Haemangioma or vascular malformation of the tympanic membrane? a case report and review of literature

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Abstract Benign vascular lesions are rarely found on the tympanic membrane. We report a case of such a lesion in a novel location, with review of the relevant literature. We also highlight the significance of accurate classification of such lesions, proposing adoption of more pathophysiologically-correct nomenclature of “haemangioma” and “vascular malformation”.

Keywords Haemangioma · Capillary haemangioma · Cavernous haemangioma · Vascular malformation · Tympanic membrane · Terminology

Introduction

Benign vascular lesions frequently occur in the head and neck. It is, however, rarely found on the tympanic membrane. To date, only 7 cases have been reported in English medical literature [1–7] (Medline search, 1968 to present), with 3 of the lesions termed “capillary haemangioma” [1, 3, 6] and the remaining 4 “cavernous haemangioma” [2, 4, 5, 7]. We present a case of a vascular lesion arising from the tympanic membrane with a review of the current literature; but also draw attention to the confusing nomenclature at-

tributed to these lesions, potentially resulting in inappropriate management.

Historically, the literature on vascular anomalies of the head and neck has lacked consistent nomenclature, contributing to improper diagnosis and inappropriate treatment, and ultimately making clinical comparisons difficult. The term “haemangioma” has often been used to describe any vascular lesion and is commonly preceded by descriptive but unhelpful terms, such as “cavernous” and “capillary”.

In 1982, Mulliken and Glowacki developed a new system of classification of cervicofacial vascular anomalies that correlated clinical presentation and growth behaviour with histopathological features [8]. Vascular anomalies were thus divided into 2 main categories: haemangiomas and vascular malformations. Subsequent revision has classed haemangiomas as vascular tumours, whilst vascular malformations were further subdivided into capillary, venous, lymphatic and arteriovenous malformations (AVM) [10]. Understanding the fundamental distinctions of these 2 classifications will provide guidelines for prognosis and facilitate appropriate management of these lesions, even if they occur on an unusual site like the tympanic membrane.

Case Report

A 49-year-old lady presented to our department with a 6-month history of hearing loss and intermittent otalgia in her right ear. She had no other otologic history; and review of her systems, family and medical histories was all unremarkable. Otoscopic examination of her left tympanic membrane was normal. On the right side, she had a deep red, well-circumscribed small protuberant lesion in the antero-inferior aspect (Fig. 1). The lesion showed no evidence of blanching with mild pressure. On tuning fork tests, Rinne’s was positive bilaterally and Weber’s was centralised. Audiological testing revealed bilaterally normal tympanograms with an essentially symmetrical pure tone audiogram. Full cranial

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Fig. 1 View of vascular lesion on the antero-inferior aspect of the right tympanic membrane

nerve examination was normal, and there were no vascular lesions elsewhere on the body.

CT scan of the temporal bone was obtained which revealed the lesion to be confined to the tympanic membrane with a normal middle ear cleft.

Due to the lack of symptom severity and reluctance on the patient's part, surgical intervention was deferred, and a watch-and-wait policy adopted. 6-monthly reviews over an 18-month period showed neither alterations in size nor in the patient's symptoms. We continue to follow this patient up at 6-monthly intervals.

Discussion

Vascular anomalies are classified into 2 main categories of haemangiomas and vascular malformations, characterised by differences in their natural history and histology (Table I).

Haemangiomas are the most common tumours of infancy, with more than 60% of all haemangiomas occurring in the head and neck [9]. The natural history is characterised by rapid proliferation followed by a slow period of spontaneous involution. Growth during the proliferative phase is embodied by endothelial hyperplasia and an increase in the number of mast cells [8]. During the involution phase, endothelial cell activity decreases with normalisation of mast cell count and infiltration of the cellular parenchyma by fibrofatty tissue [8]. Signs of involution include shrinkage, central fading and softening. Complete resolution occurs in 50% of cases by 5 years; 70% by 7 years with the remaining 30% resolving to varying degrees over time [9]. The rate and extent of regression is not predictable. Complications such as bleeding, ulceration, and in the case of the ear, refractory otitis externa or conductive hearing loss, occur in 10–20% of haemangiomas [9]. Options for definitive management include observation, surgery, laser treatment, corticosteroid or alpha-interferon (α -INF) therapy [10]. The latter two are believed to target angiogenesis. As the vast majority of haemangiomas are small, well-circumscribed and involute with no sequelae, expectant management remains the preferred option.

In contrast, vascular malformations display no proliferative cellular activity. They exhibit flat, quiescent endothelium and normal mast cell concentrations. Vascular malformations are considered errors of vascular morphogenesis, displaying abnormal vessel dilatations but no increased cell turnover [8]. Despite stability from a cellular standpoint, vascular malformations can be clinically devastating as they are always present at birth, grow in proportion to total body growth and never regress. These structural abnormalities often display interval expansion with changes in blood flow, collateral formation or hormonal modulation [9]. Many venous malformations do not require specific management apart from reassurance and explanation of the natural history of the lesion. Intervention depends on the size of the lesion and its functional or cosmetic impact on the patient. Interstitial laser therapy (eg. Nd:YAG) may prove a useful

Table 1 Comparison of haemangioma and vascular malformation

	Haemangioma	Vascular Malformation
Histology	Proliferative cellular process	Error in vascular morphogenesis
Demographics	Tends to occur in infants	Always present at birth
Progression	Rapid proliferation followed by involution	Grow in proportion to total body growth; never regresses Capacity for interval expansion
Management Options	Observation Surgery Laser Corticosteroid (intralesional or systemic) Interferon therapy	Observation Surgery Laser
Classification	“Capillary haemangioma” is a true haemangioma	“Cavernous haemangioma” is a misnomer. It should be renamed vascular malformation.

therapeutic option as pharmacological intervention plays no role in these non-proliferative lesions [4, 9].

In light of these fundamental distinctions, lesions erroneously termed “cavernous haemangiomas” are more appropriately classified as vascular malformations as they represent errors in morphogenesis rather than proliferative cellular processes, and they are best approached by considering their growth over the life of the individual. Conversely, “capillary haemangiomas” describe true haemangiomas, and they are best evaluated from a perspective of proliferation followed inevitably by involution. Furthermore, the vast majority of vascular lesions can be accurately classified into the appropriate category of haemangioma or vascular malformation based on a sound history and physical examination. Angiography and biopsy are not generally necessary to make the diagnosis [9].

Of the 7 reported cases of vascular lesions arising from the tympanic membrane [1–7], if we evaluate the case synopsis and brief histological descriptions, 2 may be classified as haemangiomas [1, 6] with the remaining 5 more appropriately renamed vascular malformations [2–5, 7]. Magliulo and Fusconi had classed their case as a capillary haemangioma [3], however, based on the lack of clinical progress over a 15-month period, this lesion may be more appropriately reclassified as a vascular malformation.

With regards to our own case, based on the patient's history and stagnant clinical progression over time, we are inclined to classify this patient's lesion as a vascular malformation; however, without the benefit of histology or information on the presence of the lesion in the patient's paediatric history, longer-term follow-up is required to conclusively label it as such. Knowing the natural history and benign course of vascular malformations, it is thus appropriate to leave such cases alone if they are asymptomatic and do not show interval growth. This is evident in both our case and that of Magliulo and Fusconi [3] where conservative follow-up over a >12-month period showed neither growth of the lesion nor progression of symptoms.

In cases where surgical excision and subsequent reconstruction was performed [1, 2, 5–7], these were carried out due to symptoms of hearing loss, otalgia or otorrhoea or for histological diagnosis due to atypical features [1]. In the case of Balkany et al [6], they chose initially to monitor the lesion conservatively; however, surgical excision was performed when the tumour doubled in size over 18 months. Virtually no information has been given on the post-operative symptomatic outcome of these patients. Hiraumi et al have, however, reported the absence of recurrence over 3 years in their case despite positive resection margins [1].

Surgically-resected lesions (both haemangiomas and vascular malformations) were found to arise from the lamina propria. This correlates with all the cases, including our own, where the lesion was not found to invade the tympanic cavity. It was thus suggested that it was possible to remove the lesion without violating the mucosal layer

facing the tympanic cavity, thereby significantly increasing the chances of successful grafting [5].

It is also known that skin and mucosal haemangiomas often appear as multiple lesions; all but one of the previously reported cases have been isolated to the tympanic membrane, as has our case. In the case of Bijelic et al, an indication of a haemorrhage of a small vascular malformation in the patient's cerebral peduncle was found on MRI of the head [2].

An interesting observation in our case is the location of the vascular lesion on the tympanic membrane. Of the cited 7 cases, 6 were found to arise from the postero-superior quadrant of the tympanic membrane [1–4, 6, 7] with the remaining case arising centrally [5]. In contrast, our case showed the lesion to arise from the antero-inferior quadrant of the tympanic membrane. The significance of the location on the tympanic membrane remains unclear as neither characteristics of the lesion nor presentation of symptoms deferred from the other cases.

In conclusion, this case demonstrates the rare presentation of a vascular lesion on the tympanic membrane in a novel location. We also discuss the need for accurate classification of such lesions, and advocate confusing terms such as “cavernous” or “capillary” haemangioma to be discarded. The more universal classification of “haemangioma” and “vascular malformation” should be employed instead.

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