CASE REPORT

A RARE CASE OF VASCULAR HAMARTOMA IN NECK OF A 50 YEAR MALE

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ABSTRACT: Neck is complex anatomical site with a large list of possible diagnoses for swelling. Most common ones being Lymph Nodes, Thyroid. Occasionally there may be Cystic lesions that are difficult to conclude. Hamartomas in the head and neck region are unusual, and few reports of such lesions are found in the literature. There are few cases of vascular hamartoma reported in children but are rare in adults. This case report describes one such case where it was difficult to conclude.

KEYWORDS: Vascular Hamartoma, Malformation

INTRODUCTION: A hamartoma (From Greek hamartion "bodily defect") is a benign (noncancerous) tumor like malformation made up of an excessive, focal overgrowth of cells and tissues native to the organ in which it occurs. Hamartomas in the head and neck region are unusual. Diagnosed by US Doppler, CT Scan, MRI, & MR Angiogram. Treated by Laser Ablation, Embolization & surgical Excision

CASE REPORT: A 50 year old male patient presented with swelling in the left side of the neck noticed 20 days back which did not show change in size since then. Swelling was associated with slight dull aching pain. Other than swelling and pain patient had no specific complaints. No history of swelling in other sites, fever, cough, hemoptysis, difficulty in swallowing/breathing, no features of hypo/hyperthyroidism & previous surgery.

On examination, a solitary swelling of 6cms x 5cms beneath the left sternocleidomastoid muscle, ill-defined borders, cystic in consistency, non-compressible, fluctuant, non-tender, with restricted mobility noted in the posterior triangle of neck.

Ultrasound of neck showed a cystic lesion measuring 5.3x4.9x3.5cms with internal echoes & septation in the posterior triangle of neck extending anteriorly into carotid triangle.

CT Scan Neck (Plain & Contrast) showed A well-defined non-enhancing cystic lesion of 5.8x5.0cms with internal septation in the posterior triangle of neck, poster lateral to carotid space. Medially abutting the left thyroid lobe compressing left internal jugular vein & laterally abutting external jugular vein. No evidence of calcification/solid components. Likely lymphangioma

FNAC of the swelling showed scantily cellular, occasional cyst macrophages & lymphocytes against an amorphous eosinophilic background suggestive of benign cystic lesion.

Informed consent for excision of cyst was taken. Under general anesthesia patient in supine position skin crease incision was put, tissues dissected in layers, a brown fluid filled lesion identified deep to left sternocleidomastoid abutting internal jugular vein, swelling was excised enmasse sent for histopathological examination though cyst ruptured and little fluid was spilt.

After confirming complete hemostasis wound closure done in layers with suction tube drain. Drain was removed on 3rd postoperative day and discharged. Suture removal done on 7th postoperative day.

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Histopathological report showed cyst wall line with Cuboidal cells and Hob nail cells. Wall is fibrovascular with few lymphoid aggregates & many congested capillaries and proliferating blood vessels. No atypia seen. Features suggestive of Vascular Hamartoma.

Swelling in the Left side of the Neck

Cystic lesion in USG Neck

Well defined Cystic lesion in the Neck

Nonenhancing Cystic lesion with Internalseptation

Per operative fluid filled lesion beneath Lt SCM

Gross specimen of Cystic lesion

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Microscopic view of lesion under Low power & Magnification View

Post-operative follow up of patient after 1 month

DISCUSSION: The term 'hamartoma' was introduced by Albrecht in 1904¹ to describe an inborn error of tissue development characterized by an abnormal mixture of tissues indigenous to the part, with excess of one or more. A hamartoma (from Greek hamartion "bodily defect") is a benign (noncancerous) tumor like malformation made up of an excessive, focal overgrowth of cells and tissues native to the organ in which it occurs. It is considered a developmental error and can occur at a number of sites.

Malformations represent primary errors of morphogenesis, in which there is an intrinsically abnormal developmental process. The line of demarcation between a hamartoma and a benign neoplasm is often unclear, as both lesions can be clonal.

Hemangiomas, lymphangiomas, rhabdomyomas of the heart, adenomas of the liver and developmental cysts within the kidneys, lungs, or pancreas are interpreted by some as hamartomas and by others as true neoplasms but are extremely rare in Neck. The frequency of these lesions in infancy and childhood and their clinical behavior give credence to the belief that many are developmental aberrations.

Vascular hamartomas are classified as capillary telangiectasis, cavernous angiomas, and arteriovenous or venous malformations. Hamartomas are usually congenital and simultaneously grow along the rest of the body. Once they reach their adult size they do not extend to involve more tissue unless there is trauma, infections, oedema, inflammation and filling of new vascular channels.³

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Lymphatic malformations or lymphangiomas of the head and neck usually involve the cervical area, in which case they are more commonly macrocystic and well demarcated.

INVESTIGATION AND DIAGNOSIS: With the increasing use of pre natal screening, congenital hemangiomas have been detected as early as 12th week of gestation. These tumors initially exhibit fast flow pattern by US and MRI. US with color flow Doppler can be extremely useful in differentiating hemangiomas from similarly appearing vascular malformations that have diminished to absent flow characteristics such as lymphatic malformations.

CT scan can delineate the extent and involvement of the hemangiomas and can also be useful in differentiating hemangiomas from lymphatic anomalies. MRI and MRA are useful to know about the extent of involvement with tissue planes and rheological characteristics.

TREATMENT: Venous malformations may be treated with laser, sclerotherapy, and/or surgical excision, depending on the depth, size, and location. Arteriovenous malformations require formal surgical resection with negative margins. Surgical excision is also required for lymphatic malformations, although superficial lesions are sometimes treatable with the CO₂ laser.

Hamartomas and Syndromes:

1. Cowden syndrome is a serious genetic disorder characterized by multiple hamartomas usually of skin, but also of thyroid, G.I tract, Bones, CNS, Eyes and G.U tract. Cowden's syndrome is considered part of PTEN hamartomas tumor syndrome (PHTS) which also includes Bannyan – Riley – Ruvalcaba syndrome, proteus syndrome, and proteus – like syndrome.
2. Tuberous sclerosis 2 is an autosomal dominant disease characterized by hamartomas of brain, retina, Pancreas, cardiac Rhabdomyomas, and mesenchymaltumors of kidney.

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