



Diagnosis and management of middle ear hemangioma

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Abstract

Hemangioma is a benign tumor of blood vessels that is commonly found in the head and neck region. Hemangioma consists of two main types, capillary and cavernous hemangiomas. Hemangioma is predominantly affects women compared to men with a ratio of 6:1. Middle ear hemangioma diagnosis can be established based on history, physical examination, and additional work-ups. Several approach of therapies that could be performed include surgery preceded by ligation, radiotherapy, steroids and embolization. The main treatment of choice for hemangiomas is surgery.

Keywords: Hemangioma, middle ear, tumor

Introduction

Hemangioma is a benign tumor of blood vessels that is commonly found in the head and neck region. These tumors are not only found on the surface of the scalp and neck but also in the mucosa. It is predominantly found in the head and neck at birth, during infancy and childhood. It is congenital in nature, which results from the development of mesenchymal remnants through the differentiation and proliferation of blood vessel endothelium.

Lesions of blood vessels according to clinical and histological characteristics can be classified into two, namely: hemangiomas vascular malformations. Histologically, hemangiomas are classified into capillary hemangioma, cavernous hemangioma and combination of capillary and cavernous hemangioma. Congenital hemangioma may occur immediately after birth or appear several weeks after birth, hemangioma in neonates can be found in 2.6% and 10% at one year of age. Hemangioma is predominantly affects women compared to men with a ratio of 6:1. Congenital hemangioma is a relatively rare disorder and may involve the skin surface of the head-neck region and mucosa.

Definitions

Hemangiomas are defined as benign tumors of blood vessels, which originate from the skin, mucosa, and deeper structures including bones, muscles, and glands. Hemangiomas consists of two main types, capillary and cavernous hemangiomas. In addition, there is a mixed-type hemangioma, combination between the two.

Anatomy

Middle ear is a dome-shaped structures with six sides. Middle ear is divided into three parts from the cephalad to caudal, where epitympanum is located above the superior margin of the tympanic membrane, the mesotympanum is also called the tympanic cavity which is located medially to the tympanic membrane, and the hypotympanum is located caudal to the tympanic membrane. The function of middle ear is mainly to transmit sound from the external into the inner ear. The anterior border of the ear space is limited by the tympanic membrane, while the inner side is limited by the oval and round window. Middle ear space consists of

several parts consist of tympanic membrane, the ossicles, and the Eustachian tube.

There are two muscles in the middle ear, namely *m. tensor tympani* and *m. stapedius*. The tensor tympani muscle originates from semi-canal of the tensor tympanic with insertion above the malleus bone, innervated by a branch of the trigeminal nerve. This muscle pulled the tympanic membrane inwards to become more tense and increases the resonant frequency of the sound- conducting system and weakens low- frequency sounds. Stapedius muscle originates in the eminence of the pyramid and inserts at the posterior end of the stapes column, this leads to stiffening of the stapes, weakens sound transmission, and increases the resonance of the ossicles. The function of these two muscles are to maintain, to strengthen the ossicle chain, and to dampen sounds that are too loud as a protective mechanisms to prevent cochlear damage.

Arteries supply to the tympanic cavity are anterior tympanic artery, stylomastoid artery, superficial petrosal artery, and inferior tympanic artery. The venous blood flows with the arterial flow accordingly and drains into the superior petrosal sinus and pterygoid plexus.

The tympanic membrane acts as a sound waves receiver. Every time a sound wave enters the ear canal, it will hit the tympanic membrane, then the tympanic membrane will bulge inward towards the middle ear, touch the ossicles or adutiroy bones, namely the malleus, incus, and stapes. These auditory bones will transmit the sound waves further into the inner ear.

The auditory ossicles consist of malleus (hammer bone), incus (anvil bone) and stapes (stirrup). These three bones form a series of bones that cross the middle ear and fuse with the tympanic membrane. Eustachian auditory tube or Eustachiusis channel is a connecting tube between the middle ear and the pharynx. Eustachian tube allows us to balance the air pressure between the middle ear cavity with the external atmosphere.

Histopathology

The general structure and composition of blood vessels is nearly similar throughout the whole cardiovascular system. The composition of vascular walls includes extracellular matrix (ECM) which contains elastin, collagen, and

glycosaminoglycan's. The vascular walls consist of three parts: tunica intima, tunica media, and tunica adventitia. The boundary between the tunica intima and tunica media is called the internal elastic lamina, while the boundary between the tunica media and tunica adventitia is the external elastic lamina. In a normal artery, the tunica intima consists of monolayer cells and the ECM which is surrounded by connective tissue, nerve fibers, and small blood vessels from the adventitia. The tunica media receives nutrients and oxygen from the vascular lumen. Blood vessels consist of the following:

- a. The tunica intima (tunica interna) consists of a layer of endothelial cells that line the inner surface of the vessels. Beneath the endothelium is the subendothelial layer, consisting of sparse smooth connective tissue containing smooth muscle cells whose role is to contract blood vessels.
- b. The tunica media consists of smooth muscle cells arranged in a circular manner. In arteries, tunica media is separated from the tunica intima by an internal elastic membrane. This membrane is composed of elastin, with fenestration so that substances are able to diffuse through fenestration of the membrane and to provide those cells lay deep in the vessel wall. In large vessels, external elastic membrane that separates the tunica media from the tunica adventitia is thinner.
- c. The tunica adventitia consists of connective tissue with elastin fibers. In the larger vessels, the vasa vasorum (vessels within vessels) branch extensively in the tunica adventitia.

Until recently, the exact pathogenesis of hemangioma is not fully understood. There are multiple theories regarding the development of hemangiomas. The current mostly accepted theory is that hemangiomas result from the development of mesenchymal remnants that underwent differentiation and proliferation as blood vessels. Histologically, hemangioma is divided into 3 major categories, namely capillary hemangioma, cavernous hemangioma, and combination of capillary and cavernous hemangioma.

Capillary hemangiomas are lesions predominantly affecting the cutaneous blood vessels. Generally, the lesions are more superficial, tend to be of the capillary type, and appear as pink-colored papular areas forming lobules. Microscopic appearance of the lesion is a clustered-epithelial-cells with multiple capillary channels. This tumor consists of new capillaries filled with blood and forms a network. This tumor only affects one segment of the blood vessels. During the involution phase, there is narrowing and occlusion of the capillary lumen, which then followed by capillary involution with an increase of connective tissue stroma.

Cavernous hemangioma is a less common lesion and located deeper compared to capillary hemangioma. The histologic feature consists of large vascular channels lined with endothelial cells. Occasionally, fibrous tissue in the channels of blood vessels can be seen. These tumors consist of sinusoidal spaces lined by endothelial cells and filled with blood. The lumen of the vessels is wide and thin-walled, irregular, and surrounded by a thick fibrous tissue. It is usually appeared as a bluish-red protrusion arising from the surface. These cavernous lesions could be permanent, especially if the lesions are not present at birth. When occurs at birth, it tends to regress spontaneously. Organs that are commonly affected are the parotid gland,

musculoskeletal, larynx, mandibula, maxillary, lip, and nose. Mixed type hemangiomas are more common-compared to cavernous hemangiomas. Usually this affects the parotid gland, more often in children, which present immediately after birth. The histological picture reveals alternating large and small blood vessels.

Etiology

Until recently, the exact etiology of middle ear hemangioma is not clearly elucidated. Vascular malformations are thought to originate from remnants of mesodermal tissue that fail to form normal connections with the vascular system. Some literature stated that hemangioma proliferation has a high level of basic fibroblast growth factors, proliferating cell nuclear antigen, type IV collagenase, vascular endothelial growth factors, urikinase, and more recently including insulin growth factors. Involution of hemangioma is characterized by the appearance of tissue metalloproteinase-inhibitors, thrombospondin, angiostatin, endostatin, in terferon- α , and reduced hemangioma proliferative factors.

Diagnosis

Middle ear hemangioma diagnosis can be established based on history, physical examination, and additional work-ups.

a. History

Symptoms and signs of middle ear hemangioma are including hearing loss, tinnitus, otorrhea, ear pain, and facial nerve palsy. Hearing loss is the main feature of those with a middle ear hemangioma, it occurs unilaterally on the side affected by the tumor. Hearing loss may progressively getting worse, and becomes the chief complaints that bring patients to seek for treatment.

b. Physical examination

On otoscopic examination, an erythematous mass may be found in the retrotympanic area which can fill 2/3 of the middle ear. On microscopic examination, clinician may observe a retrotympanic mass with a "cobblestoned" surface appearance, indicating the presence of middle ear hemangioma. Tympanometry and audiometry examinations are necessary for middle ear hemangiomas, where audiometry might reveals conduction-type hearing loss in the affected ear with normal hearing in the unaffected ear.

c. Additional Work-ups

There are no pathognomonic radiological findings of middle ear hemangioma. It has been reported that there is a possibility of bone alteration, however it can be challenging to distinguish them from malignant tumors in the setting of extensive destruction at the skull base. On radiological examination, CT scan, and MRI are required to determine the extent of the tumor and the involvement of other structures to assist the diagnosis. MRI can be used to exclude visceral involvement, while CT scan can be used to determine the tumor size and presence of invasion of surrounding structures. Vascular tumors revealed a contrast-enhanced appearance after contrast administration, indicating predominant vascularization. CT scan is reliable to detect bony structures, conversely it is difficult to distinguish tumor from inflammation, while MRI examination is superior to delineate soft tissue abnormalities. To establish a definite diagnosis of middle ear hemangioma, it must be based on pathological

examination. Generally, biopsy can be performed during surgery, due to the high risk of bleeding.

Management

In general, hemangiomas may resolve spontaneously, specifically hemangiomas that found at birth due to spontaneous involution that occurs during infancy. Further treatment should be instituted if involution does not occur and there is a possibility that expansive, ulcerative, or functional disturbances occur, making treatment more difficult. Several approach of therapies that could be performed include surgery preceded by ligation, radiotherapy, steroids and embolization. The main treatment of choice for hemangiomas is surgery.

a. Surgery

The surgical technique performed is depends on several factors including tumor involvement of the tympanic membrane, involvement of the ossicles, and tumor extension beyond the middle ear. For tumors confined to the middle ear, a transcanal approach with tympanotomy underlays tympanoplasty can be performed. Surgery for middle ear hemangioma was reported to have been carried out in 80.9% of cases, in which 65% of cases used transcanal approach with collapsed wall or intact wall approach. In 23% of cases, removal of the ossicles was performed, with most of the cases involving resection of the incus and malleus. In some cases, reconstructive measures are performed using ossicular prostheses and fascia grafts. Reconstruction can also be done with stapes osciculoplasty.

Total resection was performed in 94.1% of cases. Hiraumi *et al.*, reported that subtotal excision may be performed depending on the extent of the tumor being clearly visible at the time of surgery. Histological analysis provides a clear information regarding the tumor margins.

Kostrzewa *et al.*, reported that in addition to tumor resection, mastoidectomy and wall down tympanoplasty can be performed using a CO2 laser during the procedure. Using CO2 laser during surgery may allow to shrink tissue relatively easier during dissection, reduces the risk of bleeding and provide a good visualization during the procedure.

In addition to tumor resection and mastoidectomy with a CO2 laser, Kumbhar *et al* stated that middle ear hemangiomas can be managed with neurointervention techniques. Procedures of neurointervention techniques are carried out under general anesthesia, femoral artery is used for arterial access access. External carotid artery angiogram by submentvertical and Schuller approach shows a clear staining of tissue in the mastoid and middle ear. Hemangiomas receive their blood supply from small arterial branches from external carotid artery. Direct aspiration of the hemangioma can be performed using a 24-gauge needle in the temporal vein. After an adequate blood flow has been confirmed, embolization was performed using 2 cc of 16% n-butyl- cyanoacrylate glue followed by administration of 3 cc of 66% sodium tetradecyl sulphate. Post-embolization angiogram shows mild vascularity in the occipital area supplied by branches of the occipital artery. Embolization was further performed on the distal of occipital artery using 100-200 microns of polyvinyl alcohol. Post embolization angiogram reveals a complete resolution of the hemangioma.s

b. Conservative

In general, hemangiomas may resolve without treatment, especially hemangiomas that occur at birth due to spontaneous involution. Some cases reported the occurrence of spontaneous involution and may fully regress within 6 months, results in satisfactory hearing function. Spontaneous involution of hemangiomas in children may be explained by the different biological entities compared to adults, thus hemangiomas in children have a higher tendency towards spontaneous involution.

c. Other therapeutic modalities

In addition to surgical and conservative therapy. Pavamani *et al* reported that cases of middle ear hemangioma can be treated using targeted radiotherapy. Hemangiomas are reported to be radiosensitive and this radiotherapy can be used in hemangioma both in children and adults. In cases of middle ear hemangiomas in children who are unable to undergo surgery, radiotherapy is the acceptable treatment option.

Radiotherapy also has minimal long-term side effects.

Prognosis

The prognosis for middle ear hemangiomas is generally good when spontaneous involution occurs or total resection can be performed. Hemangiomas have a high recurrence rate (16-23%) depends on the quality of tumor resection.

Discussion

Congenital hemangioma may occur immediately after birth or appear several weeks after birth, hemangioma in neonates occur in 2.6% and 10% at the age of one. Hemangioma is more common in women compared to men with a ratio of 6:1. Congenital hemangioma is a relatively rare disorder and may affects the skin surface of the head-neck area and mucosa. According to Nouri *et al*, hemangiomas in the middle ear are rare and have an incidence rate of 0.21% of all tumors in the middle ear. Histologically, hemangioma is divided into 3 major categories, namely capillary hemangioma, cavernous hemangioma, and mixed capillary and cavernous hemangioma. According to Pistorio *et al.*, hemangioma of the capillary hemangioma is mostly found in the middle ear, however it is difficult to evaluate.

The etiology of middle ear hemangioma is not known with certainty. According to Pistorio *et al.*, middle ear hemangiomas can occur due to vascular malformations that are thought to originate from remnants of mesodermal tissue that fail to form normal connections with the vascular system.

The diagnosis of middle ear hemangioma can be established based on history, physical examination and supporting examinations. Cui-ping *et al.*, said that a definite diagnosis of hemangioma of the middle ear capillaries was established based on histopathological examination. Histopathological examination showed that the microscopic appearance was epithelial cells in groups with multiple capillary channels. This tumor consists of new capillaries filled with blood and forms a network.

Some of the therapies that can be performed include surgery preceded by ligation, radiotherapy, steroids, and embolization, where the main treatment of choice for hemangioma is surgical procedure. Hiraumi *et al.*, reported that subtotal excision may be performed depends on the

extent of the tumor being clearly visible at the time of surgery. Histological test may provide a clear information regarding the the tumor margin. Kostrzewa *et al* reported that in addition to tumor resection, mastoidectomy and collapsing tympanoplasty can be performed using a CO2 laser during the procedure. Kumbhar *et al* stated that middle ear hemangiomas can be managed using neurointervention techniques.

Conclusion

Hemangiomas are benign tumors of blood vessels, originating from the skin, mucosa, and deeper structures including bones, muscles, and glands. Until recently, the exact etiology of middle ear hemangioma is not fully understood. Vascular malformations are thought to originate from remnants of mesodermal tissue that fail to form normal connections with the vascular system. Diagnosis of middle ear hemangioma can be established based on history, physical examination, and additional work-ups. The definitive diagnosis is established according to histopathological examination. In general, hemangiomas may resolve spontaneously without any treatment, specifically hemangiomas that occur at birth due to spontaneous involution that occurs during infancy.

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