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Commentary

The Archaic Assembly - Lipochoristoma

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Preface

Commonly, neoplasms arising within internal acoustic canal are derived from the neuro-epithelium. Lipochoristoma is an exceptional, benign tumefaction emerging from cranial nerve VIII or vestibulocochlear nerve and appears confined to internal acoustic canal or the cerebello-pontine angle. Lipochoristoma is additionally denominated as lipomatous choristoma, lipomatous hamartoma, glioneural hamartoma, ectomesenchymal hamartoma, neuromuscular choristoma, neuromuscular hamartoma or lipoma.

As the neoplasm is posited to arise from mesenchymal derivative of the neural crest, tumefaction was previously and erroneously addressed as lipoma of internal acoustic canal or cerebello-pontine angle. The neoplasm demonstrates typical, diagnostic features upon magnetic resonance imaging (MRI). Cogent radiographic assessment of the exceptional neoplasm can be challenging and tumefaction can be misinterpreted as vestibular schwannoma. Appropriate differentiation of neoplasms arising within the internal acoustic canal is crucial as individual, pertinent lesions mandate distinctive and adequate therapy.

Disease characteristics

Choristoma is a tumefaction engendered by development of specific tissue subtypes aberrant to site of emerging neoplasm. Lipochoristoma comprises of around 0.15% neoplasms arising within the cerebello-pontine angle. The exceptional, indolent, gradually progressive tumefaction may be congenital and is discovered incidentally [1,2].

A male predominance is observed with an estimated 70% lesions appearing in male subjects [1,2].

Characteristically, the neoplasm emerges from cells of meninx primitiva which is a mesenchymal derivative of neural crest associated with anomalous differentiation [1,2].

Mesenchyme endogenous to vestibulocochlear nerve is posited to engender the neoplasm which is appropriately characterized as lipomatous choristoma. Lipochoristoma appears intensely concordant to the auditory nerve [1,2].

The premise of congenital origin indicates that lipochoristoma arises from an aberrant reabsorption of primitive, intrinsic meningeal tissue. Mesenchymal elements appear as integral components of the acoustic nerve, especially within preliminary embryonic development [2,3].

Tumour growth possibly occurs due to fluctuation within visceral adipose tissue as the neoplasm is predominantly composed of mature adipocytes [2,3].

Clinical elucidation

The indolent lipochoristoma engenders definitive clinical symptoms as progressive, severe, sensi-neural deafness, vestibular symptoms, vertigo, chronic headache, facial paraesthesia or tinnitus [3,4].

Pertinent, frequent, cochleo-vestibular clinical symptoms arising from neoplasms of internal acoustic canal appear as deafness, tinnitus and dizziness. Additionally, symptoms such as headache, neuralgia, paraesthesia of the trigeminal nerve, hemi-facial spasm, facial paralysis, pain within the ear, dyskinesia and dysphagia may be discerned [3,4]. Surgical resection of the neoplasm may endanger hearing and generate deafness in incriminated individuals [3,4].

Histological elucidation

Choristoma is composed of aggregates of mature, non dysplastic adipose tissue cells devoid of cellular or nuclear atypia admixed with variable quantities of fibro-vascular tissue [5,6].

Upon gross examination, the neoplasm appears to arise from cochlear or vestibular branch of cranial nerve VIII and manifests as a soft, rubbery, unilateral, ivory, pink, grey or maroon tumefaction [5,6].

Upon microscopy, normal components of incriminated cranial nerve such as myelinated nerve branches, glial cells, neurons, miniature, thin- walled vascular articulations and mature adipose tissue appear commingled with variable quantities of mature fibrous tissue, tortuous, thick- walled vascular configurations, fascicles of smooth muscle and skeletal muscle fibres [5,6].

Morphologically, tumefaction is composed of quantifiable mature adipocytes, branches of myelinated nerve, glial cells, neurons, miniature, thin-walled vascular articulations, smooth muscle cells which appear distinct from vascular articulations, mature, hyalinised fibrous tissue along with intermixed skeletal muscle fibres [5,6].

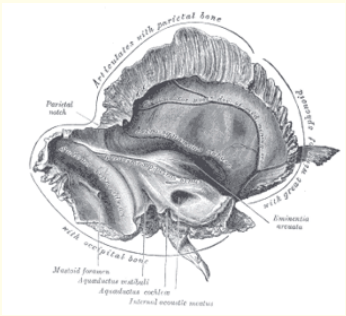

Figure 1: Anatomy of the internal acoustic canal with pertinent structures [10].

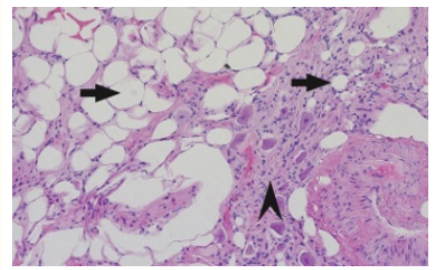

Figure 2: Lipochoristoma of internal acoustic canal demonstrating mature adipose tissue admixed with variable quantities of fibrous tissue, smooth muscle and skeletal muscle fibres and thin-walled vascular articulations [11].

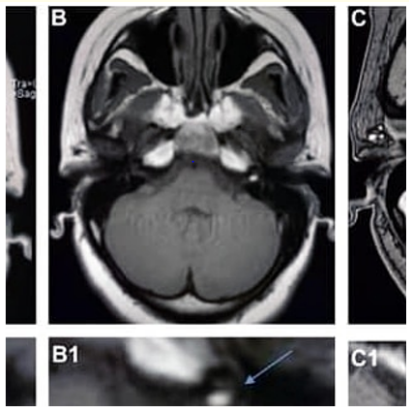

Figure 3: Lipochoristoma depicting hyper-intense signals upon T1 weighted magnetic resonance imaging [12].

Investigative assay

Magnetic resonance imaging (MRI) is an optimal radiological modality for appropriately assessing lipochoristoma. A miniature, lobulated, heterogeneous neoplasm within the internal acoustic canal with hyper-intense signal intensity is observed upon non contrast T1 weighted axial magnetic resonance imaging. Upon T1 weighted imaging, lipochoristoma displays a bright signal, in contrast to frequently discerned schwannoma [7,8].

Interval magnetic resonance imaging (MRI) with administration of gadolinium contrast medium is recommended for cogent tumour assessment [7,8].

Fat suppressed sequencing upon magnetic resonance imaging depicts a lack of enhancement of the lesion. Fat suppression upon unenhanced T1 weighted imaging is diagnostic of lipochoristoma [7,8].

MRI with non contrast T1 weighted axial imaging and fat suppression sequences can be beneficially adopted to segregate diverse lesions arising within the internal acoustic canal. Lipochoristoma can simulate a schwannoma and displays a hyper-intense signal upon T1 weighted imaging. Occasionally, lipochoristoma depicts an isointense or hypo-intense signal upon T1 weighted imaging [7,8].

Contrast assisted T1 weighted axial imaging upon MRI displays a heterogeneous enhancement of the lesion. Post-contrast images of lipochoristoma following administration of gadolinium contrast lacks enhancement, in contrast to T1 weighted imaging [7,8].

Gadolinium contrast medium demonstrates diverse patterns of image enhancement contingent to quantifiable adipose tissue component of the neoplasm. Lipochoristoma enunciating significant adipose tissue component is devoid of image enhancement. Nevertheless, non adipose tissue elements display appreciable image enhancement [7,8].

T2 weighted axial imaging demonstrates a heterogeneous lesion of variable magnitude [7,8].

Therapeutic options

Conservative management is an optimal and recommended strategy for treating lipochoristoma as hearing may be appropriately preserved. Tumour evaluation of the indolent neoplasm intimately intertwined with auditory nerve can be achieved with interval imaging which is beneficial and recommended.

Extended follow up delineates an absence of tumour reoccurrence. Persistent or permanent deafness can be treated with bone-anchored hearing aid [7,8].

Comprehensive surgical excision of the neoplasm can be employed although surgical intervention is associated with significant morbidity. Comprehensive eradication of the gradually progressive, infiltrative neoplasm with significant fibrous component may be challenging [7,8].

Besides, auditory preservation with surgical intervention may be inadequate due to preponderant interrelation between the lesion and auditory nerve [8,9].

Surgical intervention is recommended and reserved for symptomatic, progressive neoplasms associated with disabling vestibular symptoms.

Subtotal surgical resection is adopted in subjects with intractable clinical symptoms unresponsive to medical or rehabilitative therapy [8,9].

Frequently discerned postoperative complications appear as deafness, facial paresis and cranial neuropathies. Postoperatively, enhanced possible occurrence of cranial neuropathies or associated complications is observed in an estimated >70% instances. Discernible surgical complications appear as deafness, tinnitus, vertigo, deficit of trigeminal nerve, facial paresis, ataxia, hemiparesis, diplopia and additional neurological exigencies [8,9].

Lipochoristoma treated conservatively or with partial excision or resected for tissue sampling is devoid of tumour expansion or reoccurrence [8,9].

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10. Image 1 Courtesy: Wikipedia.
11. Image 2 Courtesy: Science direct.
12. Image 3 Courtesy: Semantic scholar.

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