological and histological features.

PLGA frequently exhibits an infiltrative growth pattern, lymphovascular, and perineural invasion; however, the prognosis for such patients is often favorable. The tumor is usually indolent, slow-growing, locally invasive, and rarely metastatic [7]. Wide local excision with appropriate margins is the current first-line treatment for the majority of PLGA [3]. Routine neck dissection is not recommended unless clinical evidence supporting lymph node involvement is available. The benefits of adjuvant therapy for PLGA remain controversial. Studies have reported that postoperative radiation therapy has a survival benefit for PLGA arising at unusual locations, such as the parotid glands [8]. The present case was successfully treated through surgery and adjuvant chemotherapy with postoperative radiation therapy; no recurrence was observed 25 months after the treatment.

In conclusion, PLGA is a rare cause of chronic intractable epistaxis and can be easily ignored. A fiberscopic examination is mandatory in such condition for definite diagnosis, and prompt surgery can usually improve the outcome. Additional studies are warranted to verify the role of adjuvant radiation therapy and chemotherapy in these tumors.

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Cite this article

Wang TC, Yang YT, Tsai MT (2016) Polymorphous Low-Grade Adenocarcinoma: A Rare Cause of Long-Term Epistaxis. J Ear Nose Throat Disord 1(1): 1002.