Bilateral Pulmonary Nodules after Surgery for Ameloblastoma: A Case of Misdiagnosis for 32 Years

Jie Liu
Wuxi People's Hospital
Shudong Yang
Wuxi People's Hospital
zhen yu (yuzhen1969@sina.com)
Wuxi People's Hospital
Tao Bian
Wuxi People's Hospital

Case Report

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Abstract

Background: We describe a rare case of metastatic ameloblastoma in a 68-year-old Chinese woman diagnosed with ameloblastoma of the right mandible in 1974.

Case presentation: Despite radical resection, she experienced repeated local recurrences, including asymptomatic bilateral pulmonary nodules, cough, and hemoptysis diagnosed as non-malignant inflammatory cell infiltration. The patient developed an aggravated cough, palpitations, and dyspnea and was admitted to our respiratory department in September 2018. Chest computed tomography revealed multiple bilateral lung nodules, masses, and soft tissue opacities. She was diagnosed with advanced metastatic ameloblastoma in bilateral lungs via bronchoscopic biopsy, with a misdiagnosis time of 32 years. Because her systemic condition was poor, systemic chemotherapy and radiotherapy were not recommended. Dyspnea significantly improved after interventional bronchoscopic therapy and tumor electroexcision; she died of respiratory failure 1 year after diagnosis.

Conclusions: Clinicians should carefully explore the medical histories of patients with surgical histories of ameloblastoma and subsequent unexplained pulmonary metastatic tumors to avoid misdiagnosis.

Background

Ameloblastomas are rare odontogenic tumors of epithelial origin that most frequently arise in the mandible. Radical resection is the mainstay of ameloblastoma treatment[1]. These tumors have a benign histopathological manifestation, they exhibit unique biological behaviors such as local invasiveness and repeated recurrence. Although reports suggesting the malignant potential of ameloblastoma[2], metastasis is rare. Here, we report a rare case of a patient with ameloblastoma of the right mandible who presented with bilateral pulmonary metastases 12 years after radical resection and whose case remained misdiagnosed for 32 years.

Case Presentation

A 68-year-old woman was admitted to our respiratory department on September 17, 2018 with progressively worsening dyspnea. She had a history of right mandible ameloblastoma that had been treated via surgical resection in 1974. She first developed an intermittent cough and hemoptysis in 2002, and presented with recurrences of the ameloblastoma of the mandible in 2006 and 2010. Her clinical symptoms, diagnosis and treatment are presented in Table 1.

Table 1
Patient's histories in the this case

Year	Respiratory symptom	Chest radiological manifestation	Clinical changes	Treatment
1974	Asymptomatic	/	Swelling of the right mandible, possible ameloblastoma	Partial resection of the ameloblastoma of the right mandible, iliac bone graft
1986	Asymptomatic	Scattered nodular lesions in both lungs	Palpable mass in the left lower abdomen, Diagnosis: uterine leiomyoma	Removal of the uterus and bilateral attachments
2000	Occasional cough and hemoptysis	Enlargement of and increase in the number of bilateral pulmonary nodules	Respiratory symptoms at presentation	Symptomatic treatment
2002	Recurrent cough and hemoptysis	Multiple nodules and mass shadows in both lungs	Pathology of percutaneous pulmonary puncture specimen revealed alveolar epithelial hyperplasia, interstitial fibrous tissue hyperplasia, infiltration of a small number of chronic inflammatory cells, and no cancer cells	No treatment, follow-up observation
2004	Aggravated cough, intermittent hemoptysis, decreased blood oxygen saturation (~ 85%), no dyspnea	/	/	No treatment, follow-up observation
2006	Intermittent cough and hemoptysis	/	Recurrence of ameloblastoma of the right mandible: pathological results suggested potential malignancy	Radical mandible surgery and internal titanium plate fixation
2009	Aggravated cough, intermittent hemoptysis	Enlargement and increased number of lung masses	The patient refused to undergo a repeat lung puncture	Follow-up observation

Year	Respiratory symptom	Chest radiological manifestation	Clinical changes	Treatment
2010	Aggravated cough, intermittent hemoptysis	Continuously increased number of lung masses	Second recurrence of ameloblastoma of the mandible	Curettage of right mandibular lesions under general anesthesia, removal of internal fixation
2016	Aggravated cough, dyspnea, palpitation, unable to leave home, weight loss of ~ 10 kg	Obviously increased number of and enlargement of lung masses	The patient's symptoms worsened, and two lung metastases were diagnosed; the patient refused to undergo lung puncture, and the primary tumor source was unknown	Home oxygen therapy, symptomatic treatment
2018	Frequent cough, hemoptysis, and dyspnea, more serious than those observed in 2016	Extensive mass shadows in both lungs	Bronchoscopic biopsy findings led to the diagnosis of two lung metastases from ameloblastoma (malignant ameloblastoma)	Endotracheal endoscopic interventional therapy: the tumor in the left main trachea was removed by an electric snare, and the symptoms of cough, hemoptysis and dyspnea were significantly improved

We reviewed her chest X-rays (Fig. 1A-1D). Since 1986, X-rays showed that her bilateral lung nodules increases slowly and continuously. The CT at the time of her admission in 2018 revealed soft tissue nodules in the left main trachea and right lower lobe bronchus, with enlarged mediastinal and bilateral hilar lymph nodes (Fig. 2A-2B), and electronic bronchoscopy revealed 90% blockage of the left main trachea and full blockage of the orifice of the right lower lobe basal bronchus by new lesions with abundant surface vasculature (Fig. 3A-3B). Biopsy on this tissue revealed a nest-like structure of tumor cells with surrounding cells exhibiting a high columnar shape and palisade-like arrangement and squamous metaplasia (Fig. 4A). Comparison of the recurrent right mandibular ameloblastoma with a sample resected in 2006 (Fig. 4B) led to a final diagnosis of metastatic ameloblastoma (malignant ameloblastoma [MA]) in the bilateral lungs after misdiagnosis for 32 years. Because she could not tolerate systemic chemotherapy or radiotherapy, interventional bronchoscopic therapy and electroexcision of the left main tracheal tumor were performed 2 months post-diagnosis. An electric snare was connected to a high-frequency electrocautery and ligated to the bottom of the tumor in the left main trachea through the biopsy channel of the electronic bronchoscope. High-frequency electrotomy (power, 40 W for 5–10 s) was used to remove the tumor, and biopsy forceps were used to remove tumor tissue (Fig. 3C-3D). The patient’s respiratory function was satisfactory with an St. George's respiratory disease questionnaire score that fluctuated by > 4 from before to after interventional therapy, indicating a clinically significant result. She died of respiratory failure 1 year later.

Discussion And Conclusions

Since 1923, approximately 100 cases have been reported since[3,4]. In 2005, the WHO defined MA as a histologically benign-appearing ameloblastoma that has metastasized. Ameloblastomas are locally aggressive and rarely metastasize[5]. No standard treatment for MA exists, and most treatment decisions are based on case reports[6,7]. Curettage and radical resection are common treatments, radiotherapy and chemotherapy is recommended for inoperable metastatic lesions; however, its efficacy is unproven and it is considered a palliative treatment[8,9]. In our case, The patient is contraindication for operation due to poor physical condition and she could not tolerate systemic chemotherapy or radiotherapy due to extensive masses in both lungs. Interventional treatment under tracheoscopy can effectively unblock the her airway and relieve respiratory symptoms. The findings from this case suggest that bilateral metastatic tumors should be considered in a patient with nodules or masses in both lungs, and that a careful medical history should be taken.

The mechanism of distant metastasis of ameloblastoma remains unclear; however, this tumor likely spreads via the blood, lymphatic vasculature and airway to local lymph nodes or distant organs, including the lungs, brain, and skin[5,10]. Metastasis usually follows multiple recurrences, frequently affects the lungs (≥ 80%), and may arise more than a decade after primary tumor resection[11–15]. In our case, bilateral pulmonary metastases were detected 12 years after primary tumor resection. However, the case was misdiagnosed for 32 years, likely due to the long asymptomatic period, inert growth of bilateral pulmonary tumors, and a lack of clinical understanding of the disease.

This case demonstrates several points: (1) Patient with a history of ameloblastoma who presents with pulmonary symptoms should be considered for metastases. (2) Electronic bronchoscopic biopsy can be used to confirm a diagnosis of bilateral pulmonary masses with less trauma and good tolerance. (3) Patient with intratracheal metastases of advanced ameloblastoma can achieve symptom relief from an interventional bronchoscopic treatment.

Declarations

Ethics approval and consent to participate：The case report was approved by Wuxi People's Hospital Clinical New Technology and Research Ethics Committee (KS2019008).

Consent for publication: This case report obtained the consent for publication from the patient.

Availability of data and materials： Not applicable.

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Authors' contributions: J.L. drafted the manuscript. S.Y analyzed pathological results. Z.Y. and T.B. analyzed the case and revised the manuscript.

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Figures

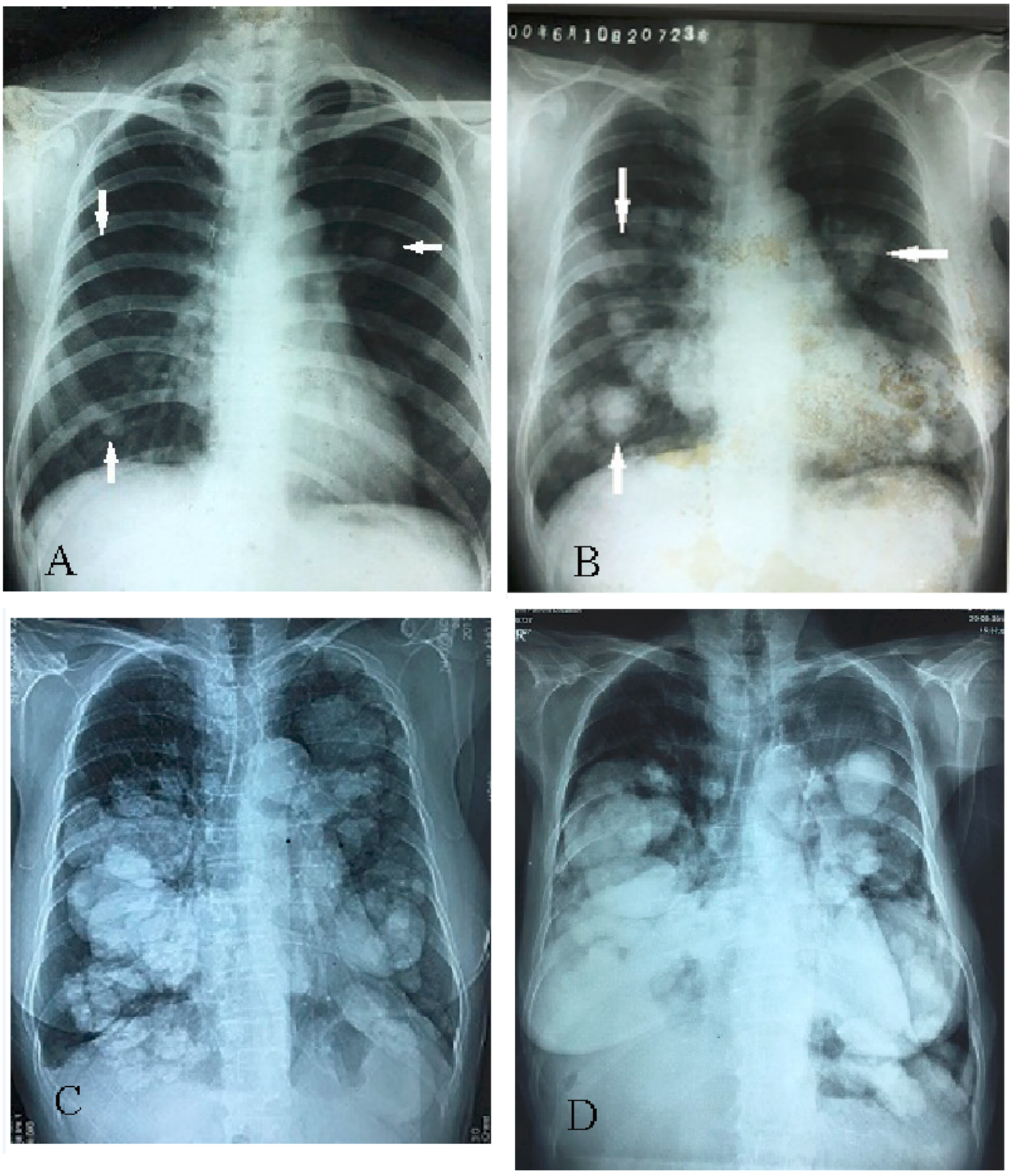

Figure 1

A1. Bilateral lung nodules (arrowheads) (1986). A2. Multiple masses and nodules in the bilateral lungs (arrowheads) (2000). A3. Significant enlargement and increases of the bilateral lung masses (2010). A4. Multiple masses in the bilateral lungs (2018).

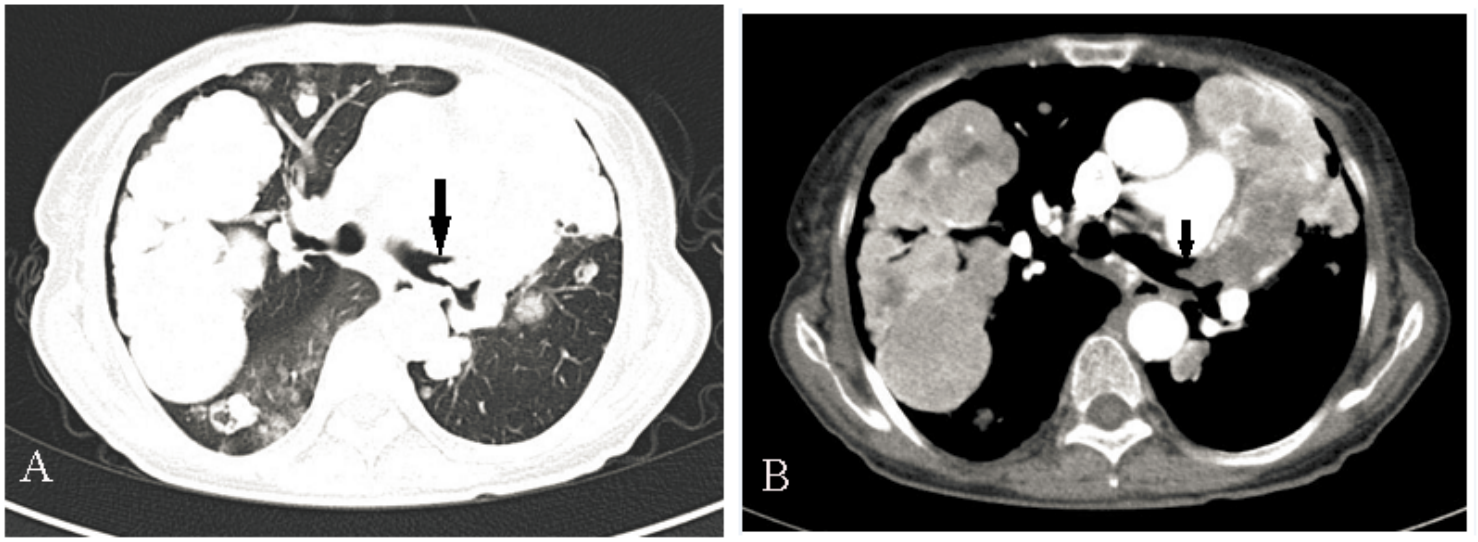

Figure 2

B1. Chest computed tomography (CT) reveals masses and nodules in the bilateral lungs and an abnormal density in the left main trachea. B2. Chest CT reveals areas of abnormally dense opacity in the left main trachea (black arrowheads).

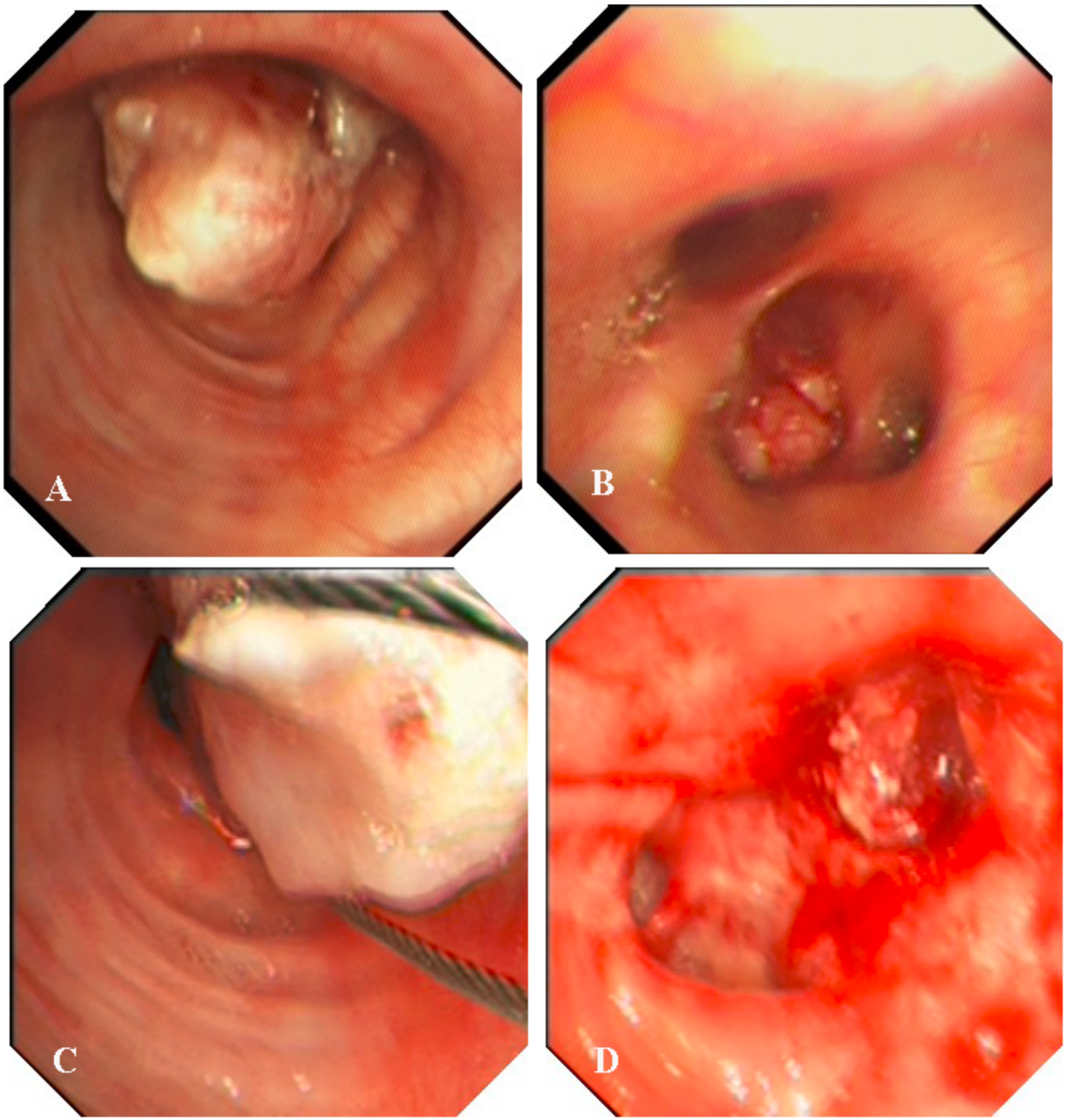

Figure 3

C1. New lesions in the left main trachea caused a 90% blockage. C2. New lesions in the basal bronchus of the right lower lobe, with abundant surface vasculature. C3. Electroexcision of the left main tracheal tumor under bronchoscopic guidance. C4. Trachea unobstructed after resection of the lesions from the left main trachea.

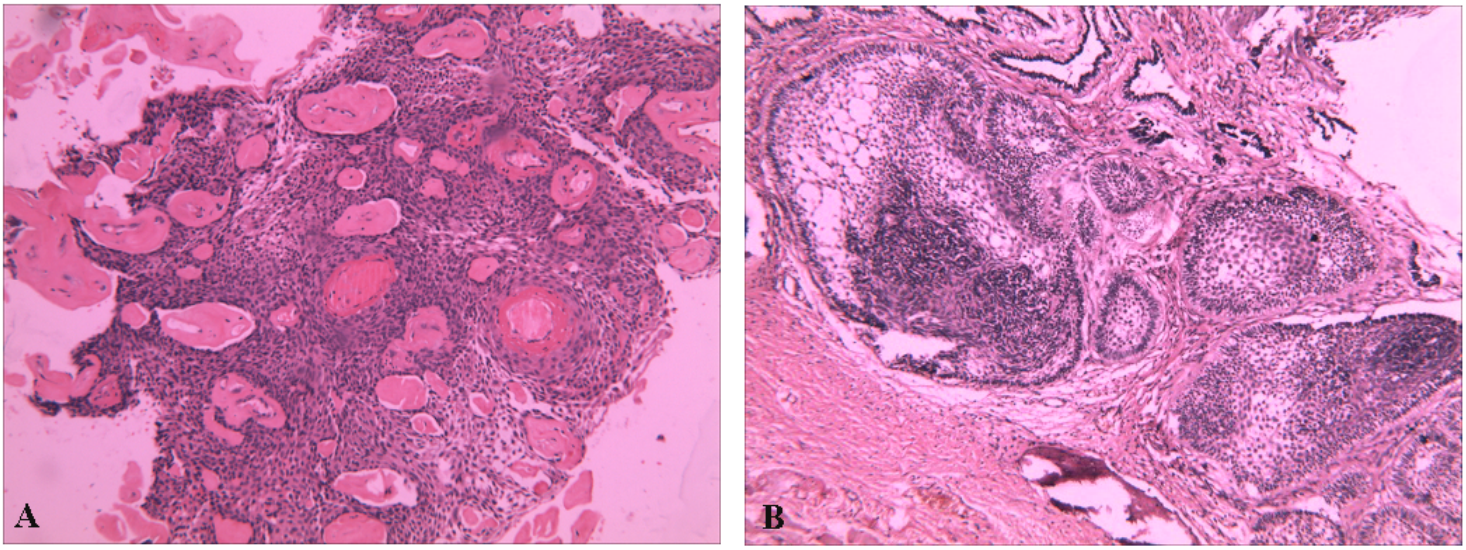

Figure 4

D1. Pathological analysis of newly biopsied tracheal lesions in 2018 revealing nest-like structures of tumor cells, with some surrounding cells exhibiting a high columnar shape and palisade-like arrangement with squamous metaplasia. The focal area exhibits a sparse reticular structure, with stellate or fusiform cells and angiogenesis. Immunohistochemistry revealed the following: Napsin A (-), thyroid transcription factor (TTF)-1 (-), creatine kinase (CK)7 (-), CK56 (+), P63 (+), and Ki-67 (15% +). D2. Pathological analysis of tumor cells resected in 2006 similarly revealing a nest-like structure of tumor cells, with some surrounding cells exhibiting a high columnar shape and palisade-like arrangement. The focal area exhibits a sparse reticular structure with stellate or fusiform cells. Pathological sections of the tracheal lesions were compared with those of the right mandibular ameloblastoma obtained 12 years earlier. The final pathological diagnosis of the new tracheal lesions was metastatic ameloblastoma (malignant ameloblastoma) in the bilateral lungs. Staining: hematoxylin-eosin unless otherwise indicated. All magnification: 100×.

Supplementary Files

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