Case Report
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Ancient schwannoma of the scalp: an infrequently diagnosed entity

Abstract

Aim: To report an highly unusual case of Ancient schwannoma (AS) in the temporal scalp region and to emphasize the importance of histopathological evaluation in diagnosing scalp lesions that mimic more common entities.

Materials and methods: An young adult male (24 years old) reported with a slow-growing non-tender scalp lesion of two years' duration in the left temporal region. Clinical examination and ultrasonography suggested a benign subcutaneous lesion, initially suspected to be a lipoma. The scalp mass was surgically excised in its entirety under local anaesthesia and sent for histopathological analysis. A systematic review of literature from PubMed and Google Scholar was also conducted to identify similar cases.

Results and conclusion: The diagnosis of ancient schwannoma was established histologically, based on encapsulated spindle cell morphology, presence of Antoni A and B areas, Verocay body formation and degenerative changes including hemorrhage and hemosiderin deposition. The patient had an uncomplicated postoperative course, and follow-up at one year revealed no signs of recurrence. Only three prior cases of AS of the scalp were found in the literature, underscoring its rarity. This case reinforces the necessity of considering AS in the differential diagnosis of atypical scalp lesions and highlights that histopathology remains the gold standard for diagnosis. Surgical excision remains the definitive and curative treatment, with excellent outcomes and minimal recurrence.

Keywords: as, scalp tumor, temporal region, schwannoma, verocay bodies, surgical excision

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Introduction

Schwannomas, also known as neurilemmomas, are benign tumors originating from neuroectodermal cells. They can develop along any nerve that contains Schwann cells, including those in the cranial, peripheral, and autonomic nervous systems. The optic nerve and the olfactory nerves are exceptions, as they are centrally derived and lack Schwann cell myelination.[1] It is estimated that 25–45% of schwannomas arise in the head and neck region.[2–4] They are usually well-demarcated, encapsulated lesions that grow slowly and remain clinically silent in many cases. Schwannomas exhibit multiple histopathological variants, such as common, plexiform, glandular, epithelioid, cellular, melanotic, and ancient schwannomas (AS).[5]

AS, a rare and distinctive histopathological variant, account for only 0.8% of all soft tissue tumors.[1] AS are rare, benign, encapsulated tumors, solitary lesion (90%) characterized by slow growth. The term may also be applied to schwannomas exhibiting marked cystic degeneration, characterized by a relative loss of Antoni A areas, along with features such as perivascular hyalinization, calcification, and cystic necrosis. Cystic growths on the scalp can be easily mistaken for common tumors like lipoma, dermoid cysts and trichilemmal cysts.[6]

We present an extremely rare case of an asymptomatic mass in the temporal region, which was identified as an AS following an excision biopsy. Upon reviewing the literature on 2 different databases (PubMed and Google Scholar), only three cases of AS of the scalp have been reported so far.

Case report

A young adult male, aged 24, reported a gradually enlarging, painless scalp swelling over the left side for the past two years. There was no history of local trauma. No relevant past medical or family history of comparable lesions was elicited. Physical examination revealed a freely mobile, soft, well circumscribed and non-tender subcutaneous mass on the left temporal region of scalp. Approximate size of the lesion is 2 cm × 1 cm size. Both subjective complaints and objective neurological findings were unremarkable. Initial differential diagnosis included lipoma.

Investigations

Ultrasonograhy of left temporal region was done which showed a well defined heterogenous lesion of size 1.7cm x 0.7 cm noted involving skin and subcutaneous plane of left temporal region.

Treatment

A hairline elliptical incision was strategically placed, enabling smooth dissection of the lesion from the perichondrium beneath and the overlying skin flaps. The mass was extracted from the galeal plane (Figure 1). Lesion was completely excised under local anesthesia and sent for histopathology. The dead space created by the exposure of galeal plane was decreased by linear closure of the skin flaps in the midline (Figure 2). By the seventh postoperative day, the sutures were removed, revealing a clean, well-healed wound without signs of infection.

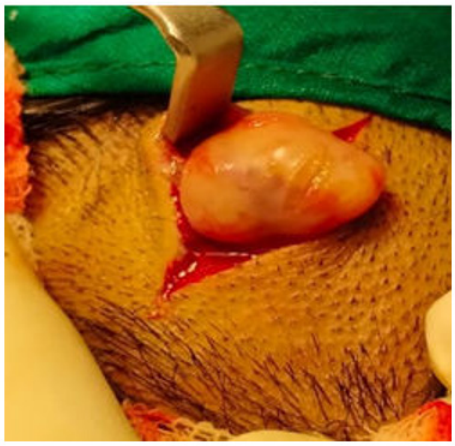

Figure 1 Extraction of the mass from galeal plane in the left temporal region.

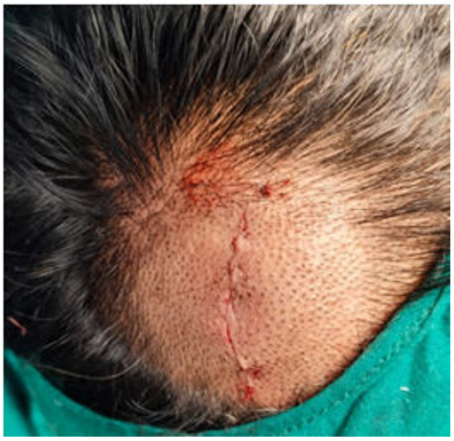

Figure 2 Linear closure of the skin flaps in the midline.

Histopathology report

Microscopic analysis demonstrated a well-defined, encapsulated tumor with characteristic Antoni A (hypercellular) and Antoni B (hypocellular) regions. Cells have narrow elongated nuclei with tapered ends (Figure 3,4). Cellular areas show palisading arrangement of nuclei consistent with verocay bodies interspersed with collagen fibres showing areas of hyalinization and myxoid degeneration. Foci of hemorrhage and hemosiderin deposition is noted (Figure 5 & 6).

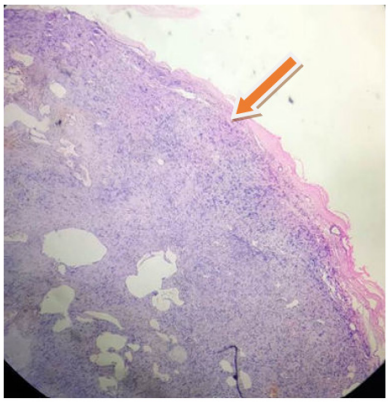

Figure 3 10X Power view of a well encapsulated tumor. Red arrow pointing the capsule.

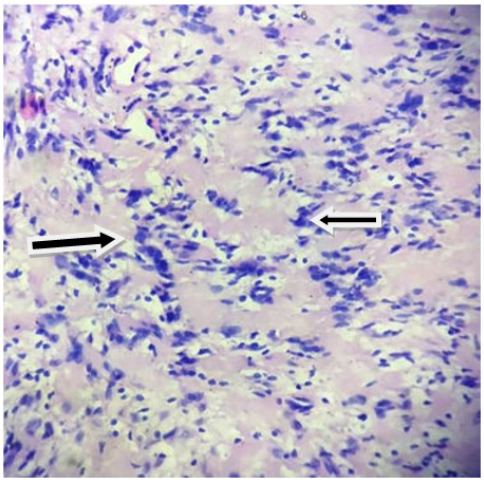

Figure 4 Antoni A areas showing Nuclear palisading with interspersed collagen fibrils(Verocaybodies in black arrows).

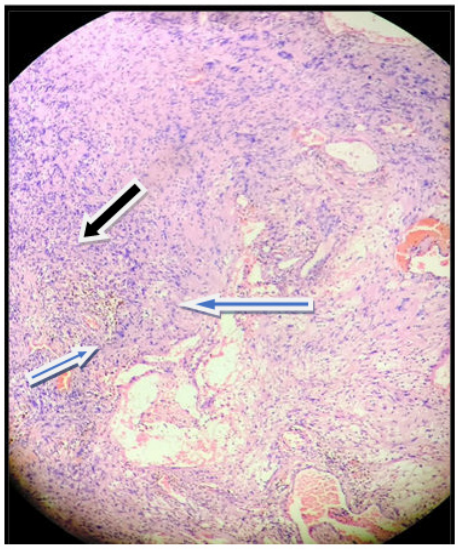

Figure 5 Low power image showing congested blood vessels, hemorrhage(blue arrows) and hemosiderin deposition(black arrows).

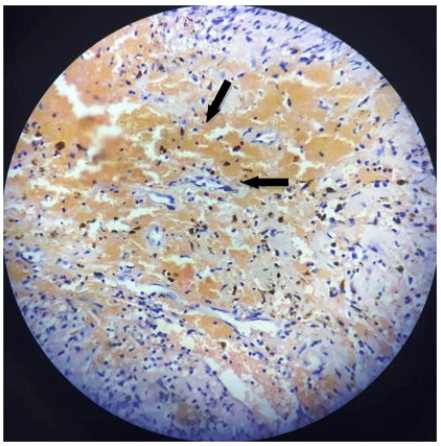

Figure 6 Areas of hemorrhage and hemosiderin deposits (black arrows).

Following the procedure, the patient showed no recurrence during follow-up visits for upto one year.

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Discussion

Neurogenic tumors are derived from neural crest cells, which give rise to Schwann cells and sympathicoblasts. Tumors such as schwannomas (also termed neurilemmomas) and neurofibromas originate from Schwann cells.[7,8] Schwannomas can develop along any nerve that possesses a Schwann cell sheath, with the exception of the optic and olfactory nerves, which do not contain such sheaths.[7] Verocay first described schwannomas as a pathological entity in 1908 and subsequently named them neurinomas in 1910.[9] The nomenclature of these tumors evolved historically, with Masson introducing the term schwannoma in 1932, followed by Stout, who proposed the term neurilemmoma in 1935.[10,11]

Schwannomas represent about 5% of all benign soft-tissue neoplasms, with 6%–8% occurring intracranially and 25%–45% extracranially in the head and neck.[12–14] The reported incidence of schwannomas is 1.2 per 100,000 population per year.[15] Approximately 90% of cases are solitary lesions, 3% are associated with neurofibromatosis type 2, and 2% are multiple, associated with schwannomatosis.[16] Schwannomas are equally seen in both genders in their third and fourth decades of life.[17]

Schwannomas are slow-growing benign neoplasms that typically present without symptoms, are non-tender on examination, and demonstrate a soft to firm consistency with mobility. Progressive growth can lead to mechanical compression of the originating nerve or adjacent anatomical structures, manifesting as pain, altered sensation, or tenderness. Although they are generally not known to ulcerate or erode, there has been a report of an external auditory canal schwannoma causing mastoid bone erosion.[1,2] The exact etiology of schwannomas is unknown, but they may arise from mesodermal or ectodermal tissues, involving the perineurium and Schwann cells, respectively.[1,18] Prior trauma may also be a contributing factor. Schwannomas typically grow over several years before symptoms become apparent.[1]

In 1985, Erlandson proposed a classification system for schwannomas comprising seven distinct subtypes: classical (Verocay), plexiform, cellular, melanotic, ancient (degenerated), cranial-nerve related and granular cell schwannomas.[19] Earlier, in 1951, Ackerman and Taylor introduced the term "AS" to describe schwannomas of long duration.[20]

Microscopically, Schwannomas display a uniform population of spindle-shaped cells with pale eosinophilic cytoplasm and tapered or oval nuclei, consistent with their neural crest origin.The key diagnostic feature is the presence of Antoni A and B areas, with Antoni A regions frequently exhibiting Verocay bodies—structures formed by palisading nuclei bordering acellular, eosinophilic zones. Chronicity of the lesion often results in degenerative changes like atypia, hemorrhage, cystic degeneration, and calcification, suggestive of the ancient schwannoma variant. As the tumor grows, vascular insufficiency may occur, and prolonged evolution of schwannomas generally leads to some degree of degenerative changes.[1,2,21,22]

In cases with degenerative changes, a thorough differential diagnosis is essential to distinguish schwannomas from malignant conditions such as leiomyosarcoma and malignant fibrous histiocytoma and others.[2,19] Analyzing the entire surgical specimen and performing immunohistochemical analysis are crucial to avoid misdiagnosis.[2] Although schwannomas with degenerative changes may present cellular atypia that can mimic malignancy, they are generally non-aggressive. However, malignant transformation to neurofibrosarcoma has been reported in less than 1% of cases.[1]

Immunohistochemistry of schwannomas typically shows positivity for S-100 protein and collagen type IV, with the capsule possibly positive for epithelial membrane antigen. Other useful immunohistochemical stains include myelin basic protein, CD68, and collagens I, III, IV, and vimentin, schwannomas lack expression of CD117, SMA, and desmin, and are generally negative for CD34.[23]

Head and neck occurrences of AS are infrequent, and scalp lesions represent a particularly rare presentation. AS of the head and neck was first reported by Eversole and Howell in 1971.[24]

"A review of the literature revealed fewer than 70 cases of AS in the head and neck region, with only three cases of 'AS' specifically involving the scalp"(Table 1). This information underscores the rarity of this tumor in the scalp. Based on the available data, our case represents the fourth reported instance of an AS in the scalp and the second specifically in the temporal region of the scalp.

Table 1 Summarizes of published case reports with ancient Schwannoma in the SCALP

S.NO.	Case Authors	Year	Age / Gender	Country	Site	Duration	Clinical symptoms	Lesion size	Clinical diagnosis	Surgical procedure	F/U	Recuurence	MT
1	Binu M. G[23]	2015	32/m	Tamilnadu, INDIA	Left parietal region	8 years	Painless swelling over scalp	6x 5 cms	Sebaceous cyst / dermoid cyst	Complete excision	unknown	-	-
2	Sang-Houn Lee[6]	2022	36/m	Korea	Left Parietal region	5 years	Painless swelling over scalp	3x2cm	Trichilemmal cyst / dermoid cyst	Complete excision	unknown	-	-
3	G. C. Ravi[1]	2022	75/F	Karnataka, India	Right mastoid region	6 month	uncontrollable profuse bleeding from a gradually progressing, painless swelling	3x1.5cm	Hemangioma	Complete excision, Feeding vessel- not identified. Cauterization of base done. Carotid and vertebral doppler- inconclusive	unknown	-	-

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Table 1Continued....

4	Present case	2024	24/M	Tamilnadu, india	Left temporal region	2 years	Asymptomatic slow growing mass	2x 0.8cm	Lipoma	Complete excision	2 months	No recurrence	-

Among the 4 cases reported in scalp, male preponderance is seen and 3 cases were reported from India.[1,6,23] The lesions appeared as well-defined submucosal swellings ranging from 2–6 cm, including in our case. All lesions, including the present case, were well-circumscribed and sharply demarcated from adjacent tissues. All were asymptomatic painless swelling except for the lesion present in mastoid which was haemorrhagic. Clinically, all cases of AS in scalp were misdiagnosed to be sebaceous cyst, trichilemmal cyst, lipoma, hemangioma and others.

Although ultrasonography, CT, and MRI cannot confirm a definitive diagnosis of schwannoma, they are instrumental in evaluating the lesion's localization, extent, and its proximity to adjacent structures.[2,3] This information is essential for planning surgery and determining further treatment. Histopathological analysis remains the gold standard for diagnosing schwannoma, with imaging and ancillary studies aiding in assessment and preoperative planning.

Complete excision of the lesion, preserving the nerve of origin is the elected treatment.[2,17,19] In the present case, the nerve of origin is likely the temporal branch of the auriculotemporal nerve, which arises from the mandibular division of the trigeminal nerve (V3). Since the tumor is well-encapsulated and non-adherent, it generally has an excellent prognosis when treated with complete surgical excision, with a low risk of recurrence.[2,17,19]

Conclusion

AS is a slow growing, benign, solitary neoplasm. This case reports a rare presentation of ASin temporal region of scalp. The clinical diagnosis of the lesion is challenging and histopathological findings are crucial for a correct diagnosis.AS, though rare, should remain a diagnostic consideration in patients presenting with scalp tumors. Identification of the distinct histopathological features of ancient schwannoma is vital to distinguish it from malignant tumors and avoid unnecessary aggressive therapy. A benign clinical course is generally observed, and complete excision is regarded as the preferred treatment modality. Recurrence and malignant transformation are rare events in the clinical course of schwannomas.

What is Already Known?

1. As are rare benign neoplasms exhibiting histologic degeneration, most frequently occurring in the head and neck region, with scalp localization being exceptionally uncommon.
2. They often mimic more common scalp lesions (e.g., lipoma, dermoid or trichilemmal cysts), making clinical diagnosis challenging without histopathological confirmation.
3. Diagnosis is based on histopathology, and treatment by complete excision yields a favorable prognosis with minimal chance of recurrence.

What additional Values in the present study?

1. This is only the fourth reported case of AS on the scalp in the literature and the second one specifically in the temporal region, highlighting its extreme rarity.
2. This case underlines the need to include ancient schwannoma in the differential diagnosis of scalp masses, especially when preoperative imaging is inconclusive.
3. A thorough literature review was conducted, comparing clinical presentations, diagnostic approaches, and outcomes, thereby contributing to the limited global data and supporting clinical decision-making in similar cases.

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None.

Conflicts of interest

The authors declare that there are no conflicts of interest.

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