



## Introduction

Pindborg tumor, also known as a calcifying epithelial odontogenic tumor (CEOT), is a rare, typically benign but locally invasive neoplasm. We report a case of an exceedingly aggressive, recurrent CEOT of maxillary origin, with malignant transformation, orbital extension, and pulmonary metastasis.

## Educational Objectives

1. Understand the key characteristics of CEOT
2. Develop an appreciation for the potential aggressive nature of CEOT
3. Review the important features and strategies of managing CEOT

## Case Report

- 71-year-old male
- 17 prior surgeries for recurrent CEOT since diagnosis in 1995
- Presented with new onset diplopia
- Prior pathology concerning for malignant transformation
- Imaging revealed tumor recurrence in the left maxillary sinus with extension superiorly into the orbital floor, inferiorly into the remnants of the pterygopalatine fossa, laterally into the medial most components of the infratemporal fossa, and posteriorly to the skull base at the fovea ethmoidalis (Fig. 1)
- Patient opted for re-resection without orbital exenteration followed by adjuvant external beam radiation therapy despite the high likelihood of subtotal resection
- Surgery was performed by otolaryngology in collaboration with oculoplastics via a combined transconjunctival, transoral, and endoscopic endonasal approach (Fig. 3)
- Orbital preservation necessitated leaving positive margins along the inferior rectus muscle and medial orbital apex
- Two years post-surgery:
  - Minimal vision or movement in the affected eye
  - Reassuring local tumor control (Fig. 2)
  - PET and subsequent biopsy revealed lung metastasis (Fig. 4)
  - Pathology showed PDL-1 positivity, started on pembrolizumab
  - Wedge resection and adjuvant radiation and chemotherapy with continued progression of metastatic disease

Figure 1. Preoperative T1-weighted coronal, axial, and sagittal view of tumor

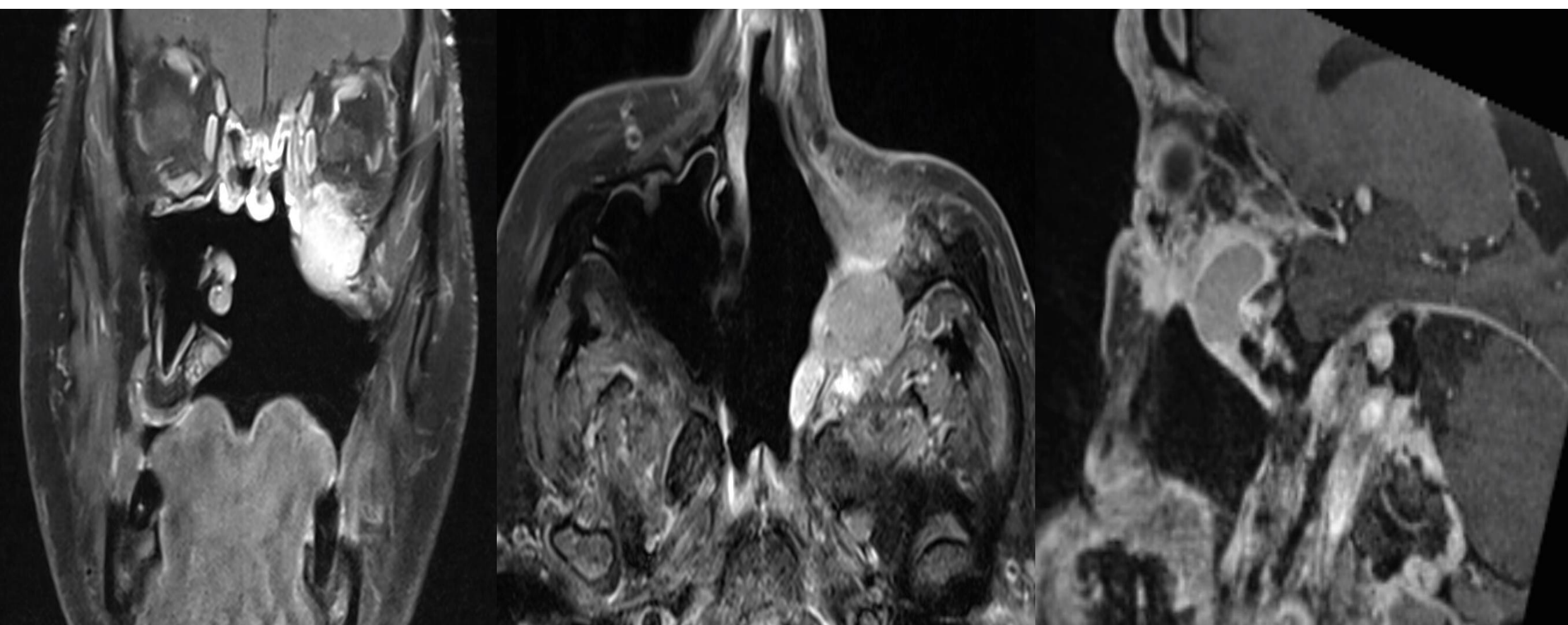


Figure 2. Two-year postoperative T1-weighted coronal, axial, and sagittal view

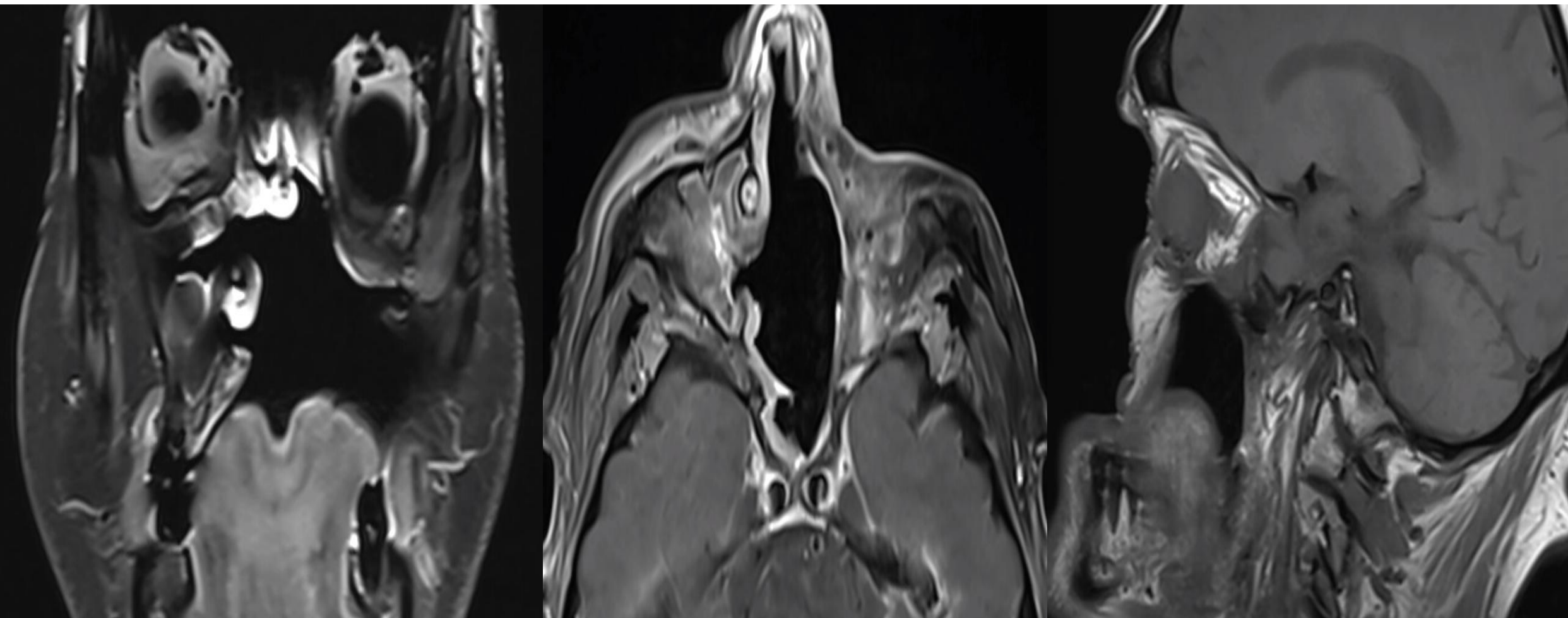


Figure 3. Intraoperative transoral endoscopic view of tumor

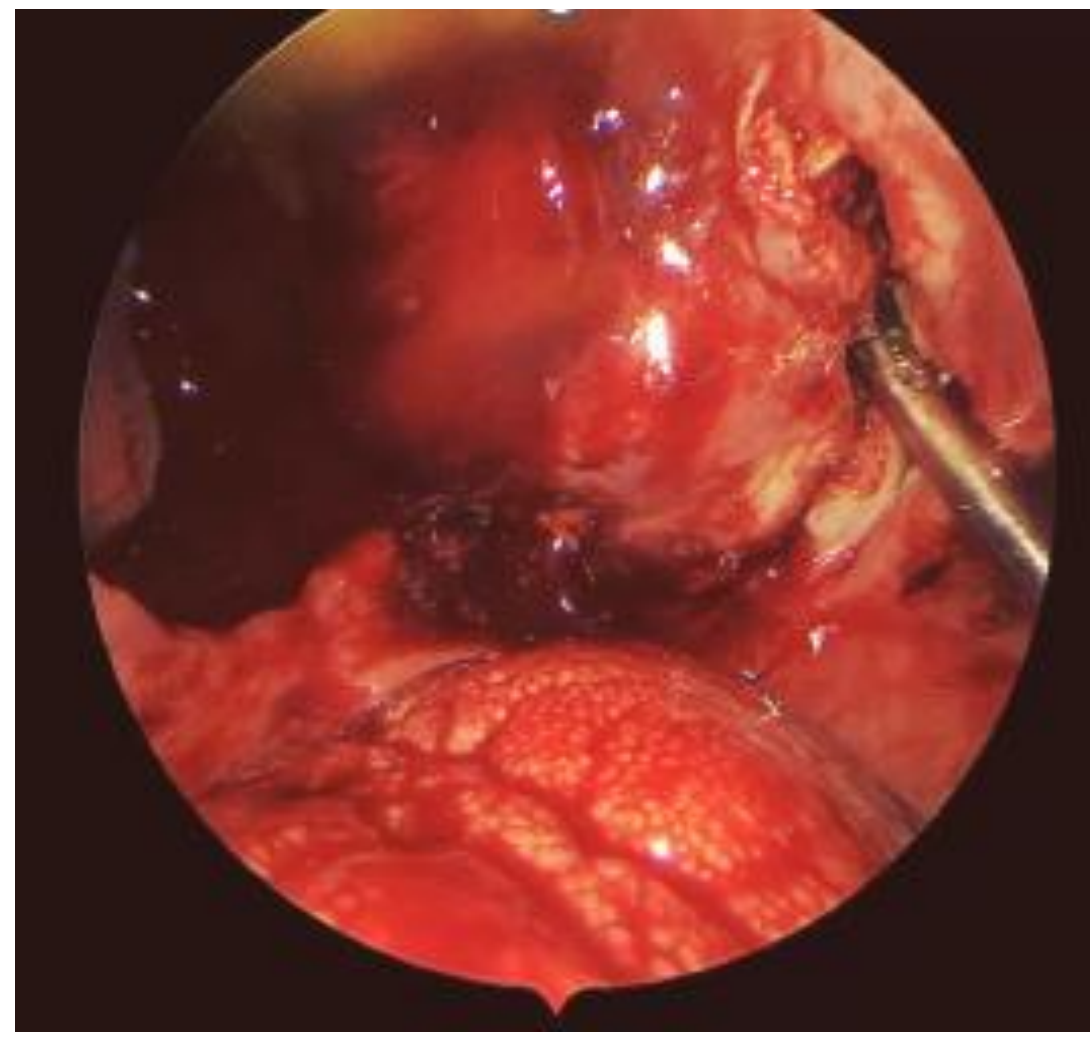


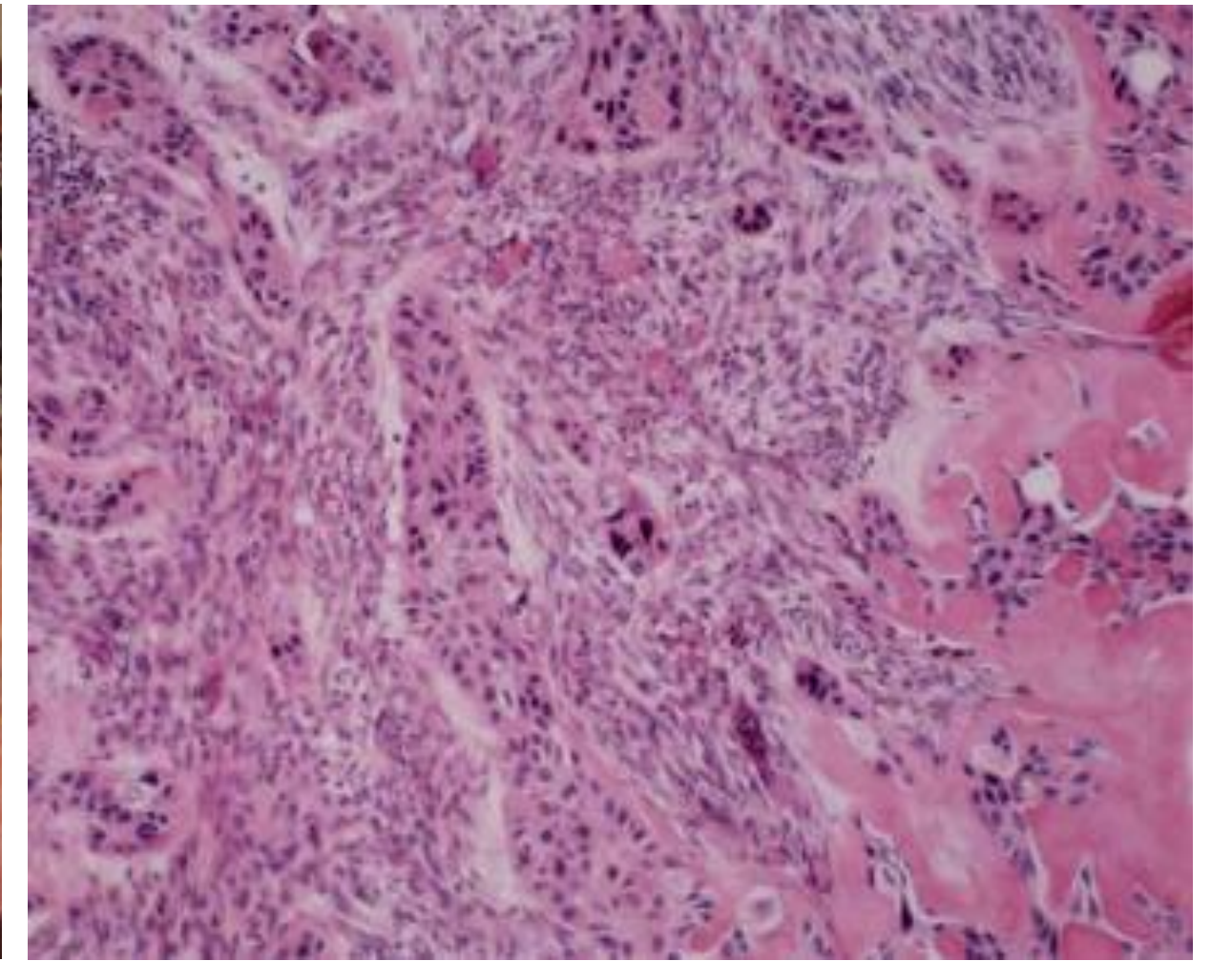
Figure 4. Axial view chest CT of pulmonary metastatic disease



Figure 5. External view of CEOT of maxillary origin<sup>1</sup>



Figure 6. CEOT pathology slide with microscopic features of malignant behavior<sup>2</sup>



Figures 5 and 6 are educational examples from the literature, not from the presented case.

## Discussion

- Calcifying epithelial odontogenic tumor (CEOT) or Pindborg tumor
- First described in 1956 by Danish pathologist Dr. Jens J. Pindborg<sup>3</sup>
- Exclusively of odontogenic epithelial origin<sup>4</sup>
- Likely arising from the stratum intermedium layer of the enamel organ in tooth development or the remnants of the primitive dental lamina<sup>5</sup>
- Mean age of 38.1 years<sup>6</sup>
- No significant gender differences<sup>6</sup>
- Constituting <1% of odontogenic neoplasms<sup>4</sup>
- Approximately <400 reported cases<sup>6</sup>
- Mandible is most common site (2:1 vs. maxilla) (Fig. 5)<sup>7</sup>
- Locally invasive, typically benign
- 10 cases of malignant transformation, more common from the mandible (Fig. 6)
- Treatment is surgical resection with radiation for malignant transformation
- 12.6% chance of recurrence, 42.9% chance with malignant transformation<sup>8</sup>
- 2 cases of orbital involvement
- 2 cases of pulmonary metastasis
- Presentation of CEOT in this case is exceedingly rare, and its aggressive nature necessitated a multidisciplinary treatment approach and close follow-up

## Conclusions

- CEOTs are rare, odontogenic neoplasms, often originating from the mandible
- While they are typically benign, there is potential for local aggression, malignant transformation, and recurrence
- Orbital extension and metastasis are possible
- Treatment involves aggressive surgical resection with or without radiation and may require a multidisciplinary approach
- Early diagnosis and intervention are crucial
- Recurrent or malignant cases should be followed closely for metastasis

## Contact

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