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Journal of Medical Case Reports (2025) 19:135
https://doi.org/10.1186/s13256-025-05041-4

Journal of
Medical Case Reports

CASE REPORT

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Metastatic choroidal pulmonary biphasic blastoma as a unique single initial metastasis to the eye: a case report

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Abstract

Background Choroidal metastasis is the most common intraocular malignancy in adults, commonly presenting in later stages of the disease and associated with poor prognosis. This is the first case to describe choroidal involvement as the sole initial metastasis of the rare biphasic pulmonary tumor.

Case presentation A 42-year-old white male patient presented to the emergency department with 3 weeks of progressive temporal visual field loss in the left eye (oculus sinister) with slight discomfort to palpation. At the emergency department, he underwent computed tomography and computed tomography angiography of the brain to rule out a central nervous system etiology of his vision loss. The imaging demonstrated lentiform dependent hyperdense material with layering hypodensity within the medial aspect of the left globe, consistent with choroidal detachment. His prior medical history was significant for biphasic pulmonary blastoma cT2N2M0 stage IIIA involving the right lung, with extension into the superior vena cava, diagnosed 3 years prior to the current presentation. He was treated with chemoradiation and excision with complete response to therapy.

Conclusion As the survival rates for cancer patients are increasing owing to better quality and more available treatments, choroidal metastasis may become more common. Early diagnosis and effective treatment of these lesions is crucial for better patient outcomes.

Keywords Choroidal metastasis, Biphasic pulmonary blastoma, Cancer, Choroid, Metastasis

Background

Owing to its profuse vascular supply, the choroid is considered the most common intraocular site for metastatic disease [1]. Breast cancer (40–47%) followed by lung cancer (21–29%) are the most common primary cancers that metastasize to the choroid [1]. Breast cancer usually leads to bilateral multifocal metastases, as opposed to lung cancer, which typically presents as unilateral and unifocal. The first case of choroidal metastasis secondary to lung disease was reported in 1872 by Perls *et al.* [2]. The treatment of these lesions varies, depending on the systemic burden of the disease, the status of the patient, and the number, location, and laterality of choroidal metastasis. Observation of ocular metastasis can be considered in an asymptomatic patient. In the case of active sites of metastasis, or whether the tumors are multifocal and bilateral, systemic chemotherapy, immunotherapy, hormone therapy, or whole eye radiotherapy is preferred. For single and active metastasis, plaque radiotherapy, transpupillary radiotherapy, or photodynamic therapy can be considered. If the extension of the tumor leads to a blind and painful eye, then enucleation is considered.

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Pulmonary blastoma is a rare and malignant neoplasm that makes up 0.25–0.5% of all primary lung tumors [3]. It tends to have poor prognosis and is considered very aggressive. These tumors morphologically resemble fetal lung tissue developing from a pulmonary blastema. They exhibit both mesenchymal and epithelial components in histology, and therefore are named biphasic. Initially, the term pulmonary blastoma was used to include fetal adenocarcinomas, pleuropulmonary blastomas, as well as biphasic blastomas, but the World Health Organization (WHO) reclassified these tumors and separated these three categories. This reclassification, along with the rarity of this tumor, makes any interpretation of the published literature challenging [3]. Despite its assumed embryonal origin, the tumor predominantly affects adults.

There are very few reports on biphasic blastoma and ocular metastasis. Shields reported a case of a ciliary body medullo-epithelioma in association with pleuropulmonary blastoma and demonstrated a germline mutation in the *DICER1* gene [4]. Costa *et al*. reported an intraocular metastasis in a 3-year-old from pleuropulmonary blastoma [5]. This study's case is the first case to report uveal metastasis of biphasic pulmonary blastoma as sole initial metastasis in an adult patient. This work importantly raises awareness for the need for thorough ocular examinations to capture possible unexpected metastases in patients with biphasic blastoma who develop ocular symptoms.

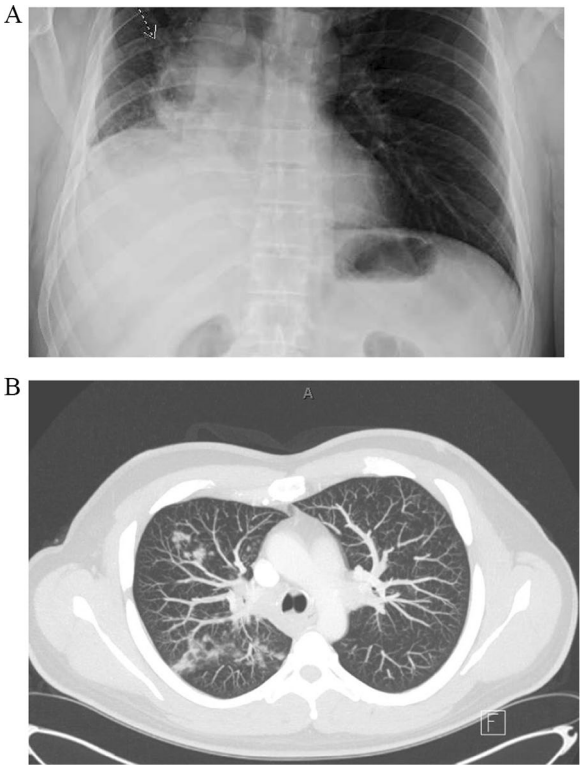


Fig. 1 **A** Chest X-ray showing volume loss at the right pulmonary base, with the appearance of a cavitary lesion in the right midlung zone (arrow), and prominent tissue to the right of the trachea; **B** Computed tomography of the chest revealing right hilar/suprahilar mass with nonocclusive extension into the superior vena cava as well as peribronchovascular patchy nodular areas of ground-glass opacity, mainly in the right upper lobe

Case presentation

A 42-year-old white male presented to the Lahey emergency department (ED) with 3 weeks of progressive vision loss and temporal visual field loss of the left eye (oculus sinister, OS) and slight discomfort on palpation. His past ocular history was noncontributory, and his past medical history was significant for pulmonary blastoma cT2N2M0, stage IIIA involving the right lung, with extension into the superior vena cava (SVC, Fig. 1), 3 years prior to his current presentation. He was treated with chemoradiation and right pneumonectomy with complete response to therapy. He had no first-degree relatives with a history of cancer nor other relevant ocular disease. He had no smoking history and drank alcohol very sparingly, roughly twice per year. In the ED, a computed tomography (CT) scan of the head and computed tomography angiography (CTA) were obtained to rule out a neurologic source of the vision loss. The study showed a lentiform dependent hyperdense material with layering hypodensity within the medial aspect of the left globe, consistent with choroidal detachment (Fig. 2). No other acute intracranial process was seen, and there was no evidence of acute infarction, hemorrhage, or mass.

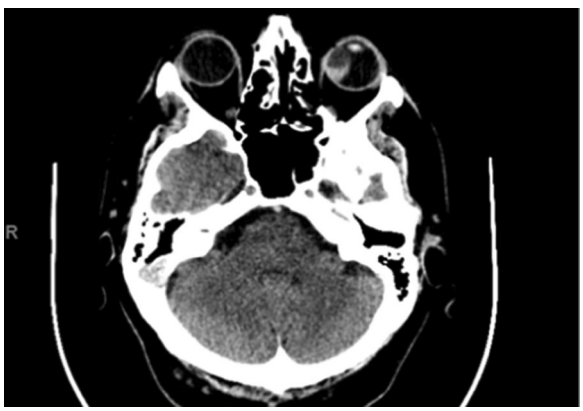


Fig. 2 Computed tomography angiography of the brain, axial cut, showing a lentiform dependent hyperdense material with layering hypodensity within the medial aspect of the left globe consistent with choroidal detachment; no acute intracranial process, no evidence of acute infarction, hemorrhage, or masses

The CTA did not show any intracranial aneurysm or other neurovascular abnormality. When the patient was referred to ophthalmology, he was only able to count fingers in the affected eye at distance, but his near visual acuity was intact at 20/30. At that point, a myopic shift was suspected and confirmed with autorefraction. His intraocular pressure (IOP) was normal at 13 mmHg. His ocular motility was orthophoric and full. His pupils were equal, round, and reactive, and there was no afferent pupillary defect. There was a left temporal visual field loss present to confrontational testing. His anterior chamber examination appeared shallow in the affected eye, later found to be due to choroidal effusions pushing the anterior chamber complex forward. The posterior pole examination in the left eye demonstrated a round, elevated, red-hued lesion which was believed to be a serous hemorrhagic choroidal detachment (Fig. 3). Visual acuity of the right eye was 20/20, and examination of the anterior and posterior poles was normal.

B-scan ultrasonography of the left eye was obtained, demonstrating medium-to-high internal reflectivity and a fluid level within a large elevated nasal lesion (Fig. 4). Subsequently, fluorescein angiography (FA) of both eyes was performed. The right eye was unremarkable, but in the left eye, a large nasal choroidal lesion was identified and found to block dye uptake (Fig. 5A–E). On the basis of this presentation, a metastatic choroidal lesion linked

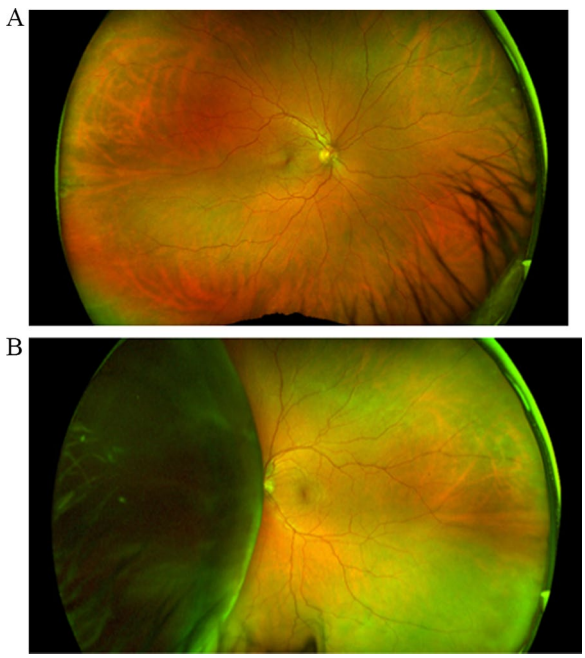


Fig. 3 **A** Optos color fundus picture of the right eye presenting as normal; **B** optos color fundus picture of the left eye, showing a reddish hue, perfectly round, serous hemorrhagic choroidal detachment versus a mass

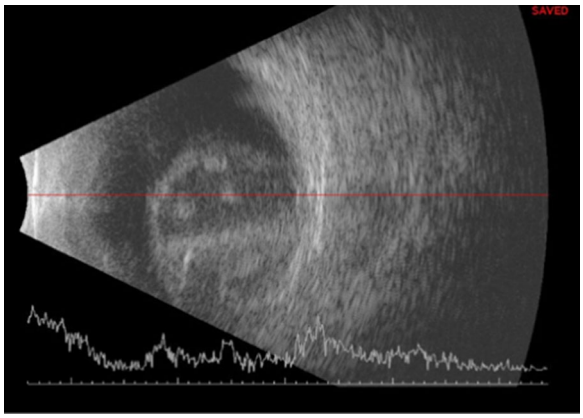


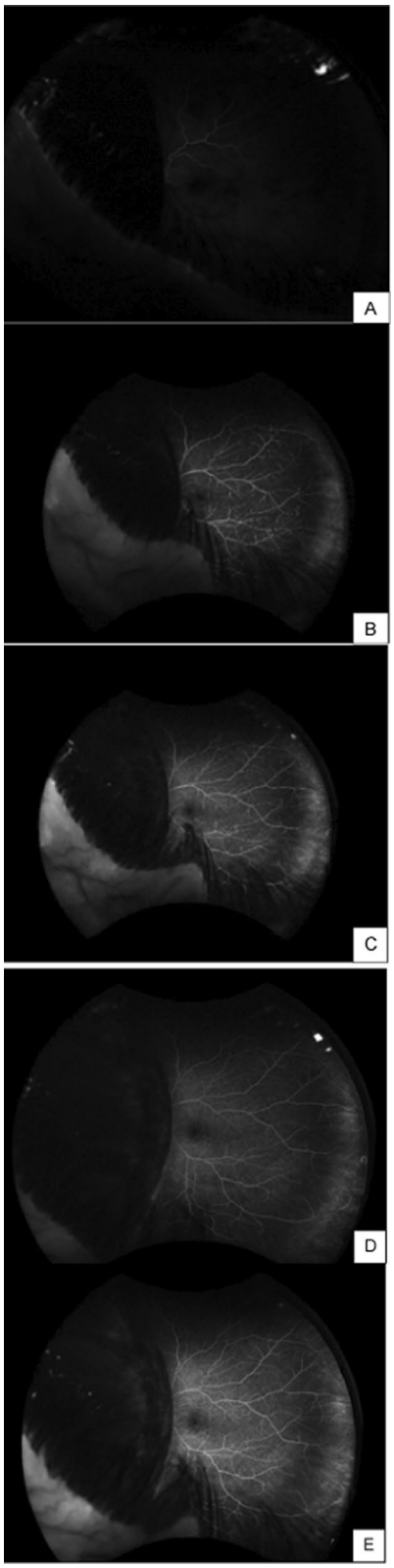
Fig. 4 B-scan ultrasonography of the left eye; medium-to-high internal reflectivity fluid level seen within large elevated nasal lesion

to his pulmonary biphasic blastoma was very high in the differential diagnosis, but a *de novo* tumor could not be excluded. Given the lack of trauma, previous surgery, or any other known ocular history such as glaucoma, a hemorrhagic etiology of choroidal effusions was unlikely. The absence of inflammation and negative B-scan findings made the diagnosis of posterior scleritis less likely.

As the suspicion for metastasis was high, a positron emission tomography (PET) scan was obtained, which was negative for metastasis. A repeat CT scan did not demonstrate any intracranial metastases. His near vision was still acceptable at 20/30, thus the patient and team opted to monitor the lesion closely. However, at a later visit, the nasal mass increased in size with an increase in subretinal fluid. In this case, there were no financial, language, or cultural barriers present. Therefore, a decision was made to proceed with an intraocular fine needle aspiration biopsy, which confirmed the suspicion of metastasis from lung origin (Fig. 6). A multidisciplinary team recommended against the need for further systemic treatment.

A total of 3 weeks after the initial presentation, the subretinal fluid increased significantly, accompanied by submacular fluid, further compromising the patient's vision. A short course of steroids was initiated to reduce the ocular inflammation and fluid. As the choroidal detachment enlarged, the vision continued to worsen due to forward shifting of the ciliary body and lens complex, ultimately causing angle closure and significant pain.

On the basis of the worsening clinical presentation and the development of a blind and painful eye, the decision was made to proceed with enucleation. On histopathologic examination, the choroid was infiltrated by a metastatic epithelial tumor, which predominantly showed the typical epithelial component of pulmonary blastoma with glandular formation (Fig. 7). The stromal component was



◀ **Fig. 5** **A**–**E** Fluorescein angiography of the left eye showing a large choroidal lesion nasally OS blocking dye uptake; each panel shows subsequent timepoints as follows: **A** 00:17 seconds, **B** 00:29 seconds, **C** 00:48 seconds, **D** 1:15 seconds, **E** 1:29 seconds

not present. The glandular epithelial cells were strongly positive for pan-cytokeratin and thyroid transcription factor 1 (TTF1) immunohistochemical stains. The findings were consistent with the diagnosis of metastatic pulmonary blastoma.

Discussion and conclusion

Pulmonary blastoma is a rare pulmonary malignancy first described in 1952 by Barnard, who initially characterized it as an embryoma [6]. Later on, it was reclassified as blastoma by Spencer [7]. The World Health Organization has classified the pulmonary blastoma into three

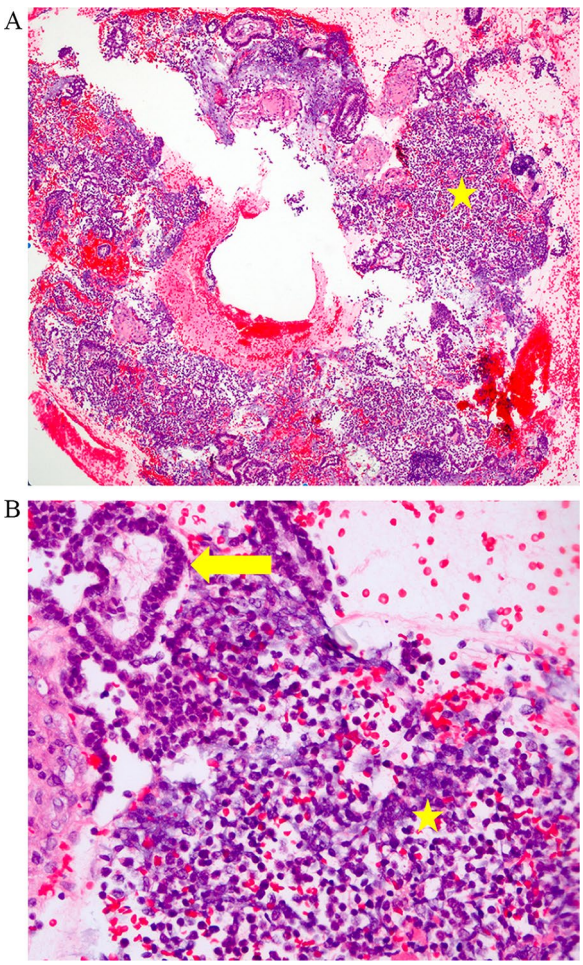
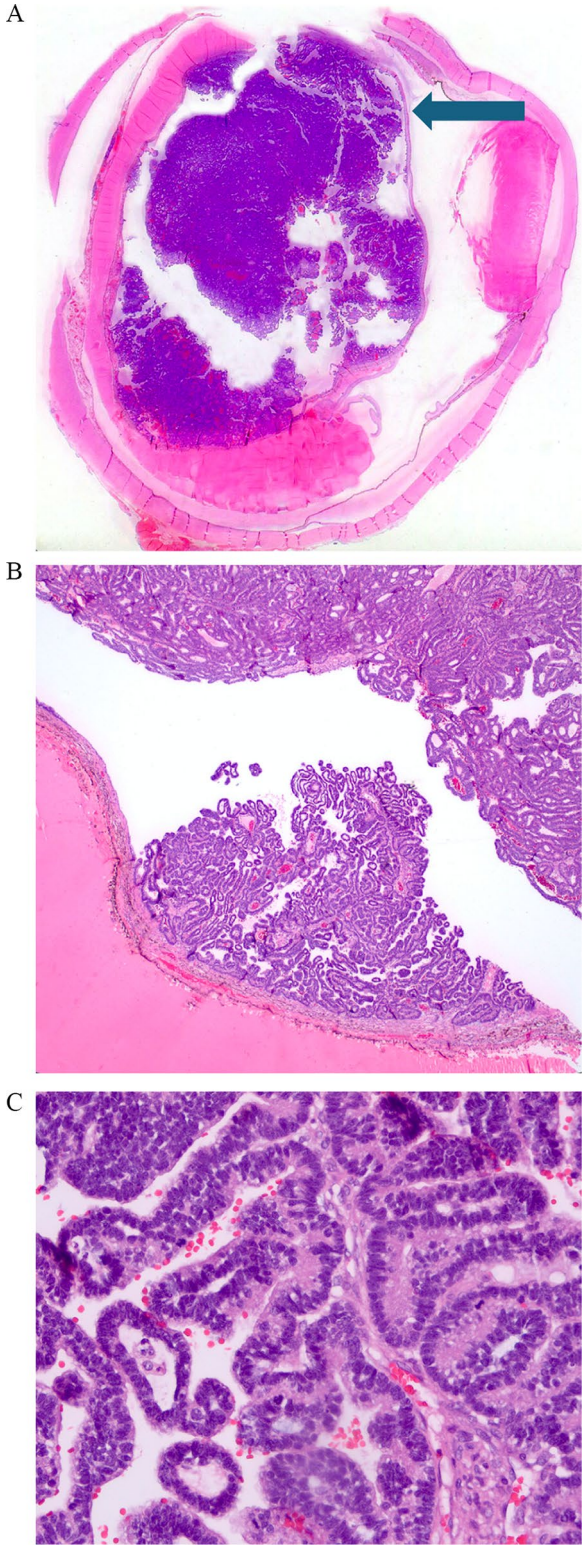


Fig. 6 Hematoxylin and eosin stain of the choroidal tissue biopsy demonstrating both the epithelial (yellow arrow) and mesenchymal (yellow star) component of the tumor; **A** 4× magnification, **B** 20× magnification



◀ **Fig. 7** **A** Hematoxylin and eosin stain of enucleated eye, showing retinal detachment (blue arrow), hemorrhage, and epithelial component (yellow arrow) of the metastatic cells, **B** 2× magnification of hematoxylin and eosin stain, **C** 20×, hematoxylin and eosin showing the glandular conformation of cells that resemble immature pulmonary tissue

groups: biphasic pulmonary blastoma (BpB), well-differentiated fetal adenocarcinoma (WDFa), and pleuropulmonary blastoma (PPB) [8]. The biphasic pulmonary blastoma is composed of two cell types, an immature epithelial component and a mesenchymal type that resembles the immature bronchi and connective tissue of the embryonic lung. Little is known about the genetic basis of biphasic pulmonary tumors. Some *p53* mutations that were identified were found to be similar to the *p53* mutations found in other lung cancers [9]. The clinical presentation varies from asymptomatic to nonspecific symptoms of pulmonary disease such as cough, hemoptysis, dyspnea, chest pain, respiratory distress, fever, anorexia, and weight loss [10]. This study's patient had progressive and persistent wheezing and shortness of breath during exertional work. The management and treatment of PpB is not well established given the rarity of the tumor; it includes surgical excision, radiation, or a combination of chemoradiation for selected cases. There is a lack of data to show whether adjunctive therapy might be beneficial.

The liver, bone, and central nervous system (CNS) are the most common sites of metastases. Overall prognosis is poor, with a 5-year survival of approximately 16% [11]. There are very few cases of pulmonary blastomas that have been associated with intraocular tumors. Shields and Hubbard reported a case of pleuropulmonary blastoma with a germline mutation of *DICER1* giving off a metastasis to a ciliary body meduloepithelioma [4]. However, to the authors' knowledge, there are no cases that report ocular metastasis secondary to the rare biphasic pulmonary blastoma. Choroidal metastasis manifests late during the disseminated stage of disease, which makes it a poor prognostic sign when present [1]. This study's case describes a patient who was treated for biphasic pulmonary blastoma and 3 years later developed metastasis to the choroid as the sole initial presentation (Fig. 8).

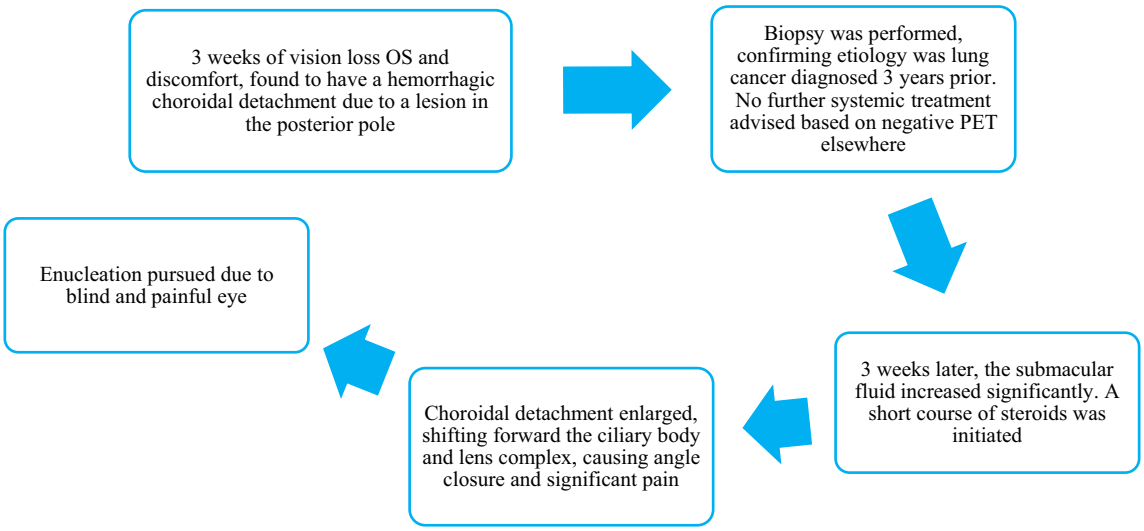


Fig. 8 Timeline and summary of the case presentation, beginning with symptom onset 3 years after the initial lung cancer was diagnosed

Abbreviations

ED	Emergency department
OS	Left eye (oculus sinister)
CT	Computed tomography
CTA	Computed tomography angiography
CNS	Central nervous system
SVC	Superior vena cava
IOP	Intraocular pressure
FA	Fluorescein angiography
WHO	World Health Organization
PET	Positron emission tomography
BpB	Biphasic pulmonary blastoma
WDFa	Well-differentiated fetal adenocarcinoma
PPB	Pleuropulmonary blastoma

Acknowledgements
Not applicable.

Author contributions
All authors were involved in data extraction for the patient. All authors were involved in writing, editing, reading, and approving the final manuscript.

Funding
Funding for publication was sourced from the authors of the publication.

Availability of data and materials
Not applicable.

Declarations

Ethics approval and consent to participate
Not applicable.

Consent for publication
Written informed consent was obtained from the patient's next-of-kin for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests
The authors declare that they have no competing interests.

Received: 2 October 2024 Accepted: 22 November 2024
Published online: 24 March 2025

References

1. Arepalli S, Kaliki S, Shields CL. Choroidal metastases: origin, features, and therapy. Indian J Ophthalmol. 2015;63(2):122–7. https://doi.org/10.4103/0301-4738.154380.
2. Perls M. *Beitrage zur geschwulstlehere*. Virchows Arch. 1872;1:437–67 (**abstract only**).
3. Smyth RJ, Fabre A, Dodd JD, *et al*. Pulmonary blastoma: a case report and review of the literature. BMC Res Notes. 2014;7:294. https://doi.org/10.1186/1756-0500-7-294.PMID:24885892;PMCID:PMC4026592.
4. Kramer GD, Arepalli S, Shields CL, Shields JA. Ciliary body medulloepithelioma association with pleuropulmonary blastoma in a familial tumor predisposition syndrome. J Pediatr Ophthalmol Strabismus. 2014;51:e48-50. https://doi.org/10.3928/01913913-20140709-03.
5. Costa J, Klijanienko J, Desjardins L, Cassoux N, Machet MC, Pacquement H. Fine needle aspiration in intraocular metastasis from pleuropulmonary blastoma. A case report and a review of the literature. Diagn Cytopathol. 2017;45(2):156–60. https://doi.org/10.1002/dc.23618.
6. Barnard WG. Embryoma of lung. Thorax. 1952;7(4):299–301.
7. Spencer H. Pulmonary blastomas. J Pathol. 1961;82:161–6. https://doi.org/10.1002/path.1700820119.
8. Travis WD, Brambilla E, Nicholson AG, *et al*. The 2015 World Health Organization Classification of lung tumors: impact of genetic, clinical and radiologic advances since the 2004 classification. J Thorac Oncol. 2015;10(9):1243–60. https://doi.org/10.1097/JTO.0000000000000630.
9. Bodner SM, Koss MN. Mutations in the p53 gene in pulmonary blastomas: immunohistochemical and molecular studies. Hum Pathol. 1996;27(11):1117–23. https://doi.org/10.1016/s0046-8177(96)90302-0.
10. Shah SU, Mashayekhi A, Shields CL, *et al*. Uveal metastasis from lung cancer: clinical features, treatment, and outcome in 194 patients. Ophthalmology. 2014;121(1):352–7. https://doi.org/10.1016/j.ophtha.2013.07.014.
11. Siegel RL, Miller KD, Jemal A. Cancer statistics, 2020. CA Cancer J Clin. 2020;70(1):7–30. https://doi.org/10.3322/caac.21590.

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