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Case Report

Section: General Surgery

A Rare Neoplasm With A Deceptively Benign Face: A Case Report

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ARTICLE INFO

Article History:

Received: 12-05-2025

Accepted: 21-06-2025

Key words:

Retiform Hemangioendothelioma (RHE)
Parotid Gland
Vascular Tumor
Histopathology
Rare Neoplasm
Surgical Margins
Case Report.

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ABSTRACT

Aim : To report a rare case of retiform hemangioendothelioma arising in the parotid region and to emphasize the importance of histopathological evaluation in diagnosing atypical vascular neoplasms. **Introduction:** Retiform hemangioendothelioma (RHE) is a rare, low-grade malignant vascular tumor that resembles the rete testis. It is primarily affects young to middle-aged adults with a mean age of onset around 36 years, with a higher incidence in females. RHE presents as a slow-growing, painless mass, often in the distal extremities. Despite its low metastatic potential, RHE is locally aggressive and has a high recurrence rate. Accurate diagnosis relies on histopathological examination and immunohistochemical staining for endothelial markers. The most common locations include the skin and subcutaneous tissue of the lower extremities. **Case Description:** A 31-year-old male from West Bengal, a shopkeeper, had a gradually progressive, non-tender swelling in front of his right ear for the past 8 years. The swelling, initially small and asymptomatic, grew to 8 x 4 cm. Despite two previous surgical interventions in 2012 and 2015, the swelling recurred and enlarged, indicating a persistent underlying pathology. The patient's swelling was painless and non-tender, without any associated symptoms. The recurrence suggested a persistent pathology, possibly due to incomplete excision or the lesion's nature. **Clinical Significance:** The present case underscores the need for histopathological confirmation in atypical parotid lesions to avoid misdiagnosis and ensure appropriate surgical management of rare vascular tumors like retiform hemangioen dothelioma. **Conclusion:** This case highlights the diagnostic challenge posed by rare vascular neoplasms such as retiform hemangioendothelioma, especially when presenting at atypical sites such as the parotid region. Accurate diagnosis through histopathological evaluation is crucial for guiding appropriate surgical management and ensuring long-term disease control.

INTRODUCTION

Retiform hemangioendothelioma (RHE) is an exceptionally rare, low-grade malignant vascular tumor characterized by its unique histological pattern that resembles the rete testis [1]. First described by Calonje et al. in 1994, RHE lies intermediate on the biological spectrum between benign hemangiomas and highly malignant angiosarcomas [2]. It predominantly affects young to middle-aged adults, with a higher incidence reported in females [1]. Clinically, RHE typically presents as a slow-growing, painless mass, most commonly in the distal extremities [3]. However, cases involving unusual locations such as the trunk, scalp, and head and neck region have also been documented [1]. Despite its low metastatic potential, RHE is locally aggressive and demonstrates a high recurrence rate,reported in up to 60% of cases [4]. Due to its rarity and morphological overlap with other vascular neoplasms, accurate diagnosis relies heavily on histopathological examination supplemented by immunohistochemical staining for endothelial markers such as CD31, CD34, and ERG [1, 5].

Histologically, RH is characterized by arborizing vascular channels resembling the normal rete testis, lined by a single layer of endothelial cells with a prominent hobnail appearance. The tumor typically exhibits low mitotic activity and minimal pleomorphism [6]. A prominent lymphocytic infiltrate is often present in the stroma. Differentiation from other vascular tumors, such as Kaposiform Hemangioendothelioma and Dabska's tumor, is crucial due to overlapping features [7].

Immunohistochemical staining plays a vital role in confirming the diagnosis of RH. The tumor cells typically express endothelial markers such as CD31, CD34, and factor VIII-related antigen [8]. These markers aid in distinguishing RH from other vascular neoplasms. RH predominantly affects young adults, with a mean age of onset around 36 years. Both males and females are equally affected, and the tumor often presents in the second to fourth decades of life [9].

The most common locations include the skin and subcutaneous tissue of the lower extremities, though cases have been reported in the head and neck region, trunk, penis, and pleura [10]. Clinically, RH typically manifests as a slow-growing, painless, well-defined nodule or plaque. The lesions are often red or bluish in color and may be asymptomatic. While most cases are localized, some reports indicate regional lymph node metastasis and local spread to soft tissue.

CASE DESCRIPTION

A 31-year-old male, a shopkeeper by occupation from West Bengal, presented with a chief complaint of a gradually progressive swelling located in front of the right ear, persisting for the past 8 years. The swelling was painless and non-tender, with no associated symptoms such as facial weakness, fever, or discharge. The onset was insidious, with the swelling initially being small and asymptomatic. Over time, the lesion had a gradual and progressive increase in size, currently measuring approximately 8 × 4 cm. The patient also reported a history of two prior surgical interventions for the same complaint, performed in 2012 and 2015. Despite these procedures, the swelling had recurred and continued to enlarge, suggesting a recurrent or persistent underlying pathology.

Preoperative Condition

The patient currently had a 8 x 4 cm swelling, with no complaints of pain. There was no history of fever, chronic cough, or unintentional weight loss. The patient was not on any regular medications and had no known comorbidities. Previous FNAC reports were not available for review.

General Examination

The patient is a middle-aged male, moderately built and nourished, conscious, cooperative, and well-oriented to time, place, and person.

Vital Signs

The patient's vital signs were within normal limits. He was afebrile, blood pressure was recorded at 110/70 mmHg, which falls within the normal range. The pulse rate was 80 beats per minute, suggesting a normal resting heart rate. Oxygen saturation (SpO2) was 98% on room air, indicating adequate oxygenation without the need for supplemental oxygen.

Local Examination (Physical Appearance)

A solitary swelling measuring approximately 8 × 4 cm was noted over the right pre-auricular region, extending from the zygomatic arch superiorly to the angle of the mandible inferiorly. The swelling had well-defined borders and an irregular contour.

Palpation

On examination, the area was non-tender, firm in consistency and had a nodular surface. There was no local rise in temperature, suggesting absence of active inflam mation or infection.

Additional Local Findings

Two well-healed surgical scars were noted over the swelling. One scar measured approximately 4 × 0.5 cm, while the other measured around 2 × 0.5 cm, both scars were healed by primary intention, with no signs of discharge, inflammation, or induration. Additionally, the right ear lobe was elevated, likely due to the underlying mass effect of the swelling.

Figure 1: Illustration showing the patient's front and side views used during physical examination for diagnostic assessment. These views help in evaluating the location, size, and surface characteristics of the swelling or lesion, as well as in identifying any visible asymmetry, skin changes, or surgical scars relevant to the clinical diagnosis.

Clinical Diagnosis

A recurrent benign tumor involving the right parotid gland, likely to be a pleomorphic adenoma.

Hematological Profile

Investigations revealed a hemoglobin level of 14.8 mg/dL and a packed cell volume (PCV) of 36.4%, both within normal limits. The total white blood cell count (TWBC) was 6,200/µL, and the platelet count was 189,000, indicating a normal hematological profile. Differential count showed neutrophils at 51.8%, lymphocytes at 36.6%, monocytes at 4.6%, eosinophils at 6.5%, and basophils at 0.5%. Serology results were negative or non-reactive. The patient's random blood glucose (GRBS) was 116 mg/dL. Blood urea nitrogen (BUN) was 10.5 mg/dL, and urea was 22.47 mg/dL.Serum creatinine was 0.59 mg/dL, suggesting normal renal function. Electrolyte analysis showed sodium at 139.5 mEq/L, potassium at 4.79 mEq/L, calcium at 9.53 mg/dL, and chloride at 104.3 mEq/L, all within normal ranges.

Radiological Investigation

Ultrasound Report

The lesion appeared hypoechoic on ultrasound imaging (Fig. 2), meaning it reflects fewer sound waves and therefore appears darker than the surrounding tissues. Additionally, it demonstrated posterior acoustic enhancement, a pheno menon where increased brightness is seen behind the lesion due to the ultrasound waves passing through a structure that transmits sound more effectively than the surrounding tissue suggesting the lesion is likely fluid-filled or has low internal density.

Ultrasound report

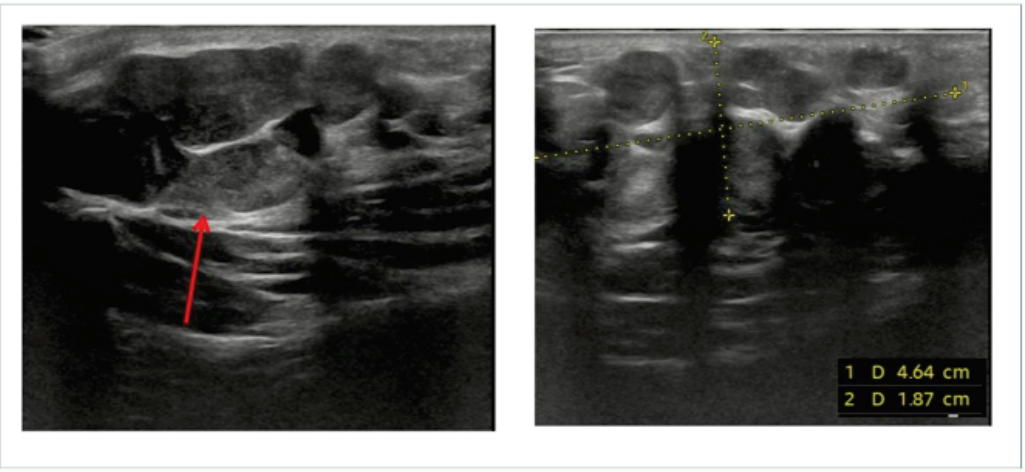


Figure 2: Illustration of ultrasound report, showing the lesion appeared hypoechoic

CT Scan Imaging

On CT imaging (Fig. 3) the lesion exhibited homogeneous attenuation, meaning its internal density appears uniform throughout, without areas of calcification, necrosis, or cystic change. Additionally, it showed prominent enhan - cement after contrast administration, indicating that the lesion takes up contrast material well often a sign of increased vascularity or active tissue within the lesion (Fig. 4).

CT Scan

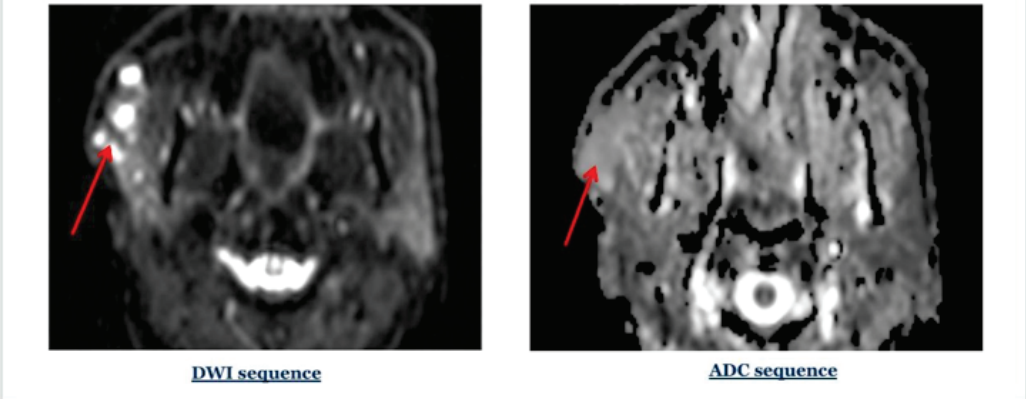


Figure 3: Illustration of CT imaging showing the lesion with homogeneous attenuation. This radiological appearance is suggestive of a well-defined, likely benign pathology, with uniform density and no evidence of calcification, necrosis, or irregular margins.

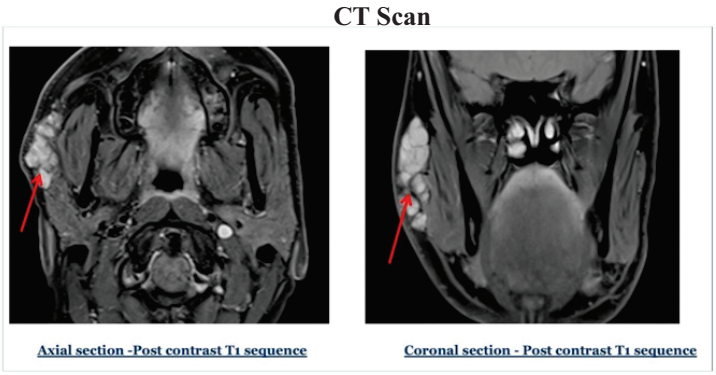


Figure 4: Illustration of CT imaging showing the lesion with homogeneous attenuation. This radiological appearance is suggestive of a well-defined, likely benign pathology, with uniform density and no evidence of calcification, necrosis, or irregular margins.

MRI imaging

On MRI, the lesion typically appeared hypointense (dark) on T1-weighted images, suggesting it has low fat or protein content. On T2-weighted images, it appeared hyperintense (bright), which is characteristic of fluid-rich or cystic com-ponents. Surrounding the lesion, was a rim of decreased signal intensity on T2, which likely represents a fibrous capsule. Following contrast administration, T1-weighted images showed homogeneous enhancement, indicating uniform contrast uptake throughout the lesion, which suggested a well-vascularized and solid nature.

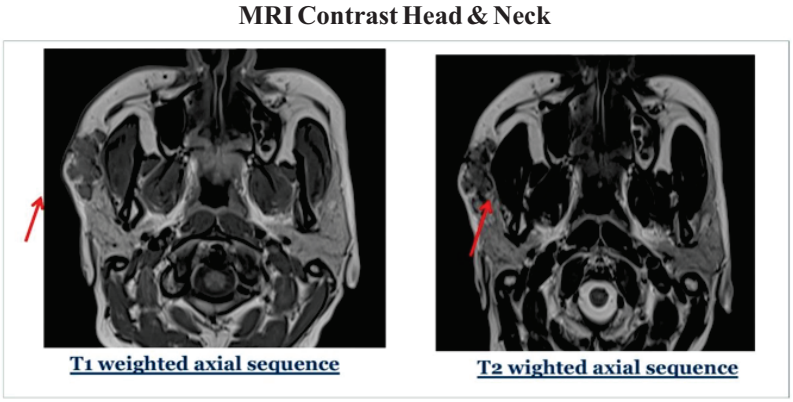
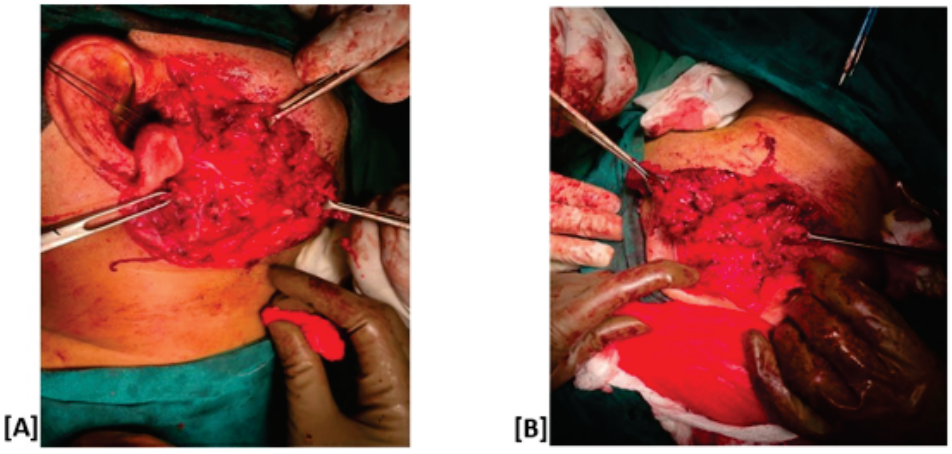


Figure 5: Illustration of MRI image, the lesion appears hypointense (dark) on T1-weighted images, suggesting it has low fat or protein content and on T2-weighted images, it appeared hyperintense (bright).s.

Surgical Procedure

Based on the clinical, pathological, and radiological findings, superficial parotidectomy was performed, following a complete pre-anesthetic evaluation and after obtaining written informed consent. Under general anes thesia, the patient was placed in supine position. A lazy S-shaped incision was made over the right preauricular region to provide optimal exposure of the parotid gland. After visualisation and identification of the facial nerve trunk, careful dissection was carried out along the fasciovenous plane of Patey, and the gland was excised in toto and forwarded for biopsy. After closure, the patient was transferred to the recovery room for postoperative monitoring.



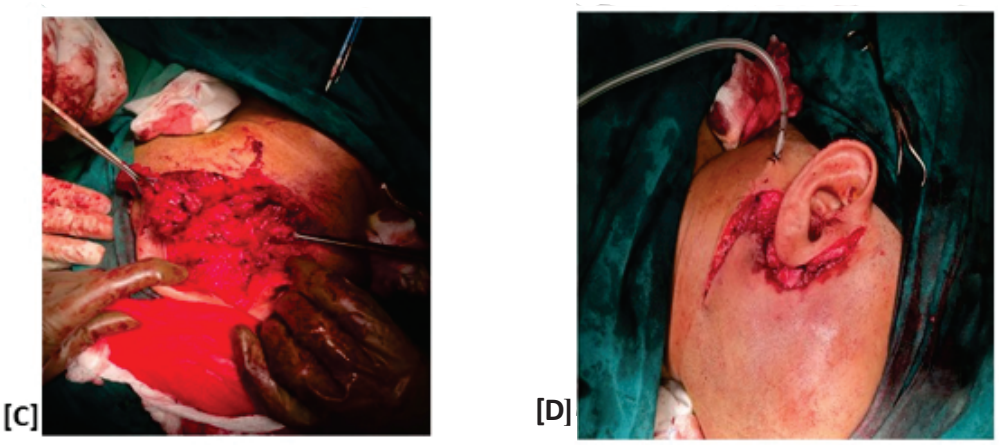


Figure 6 : Sequential illustration of the operative procedure. [A] A precise surgical plane was established above the parotid fascia, and meticulous dissection was undertaken to locate the main trunk of the facial nerve. [B] Upon successful exposure, the facial nerve was clearly visualized and carefully preserved throughout the surgery to prevent any functional impairment. [C] An adenomatous enlargement of the right superficial parotid gland, measuring approximately 5 × 3 cm, was identified. Dissection was extended to reveal all five branches of the facial nerve, which were intricately embedded within the glandular tissue and were meticulously preserved during excision. [D] The surgical field was irrigated thoroughly with saline, and a suction drain was placed and securely fixed to the skin. Layered closure of the incision was performed, followed by the application of a sterile dressing.

Postoperative Procedure

The postoperative period was uneventful and without complications. The suction drain was removed on the third postoperative day, and the surgical wound appeared healthy and showed good signs of healing at the time of discharge. Due to the presence of neuropraxia, the patient was initiated on a course of oral Prednisolone during the postoperative phase to aid in nerve recovery and reduce inflammation. Regular follow-up was advised to monitor neurological function and wound healing.

DISCUSSION

RHE represents a rare and diagnostically challenging vascular neoplasm that occupies an intermediate space between benign hemangiomas and highly malignant angiosarcomas [1]. The tumor is characterized histologically by elongated, arborizing vascular channels arranged in a retiform pattern resembling the rete testis, lined by hobnail endothelial cells [12]. Originally described by Calonje et al. in 1994, RHE has since been recognized for its distinctive morphological features and locally aggressive behavior with a high recurrence rate, but limited metastatic potential [13].

In this case the patient, a 31-year-old male, presented with a longstanding swelling in front of the right ear, persisting for approximately 8 years. Such a chronic and slow-growing mass in the parotid region is most commonly suggestive of a benign salivary gland tumor, with pleomorphic adenoma being the most frequent [14]. Ultrasound imaging of the lesion showed it to be hypoechoic with posterior acoustic enhancement, features typically associated with cystic or low-density solid lesions. Hypoechogenicity indicates that the lesion reflects fewer ultrasound waves compared to surrounding tissues, appearing darker on the scan, while posterior enhancement suggests that the lesion transmits sound waves more efficiently, further supporting the likelihood of a benign pathology with a cystic component or low internal density [15].

The swelling was insidious in onset, initially small and asymptomatic, and had progressively increased in size over time, now measuring approximately 8 × 4 cm. It remained painless and non-tender throughout its course, with no associated symptoms such as facial weakness, fever, or discharge. Of particular significance is the patient's history of two prior surgical interventions for the same complaint, performed in 2012 and 2015. The recurrence and continued enlargement of the swelling despite these procedures suggest the presence of a persistent or recurrent pathology, possibly due to incomplete excision or the nature of the lesion itself [16]. Recurrent pleomorphic adenoma is a common consideration in such cases, given its tendency to recur if not completely excised with an adequate margin.

The chronicity, slow growth, and recurrence all point toward a benign but persistent salivary gland tumor, most likely arising from the superficial lobe of the parotid gland. These features warrant a thorough evaluation and a carefully planned surgical approach with particular attention to preserving the facial nerve and minimizing the risk of further recurrence. Although RHE most commonly affects the distal extremities, its occurrence in the head and neck region is exceedingly rare. The involvement of the parotid region, as in this case, is even more atypical and can easily be mistaken for salivary gland tumors or benign skin adnexal lesions [17]. This case heightened clinical suspicion and broad differential diagnosis when evaluating soft tissue masses in uncommon sites.

Given the imaging characteristics and the clinical history, the findings were consistent with a benign lesion of the superficial lobe of the parotid gland. Surgical excision remains the treatment of choice, not only for definitive diagnosis through histopathological evaluation but also to prevent further growth and potential malignant transfor mation. Careful identification and preservation of the facial nerve and its branches during surgery are crucial to minimize the risk of postoperative facial nerve dysfunction.

The tumor's locally infiltrative nature and predilection for recurrence reported in up to 60% of cases warrant complete surgical excision with negative margins as the mainstay of treatment [18]. In this case, surgical margins were free of tumor, offering an optimistic prognosis. While regional lymph node metastasis has only been reported in isolated cases, long-term surveillance remains essential due to the unpredictable clinical course [19].

Immunohistochemistry, although not detailed in this case, would play an important adjunctive role in confirming the vascular origin of the tumor, typically showing positivity for endothelial markers such as CD31, CD34, ERG, and VEGFR2 [20]. These markers not only aid in differentiating RHE from angiosarcoma and other mimickers but also reinforce the low-grade malignant potential of the tumor.

In totally, the present case underscores the diagnostic complexity and clinical significance of recognizing RHE, particularly when arising in an unusual location such as the parotid region. Early and accurate diagnosis, achieved through thorough histopathological analysis, is paramount to guiding appropriate surgical management and minimizing the risk of recurrence [21]. This case also enriches the existing literature by documenting a rare site of involvement, thereby expanding the clinical spectrum of this unique vascular tumor.

CLINICAL SIGNIFICANCE

This case of RHE holds considerable clinical importance due to the tumor's rarity, its atypical presentation in the parotid region, and the diagnostic challenge it poses. While RHE predominantly affects the distal extremities, this case illustrates that clinicians and pathologists must consider it in the differential diagnosis of slow-growing, painless swellings in uncommon anatomical locations such as the head and neck region.

Early clinical misinterpretation, as seen here with the initial cytological impression of a pleomorphic adenoma, can lead to underestimation of the lesion's aggressive potential. This underlines the critical value of histopathological confirmation, especially in lesions that do not resolve or change over time. Furthermore, the locally infiltrative nature of RHE necessitates complete surgical excision with tumor-free margins to reduce the risk of recurrence, which is reported in up to 60% of cases.

Recognizing RHE is not only essential for ensuring appropriate surgical intervention but also for avoiding over-treatment. It must be distinguished from angiosarcoma, a highly malignant vascular tumor, as overtreatment in the form of extensive resections or unnecessary adjuvant therapy may otherwise be considered.

Additionally, this case adds to the growing but limited body of literature on RHE in the parotid region, contributing to a broader understanding of its clinicopathological spectrum. Increased awareness among clinicians, radio - logists, and pathologists about such rare vascular tumors can significantly improve diagnostic accuracy and patient outcomes through timely, precise, and tailored management strategies.

CONCLUSION

The present case study highlights the importance of thorough histopathological evaluation and clinicopatho logical correlation in diagnosing rare vascular tumors like retiform heman -gioendothelioma in atypical sites such as the parotid region. Early detection and complete surgical excision with clear margins are key to preventing local recurrence. Ultrasound imaging revealed a hypoechoic lesion with posterior acoustic enhancement, indicating a structure that is likely fluid-filled or of low internal density. These findings are consistent with a benign lesion of the parotid gland, warranting further evaluation and appropriate management.

Conflicts of Interest

Authors declared that there is no conflict of interest.

Funding

None

Ethics Approval And Consent To Participate

All necessary consent & approval was obtained by authors.

Consent For Publication

All necessary consent for publication was obtained by authors.

Data Availability

All data generated and analyzed are included with in this research article.

Author Contributions

All authors contribute significantly in this manuscript.

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How to cite: Sriya Gollamudi, Rahul Varma Datla, Ramesh Reddy G., Vanaja Reddy Banda, Amrutha T., Sindhuja R., A Rare Neoplasm With A Deceptively Benign Face: A Case Report. *International Medicine*, 2025;11 (1) :1-7