ASIDE Case Reports

Case Report

Lamellar Intraocular Ossification encasing the Optic Nerve Head: A Case Report

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ABSTRACT

Intraocular ossification is a rare end-stage sequela of chronic retinal detachment, inflammation, trauma, or phthisis bulbi. It typically presents as minor localised choroidal or retinal ossification. We report an exceptionally rare case of mature lamellar intraocular bone encasing the optic nerve head. A 23-year-old male presented three days after sustaining a knife injury to the right eye. The eye had been blind with no light perception for approximately five years prior, with a history of longstanding visual impairment since childhood. Examination revealed an open globe with corneal-limbal laceration and prolapse of uveal and lenticular tissue. The open-globe injury and extensive tissue prolapse precluded posterior segment assessment and B-scan ultrasonography. The contralateral eye demonstrated a morning glory disc anomaly with a shallow chronic retinal detachment, which was treated conservatively. During routine evisceration, a near-round 12 mm lamellar bony mass was identified encasing the optic nerve head, forming a central invagination in which the optic nerve was seated. The mass felt like bone and was non-adherent; it was removed intact. Post-operative CT imaging showed no additional intra-orbital ossification or malignancy. Histopathology demonstrated mature lamellar bone with osteoblastic rimming, marrow spaces containing adipocytes, and psammomatous calcifications, consistent with long-standing heterotopic ossification. No malignant features or organisms were identified. The postoperative course was uncomplicated, and the patient remained clinically well at six months' follow-up. This case represents an exceptionally rare report of mature lamellar intraocular bone fully encasing the optic nerve head, highlighting the remarkable metaplastic potential of intraocular tissues in chronic retinal pathology.

1. Introduction

Intraocular ossification is a rare end-stage sequela of long-standing retinal detachment, inflammation, or trauma, and may manifest as osseous metaplasia or heterotopic bone formation within the globe [1]. Several clinicopathologic series of enucleated eyes have reported osseous metaplasia in 3% (n = 3,833) [2], 4.8% (n = 2,486) [3], and 5.2% (n = 151) [4] of eyes. This highlights its uncommon occurrence, even in globes removed for chronic ocular pathology rather than in the general population. Sparse case reports describe similar peri-papillary and choroidal tumours, but these are exceptionally rare. These described cases do not encompass the optic nerve, nor do they contain such substantial bony elements as described in this case [5, 6, 1]. The retinal pigment epithelium (RPE) and glial tissue are considered a multipotent cell layer, and are known to undergo mesenchymal transdifferentiation into osteoblast-like cells under pathological stimuli such as chronic inflammation or detachment [6, 1]. Glial tissue, particularly Müller cells, has also been shown to undergo osseous metaplasia, via reactive gliosis, but this typically occurs in conjunction with, rather than independently of, RPE-derived transdifferentiation [4]. While often associated with phthisical eyes, ossification may also occur in eyes without overt phthisis [4]. Here, we report an unusual case of intraocular lamellar bone formation encased around the optic nerve head. It was incidentally discovered during a routine evisceration for a traumatic anterior globe injury. To our knowledge, ossification of this size encasing the optic nerve head has not been reported, based on a targeted PubMed and Google Scholar search using relevant terms.

2. Patient Presentation

A 23-year-old male presented three days after an assault with a sharp, horizontal knife injury to the right eye. He reported longstanding poor vision in both eyes since birth, with the right eye blind (no light perception, NLP) for approximately five years prior to the trauma. He also recalled a prior evaluation in childhood for bilateral congenital ocular abnormalities, although medical records were unavailable. He had no history of prior ocular infections or surgeries, no systemic comorbidities, and was not on any regular medications. On presentation, the injured right eye had NLP, while the left eye could count fingers (CF) at 3 meters. Anterior segment examination revealed intact eyelid margins and a corneal-limbal laceration extending from 5 to 8 o'clock, with prolapse of the lens and uveal tissue preventing full eyelid closure. No posterior segment structures were visualized, and prolapsed ocular contents precluded ultrasound imaging. There were no clinical signs of infection, and given the urgent need for surgical intervention, the index clinician did not administer prophylactic antibiotics.

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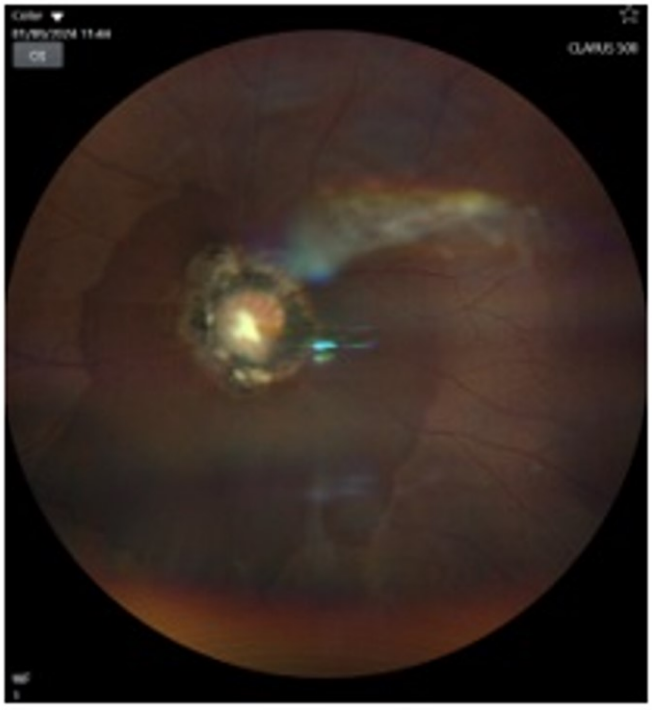

Figure 1: Contralateral eye fundus photo showing MGDA with shallow retinal detachment. Visual acuity was CF at 3m.

The contralateral eye was diagnosed with a morning glory disc anomaly (MGDA) associated with a shallow, chronic retinal detachment. Given the longstanding visual impairment, absence of acute symptoms, and poor visual prognosis, no surgical or prophylactic retinal intervention was undertaken. In our public-sector setting, access to vitreoretinal services is limited and prioritised for patients with a reasonable expectation of visual improvement. The patient was counselled regarding the guarded prognosis, educated about potential progression symptoms, and advised to continue routine clinical follow-up.

Based on this history and the confirmed presence of MGDA in the left eye, it was considered plausible that the right eye had also been affected; however, this remains speculative and represents a limitation of the study due to the absence of confirmatory pre-injury documentation or imaging. On examination, the right eye demonstrated an anterior globe rupture with prolapse of uveal and lens tissue. In contrast, the left eye showed a classic MGDA with a shallow chronic retinal detachment **(Figure 1)**. Although unconfirmed, the history suggests bilateral MGDA, which itself is a very rare entity, approximately 15.9% of MGDA cases [7].

No B-scan was performed due to the extent of uveal components prolapsing through the open globe. Due to resource limitations and prolonged waiting times, a pre-operative CT scan could not be performed. Evisceration of the right globe was performed 8 hours after presentation to the ophthalmology department. Intraoperatively, a 12 mm near round, lamellar bony mass was incidentally discovered encasing the optic nerve head **(Figure 2)**. The optic nerve measured 5 mm in diameter with a 1 mm thick sheath (measured and photographed fresh intra-operatively, immediately after excision). The bony element was non-adherent within the scleral shell and was easily removed without any additional surgical manoeuvres. The bony mass spontaneously separated from the nerve, revealing a central channel where the optic nerve was attached. The entire sclera was routinely divided into two segments to allow passage of the silicone implant into the orbit. The bony sphere was solid, with an invagination where the optic nerve entered; the opening was around 1.5mm before ending blindly. The optic nerve was excised as proximally as possible to allow inclusion in histological evaluation. There were no intraoperative features suggestive of malignancy, and the mass appeared grossly benign. A 20 mm non-porous silicone orbital implant, the only size and type available at our institution, was placed, and the scleral shell was closed. Due to diagnostic uncertainty, a postoperative CT was performed to evaluate the remaining orbit for osseous or malignant lesions. The postoperative CT scan revealed no other intra-orbital bony elements or malignancies **(Figure 3)**.

The patient's postoperative course was uncomplicated. He was discharged on postoperative day one with oral analgesia and topical chloramphenicol ointment. Socket healing was satisfactory, with no evidence of infection, implant exposure, migration, or wound dehiscence. The conformer was removed at four weeks post-evisceration, and a prosthetic eye was successfully fitted thereafter. At six months of follow-up, the socket remained healthy, with no complications reported. Visual acuity in the contralateral eye remained unchanged from baseline. The patient was subsequently discharged from ophthalmic care at this facility.

No formal genetic testing or systemic evaluation was performed due to resource limitations. Clinically, he had no dysmorphic features and was systemically and biochemically healthy. The post-operative CT brain showed no midline defects or other anomalies. This represents a limitation of the study regarding potential syndromic associations.

2.1. Histopathological Findings

Microscopy demonstrated disorganized intraocular tissue, including lens capsule remnants, pigmented epithelium, and neural tissue consistent with a disrupted retina. A well-demarcated lamellar bone fragment was observed **(Figure 4)**, lined by osteoblasts and

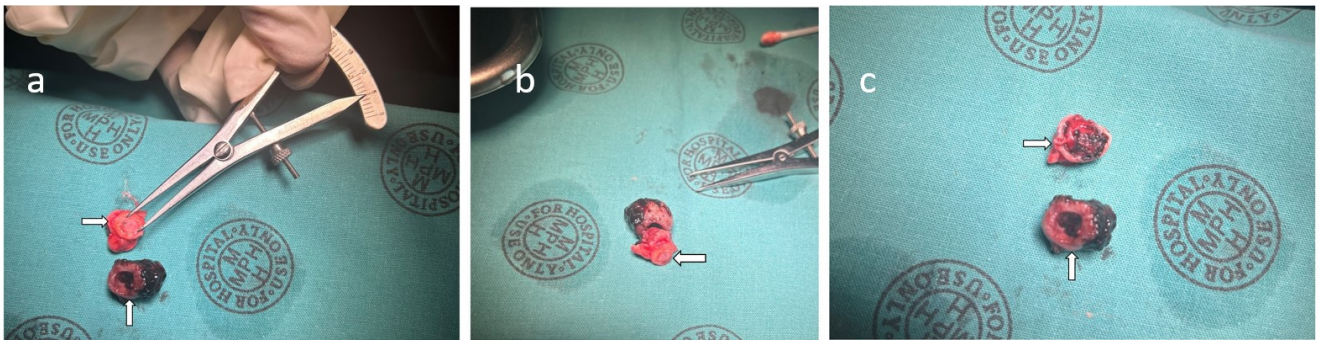

Figure 2: Gross specimen of lamellar intraocular bone with attached optic nerve. (a) Posterior aspect of the optic nerve (horizontal arrow) with the adjacent 12 mm bony structure (vertical arrow) (calliper at 5 mm). (b) Posterior view showing the anatomical position of the ossified mass as it was situated within the globe. Horizontal arrow indicating the posterior section of the optic nerve directed away from the globe. (c) Anterior view of the optic nerve where it invaginates into the base of the lamellar bony structure (horizontal arrow). Posterior section of bony element (vertical arrow).
*Images taken intra-operatively, immediately after excision.

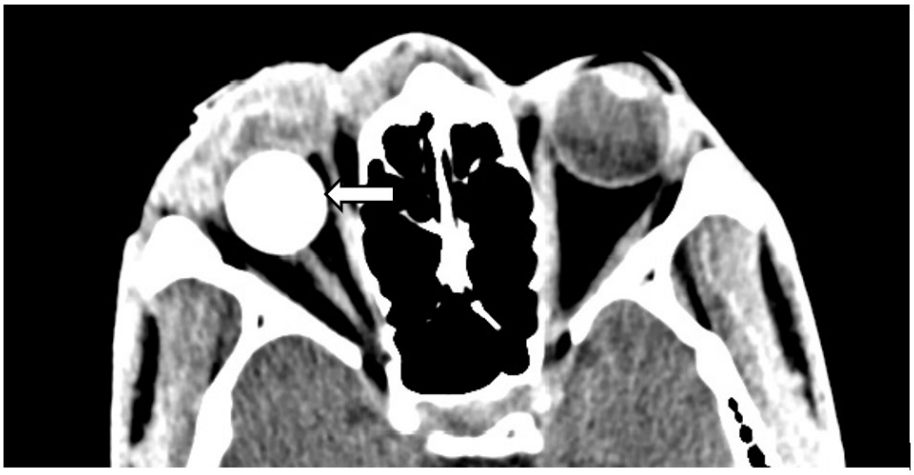

Figure 3: Postoperative CT scan revealed no other intra-orbital bony elements or malignancies. A 20 mm orbital implant was noted in the right socket (horizontal arrow).

containing mature adipocytes in marrow spaces **(Figure 6)**. Psammomatous and cementum-like calcifications were noted **(Figure 5)**. These calcifications, occurring within mature lamellar bone with marrow, reflect chronic degenerative ossification and help exclude neoplastic or congenital lesions, given their absence of atypia and organized cellular proliferation.

S100 immunostaining confirmed neural tissue. No malignant cells were identified. No fungal organisms or acid-fast bacilli were identified. No malignant features were seen.

The specimen required decalcification prior to histologic sectioning and was processed according to the College of American Pathologists' standard protocols. Routine laboratory practice employs an acid-based decalcification solution of distilled water, hydrochloric acid, and nitric acid in approximately a 17:1:1 ratio. As a limitation of this study, EDTA-based decalcification, which better preserves tissue architecture and antigenicity, was not used.

Additional immunohistochemical stains were not performed. Mature bone formation was readily identifiable on routine haematoxylin and eosin staining and did not require further immunohistochemical confirmation. Furthermore, in established osseous metaplasia, the tissue typically assumes the immunophenotype of mature bone, limiting the utility of epithelial or glial markers in reliably determining cellular origin. Osteocalcin staining was unavailable at our institution, and retrospective immunohistochemical analysis is not feasible due to constraints on tissue retrieval.

Given the presence of both pigmented epithelial and neural elements, and in keeping with existing literature, the osseous metaplasia likely arose via transdifferentiation of retinal pigment epithelium under chronic inflammatory or degenerative conditions.

3. Discussion

The diagnosis of MGDA in the contralateral eye was based on characteristic funduscopic features, including an enlarged, funnel-shaped optic disc with a central glial tuft, surrounding peripapillary excavation, radial configuration of retinal vessels, and peripapillary pigmentation, which together distinguish MGDA from optic disc coloboma, optic disc pit, and peripapillary staphyloma. These findings are consistent with established clinical diagnostic criteria for MGDA. Ancillary imaging of the left eye was limited; optical coherence tomography, fluorescein angiography, and OCT-angiography were not available at our facility at the time of presentation, and a B-scan ultrasonography was inadvertently not

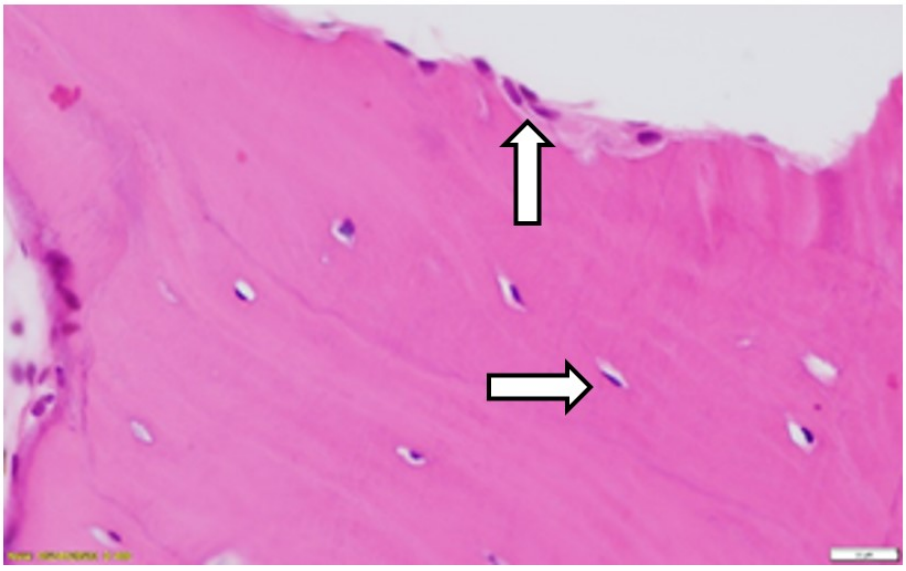

Figure 4: Prominent rimming of bone trabeculae by osteoblasts (vertical arrow) is evident, indicating active bone formation. Horizontal arrow indicating osteocytes. (Haematoxylin and eosin stain; original magnification ×40).

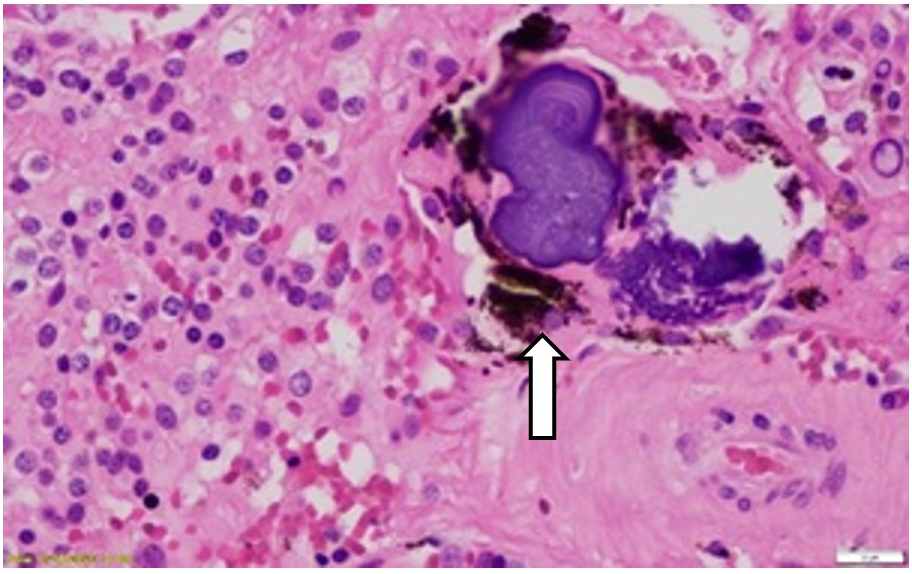

Figure 5: Psammomatoid calcifications are present (vertical arrow), characterised by basophilic, concentrically laminated structures. (Haematoxylin and eosin stain; original magnification ×40).

performed, which would have aided the diagnosis. While additional imaging would have aided further anatomical characterisation of the disc anomaly and the extent of the associated shallow retinal detachment, the diagnosis was made clinically. This is a limitation, but it was unlikely to change the clinical course and/or management of this patient.

Intraocular ossification is typically associated with chronic retinal detachment, inflammation, trauma, or phthisis bulbi [1, 4]. The present case is distinctive for several reasons: the ossified structure was focally centred on the optic nerve head, forming a bony "cap" with a central channel; the globe had no overt signs of phthisis at the presentation. Interestingly, the bony mass appears to be centred on the lamina cribrosa/optic nerve head. One plausible hypothesis is that chronic retinal detachment and subsequent folding of retinal tissue around the optic nerve head led to localized epithelial–mesenchymal transition, with the wrinkled retina serving as a scaffold for lamellar bone formation and the development of the observed bony “cap.”

Also, considering the history, the longstanding bilateral visual impairment, and the confirmed MGDA in the left eye, it is a plausible hypothesis that the patient had bilateral MGDA, with a more severe and unrecognized retinal detachment in the right eye, leading to chronic inflammation and ultimately osseous metaplasia. MGDA is frequently associated with retinal detachment, due to structural abnormalities at the optic nerve head and surrounding retina that predispose to tractional or exudative separation [8].

While chronic retinal detachment associated with MGDA provides a plausible mechanism for the observed ossification, alternative

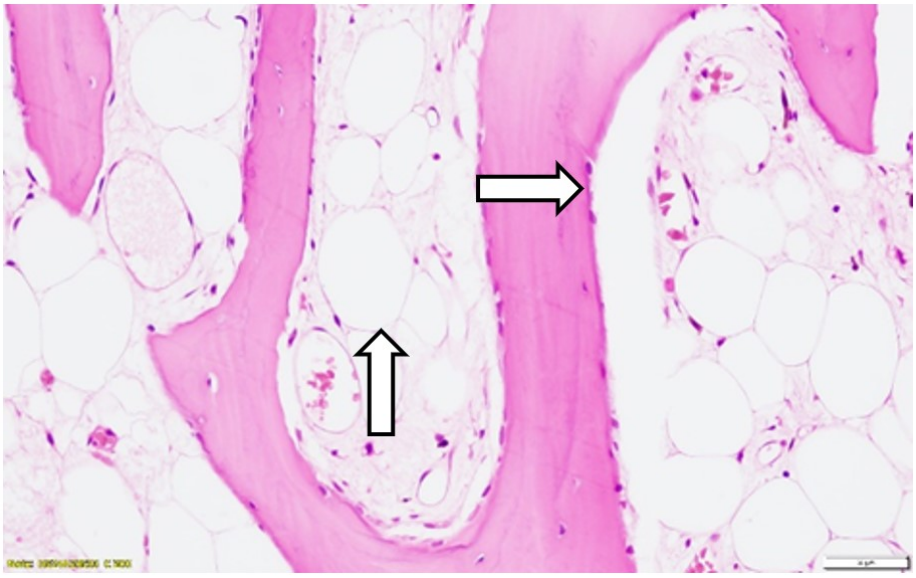

Figure 6: Prominent rimming by osteoblasts is noted around the bone trabeculae (horizontal arrow). In addition, loose connective tissue with mature adipocytes is noted in the marrow expanses (vertical arrow). (H/E micrograph, 20× magnification).

contributors, such as prior trauma, chronic intraocular inflammation, or unrelated congenital anomalies, cannot be excluded. Given the lack of pre-injury imaging, prior records, and comprehensive systemic evaluation, these remain speculative. In contrast, the presence of mature lamellar bone with marrow provides direct histopathologic evidence of a long-standing metaplastic process.

Cases of intraocular ossification are exceptionally rare, with the majority of reports describing partial or localized ossification, most often confined to the choroid or presenting as small osseous deposits rather than widespread bone formation [5, 9, 6, 10, 1]. The two most extensive ossification cases were reported by Alam et al (2020) [5] and Munteanu et al (2013) [10]. Munteanu et al described a flat, sickle-shaped cup of ossified choroid at the poster pole [10]. The most similar to this report was a case by Alam et al., in which an oval bony mass measuring 1cm was also eviscerated, but it did not encase the optic nerve head [5]. The interval between an inciting ocular pathology and enucleation ranged from 6 months to 22 years, with a mean of approximately 10 years [4]. Mature lamellar bone formation, as opposed to immature woven bone, reflects an advanced stage of bone maturation requiring prolonged remodeling and organization, consistent with a chronic process. Lamellar bone replaces woven bone over a prolonged remodeling phase and is indicative of long-standing osteogenic stimulation. Although the patient's reported duration of NLP vision is shorter than that reported by many, inflammation and degeneration likely preceded it by several years. The presence of fully mature lamellar bone with marrow elements supports a prolonged, chronic metaplastic process likely driven by sustained retinal pathology.

The presence of mature adipocytes within marrow spaces is a particularly significant finding, as it indicates complete heterotopic bone organ formation rather than simple calcification or partial osseous metaplasia. True marrow formation implies a prolonged, biologically active process requiring vascularisation and stromal maturation, suggesting that the ossification developed over many years and pre-dated the acute traumatic event. Reports of fully developed lamellar bone with marrow components are exceedingly rare and typically associated with long-standing retinal detachment and chronic inflammation.

Traditionally, the RPE has been regarded as the primary progenitor for intraocular bone formation through epithelial–mesenchymal transdifferentiation [11]. Bone morphogenetic proteins 7 and growth differentiation factors 5 have been implicated in this transformation [11]. Through fibroblast metaplasia and osteogenic transformation, glial tissue has also been reported as an alternative, but far less common, cellular progenitor source [4].

4. Conclusion

This case describes an exceptionally rare example of mature lamellar intraocular bone with marrow formation encasing the optic nerve head. The size, degree of maturation, and distinctive morphology make this presentation unique among reported cases of intraocular ossification. For clinicians and surgeons, this case highlights that profound heterotopic bone formation may arise silently in the setting of chronic retinal pathology. Ultimately, this case serves to illustrate the remarkable metaplastic capacity of ocular tissues, most plausibly through epithelial–mesenchymal transdifferentiation, and expands current understanding of the end of intraocular tissue remodelling.

Conflicts of Interest

The author declares no conflicts of interest.

Funding Source

This case report was conducted independently as a single observational study. No funding was received, and there are no financial disclosures to declare.

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Informed consent

Written consent for publication of this case report was obtained. The report does not contain any personal identifying information, and all data are presented anonymously in accordance with ethical publishing guidelines.

Large Language Model

None.

Authors Contribution

None. CG conceived the report, collected and interpreted the clinical, imaging, and histopathological information, drafted the manuscript, and performed critical revision. The author approved the final version for publication.

Data Availability

The data supporting the findings of this case report are contained within the article. No new datasets were generated or analyzed. Additional clinical details cannot be shared publicly to protect patient privacy; limited clarifications may be provided by the corresponding author upon reasonable request, subject to institutional and ethical approval.

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