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Case report

Felicitous surgical management of a ponderously rare diagnosis of a Vascular Hamartoma in the neck of an adolescent female - A Case Report

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ABSTRACT

Introduction and importance: Hamartomas are defined as abnormal nonmalignant tissue malformations and are characterized by defected propagation of fully differentiated cells and soft tissues that are native to the affected organ. Etiology is either sporadic or congenital. Established incidence rates are only known for Pulmonary Hamartomas and are 0.25 %. Vascular Hamartomas are rarer and possess no known incidence rate. We must bear that diagnosis in mind when presented with such presentations.

Case presentation: We present the case of a 20-year-old Middle Eastern female, who presented to the General Surgery clinic with a two-month history of a gradually expansive bulge in the patient's right side of the neck. The bulge was painless and slowly increased in size. Preoperative radiological analysis demonstrated a cystic formation conformant with a vascular anomaly. Complete surgical excision of the mass was done, and histopathology revealed a Vascular Hamartoma.

Clinical discussion: Surgery was the modality of choice for treatment of our patient. Meticulous radiological analysis accompanied by informed clinical judgement were the gold standards for preoperative assessment. The patient underwent complete postoperative recovery and has been followed-up for 5 months thus far with no evidence of recurrence or complications.

Conclusion: In general, Hamartomas are profoundly rare occurrences, especially Vascular Hamartomas, and are even rarer in adolescent females. It is crucial to study and document such a rare diagnosis. This would facilitate performing epidemiological studies and enable surgeons to choose appropriate individualized therapeutic options.

1. Introduction

The nomenclature Hamartoma was first derived from the Greek term “Hamartia”, which means a character defect. Hamartomas are not classified as true neoplasia, rather are characterized by excessively defective hyperplasia and hypertrophy of one or multiple cellular classes. However, their cellular formation is disorganized unlike the organ tissue from which they arose. Additionally, they can originate from several organ sites as described in the literature [1].

The classical definition of Hamartoma in the current literature is an abnormal proliferation of fully differentiated cells in a nonneoplastic fashion. They are representative of the specific organ tissue of emergence. Multiple examples of occurrence sites exist. The most common sites are the lungs followed by the gastrointestinal tract, liver, skin, chest wall, and kidneys. They can further be organized into distinct classes based on which tissue constituent is prevalent. These include neurogenic, lipomatous, angiomatous, vascular, chondroid, and osseous origins [2].

With regards to incidence rates of Hamartomas, they're still poorly defined because they are eminently rare diagnoses. The sole exception is the pulmonary hamartoma as its incidence rate is estimated to be 0.25 % [3].

When diving into the subclassification of Vascular Hamartomas in

Abbreviations: BMI, Body Mass Index; CT, Computed Tomography; IV, Intravenous; MRI, Magnetic Resonance Imaging.

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Abbreviations

BMI	Body Mass Index
CT	Computed Tomography
IV	Intravenous
MRI	Magnetic Resonance Imaging

the young population groups, such as pediatrics or adolescents, most of those lesions occur in the form of a hemangioma. Furthermore, the most common occurrence location is the face [4].

These types of lesions are major causes for cosmetic complaints for affected patients. It is more dangerous when they arise from ocular tissues. This leads to numerous visual defects in said patients [5,6].

It is documented in the literature that the course which leads to the spontaneous regression of such lesions occurs at 7 years of age in 70 % of the cases. Nevertheless, we cannot allow for such potentially dangerous lesions to continue their progression hoping that they might spontaneously regress [7].

The work has been reported in line with the SCARE criteria and the revised 2020 SCARE guidelines [8].

2. Presentation of case

2.1. Patient information

Herein, we present the case of a previously healthy 20-year-old Middle Eastern female. She presented to our hospital's General Surgery outpatient clinic with the chief complaint of a bulge in her right supraclavicular region. Her story started two months prior to her admission when she noticed a gradually expanding bulge in the right side of her neck. It was sudden, painless, and gradually surging in size, particularly two weeks before her clinical presentation. Throughout the course of her complaint, our patient did not report any remarkable alterations in the skin overlying said neck mass, such as redness, hotness, ulceration, or hyper-/hypopigmentation.

No loss of appetite, hoarseness of voice, fatigue, malaise, fever, alterations of bowel habits, night sweats, or genitourinary complaints were reported by the patient.

The patient's surgical history was negative except for the presence of a similar occurrence at the same site 15 years ago. At that time, it was treated by needle aspiration in a rural hospital without any proper documentation. This hindered our ability to state whether this is a recurrence or not.

Our patient's family, drug, psychosocial, and allergic histories were negative.

She denied any exposure to chemotherapy or irradiation. Her Body Mass Index (BMI) was 25 kg/m^2.

2.2. Clinical findings

We started our **physical examination** by taking vital signs readings. Their results turned-out normal. **Via inspection** of the neck, we noticed a slight bulge in the patient's right supraclavicular region, specifically in the posterior triangle. We did not see any overlying skin ulceration, discoloration, nor hyper-/hypopigmentation. Inspection of the patient's left side of the neck was insignificant. **Through deep palpation,** a mass was felt. It was soft, immobile, with well-demarcated edges, and unattached to the overlying skin. Nonetheless, there weren't any palpable ipsilateral and/or contralateral axillary or supraclavicular lymph nodes. **Laboratory investigations** were done and yielded normal values.

2.3. Diagnostic assessment

Preoperative neck ultrasound was performed by a specialist and yielded positive findings. The report stated that there was a thin-walled cystic formation with clear contents. The demonstrated borders were well-demarcated without vascular inflow from another source. The lesion was situated behind the lower third of the Sternocleidomastoid muscle and measured approximately (4.7 × 3.3 × 3.5 cm). The extent of said cyst couldn't be vividly marked. Moreover, a second cystic formation with identical features was found lateral to the right internal jugular vein and estimated to measure (1.5 × 1 × 2 cm). No lymphadenopathy was demonstrated.

Based on the findings of the ultrasound, a form of lymphatic or vascular malformations was suspected.

To fully visualize the lesion and to carefully plan the surgical intervention, a **Computed Tomography (CT) scan** was done. It demonstrated findings which conform with those of the ultrasound. There was a right supraclavicular mass formation with clear content measuring (9 × 5 cm) (Fig. 1A–B).

No mediastinal nor supraclavicular lymphadenopathies were demonstrated. Furthermore, no ipsilateral pleural effusion was seen.

Based on what is common, what the radiological analysis revealed, and what was suitable to consider here, multiple differential diagnoses were thought of. Those were Lipoma, Dermoid Cyst, Lymphangioma, Hemangioma, Cystic Hygroma, or an Angiofibroma. A malignant differential diagnosis was -to a large extent- excluded because the visualized mass had clear content, it had well-demarcated borders, there was no vascular inflow into it from another source, no calcifications were seen, it wasn't invasive of neighboring soft tissues, and no family history of neoplasia was reported.

Presurgical optimization involved the administration of suitable preoperative antibiotics, setting-up an Intravenous (IV) access, and drawing blood samples for crossmatch.

Notable challenges were the unavailability of a Magnetic Resonance Imaging (MRI) device in the hospital at that time. In addition, the patient couldn't undergo an examination by an MRI or contrast-enhanced CT scan due to her low socioeconomic status.

2.4. Therapeutic intervention

The presenting clinical scenario warranted surgical excision of the lesion. The surgical procedure was successfully performed at our tertiary university hospital. Surgery was achieved by a General Surgery specialist and a senior resident General Surgery physician with 12 and 5 years of experience, respectively. It is worthy to note that the operation was seen through under general anesthesia with no perioperative complications.

A longitudinal incision was done along the extent of the mass to achieve proper anatomical exposure and to establish adequate surgical access. A cystic mass was found in the posterior triangle of the right side of the neck. It was medially bordered by the Sternocleidomastoid muscle and the Carotid Artery sheath. Posteriorly and laterally, it was bordered by the Trapezius muscle. Inferiorly, it was bordered by the Scalenus muscle. Furthermore, the mass was tightly attached to the surrounding structures. Its size was estimated to be (8 × 5 cm). We isolated it from its surroundings, and we noted the presence of its heavy vascularization along with the existence of oozing lymphatic discharge.

Intraoperative image was taken to demarcate the surgical site after resection of the mentioned mass (Fig. 2). We further demonstrate an intraoperative postresection image depicting the excised specimen (Fig. 3).

Said excised mass was directly sent for histopathological analysis.

The definitive diagnosis was reached postoperatively via histopathological analysis of the resected mass. Detailed histopathology report stated that the excised specimen grossly measured (5 × 3 × 2.5 cm). As for the microscopic examination, it revealed the presence of adipose

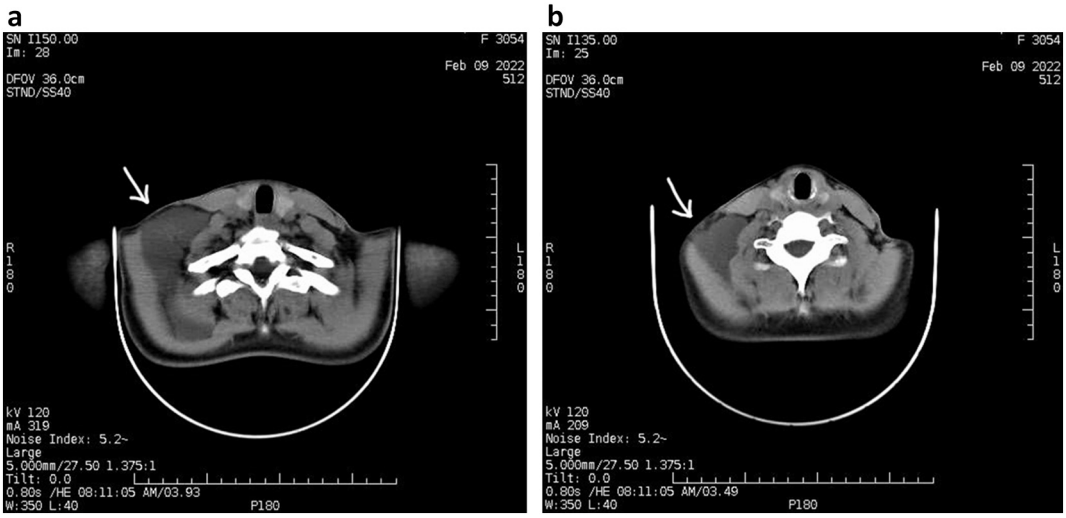

Fig. 1. (A–B): Preoperative **CT scan** demonstrating a clearly defined right supraclavicular mass formation with clear content measuring almost (9 × 5 cm) in length**.** The mass is depicted by the ***(Arrow).***

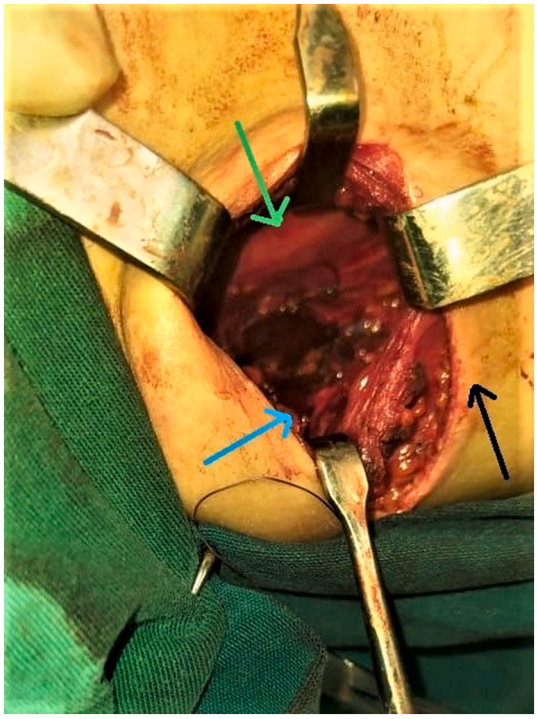

Fig. 2. Intraoperative postresection image depicting the site of the mass with its surrounding structures.
Black Arrow points towards the Clavicle. Blue Arrow points towards the Trapezius muscle. Green Arrow points towards the Sternocleidomastoid muscle.

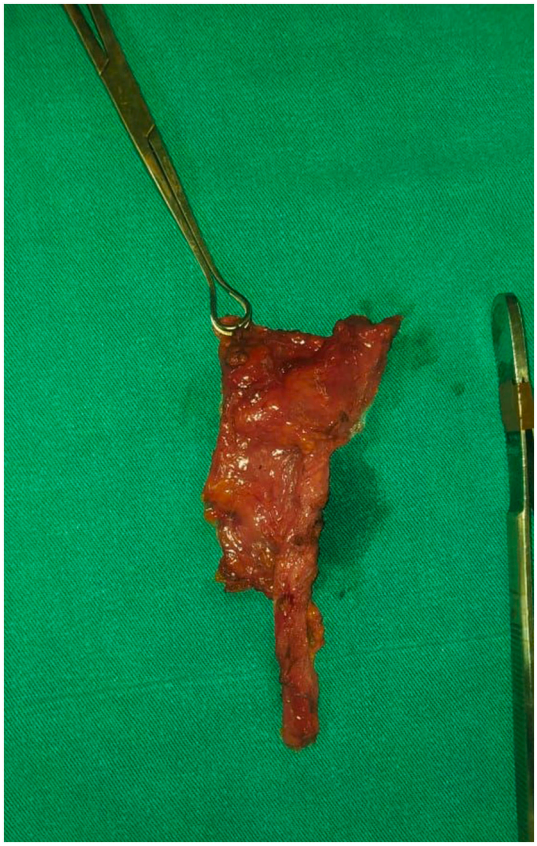

Fig. 3. Intraoperative postresection image revealing the excised specimen.

tissue with large, thickened blood vessel walls. In addition, we visualized small blood vessels lined by normal endothelial cells. Moreover, lymphatic vessels, fibrous tissue, muscle tissue, and nerve fibers were also seen. There were no traces of malignancy or atypia. In conclusion, the mass conforms with the diagnosis of a Vascular Hamartoma (Fig. 4A–E).

The patient underwent complete postsurgical recovery. She was discharged to the outpatient settings 3 days after her surgery. Furthermore, the concise nature of the lesion was thoroughly explained to her. We performed regular wound dressings for her in our clinic to aid in wound healing and in avoiding any wound complications.

Afterwards, we referred her to an oncologist specialized in such vascular anomalies for any necessary follow-up routine from an oncological standpoint.

Finally, she has been followed-up in our General Surgery clinic for 5 months so far where we meticulously examined her and the surgical site to ensure no recurrences have taken place. Her clinical and radiological examinations yielded normal results.

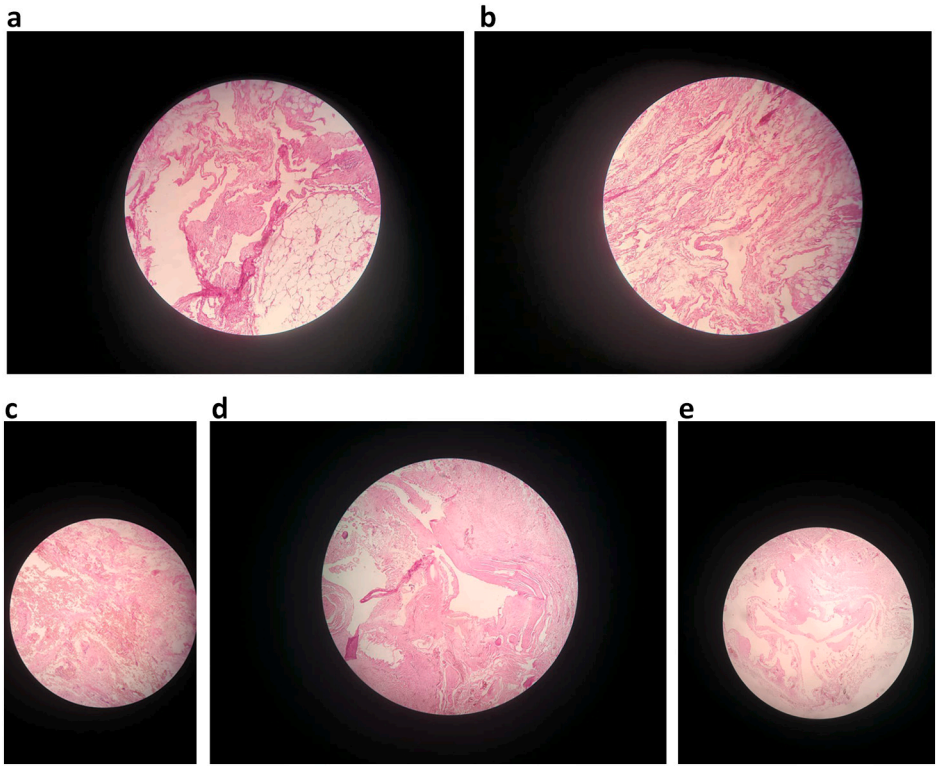

Fig. 4. (A–E): Histopathological Hematoxylin and Eosin staining. Detailed that the excised specimen grossly measured (5 × 3 × 2.5 cm). Microscopic findings revealed the presence of adipose tissue with large, thickened blood vessel walls, in addition to small blood vessels which are lined by normal endothelial cells, along with lymphatic vessels, fibrous tissue, muscle tissue, and nerve fibers. There were no traces of malignancy or atypia. The mass conforms with the diagnosis of a Vascular Hamartoma.

3. Discussion

Albrecht was the first researcher who coined the term "Hamartoma" in 1904 because he wanted to define the pathological combination of normal with congenitally abnormal cellular components of a tissue organ [9].

Afterwards, Hart was the pioneer to first to give pulmonary neoplasia such a term in 1906. As a result, it was defined as an abnormal nonmalignant tissue malformation branded by the defected propagation of fully differentiated cells and soft tissues native to the diseased organ [10].

It is described in the published literature that Hamartomas possess the capability to originate from any layer of the three known germ cell layers. Vascular Hamartomas could arise from any of those 3 main layers [10].

In 1982 came two renowned researchers called Mulliken and Glowacki, who in turn, were the first to depict a general categorization of abnormal vascular lesions based on histological, clinical, and cytological characteristics [11]. Said categorization was later validated by the general membership committee at the June Workshop on Vascular Birthmarks, which took place in Rome in 1996 [12,13].

It was hypothesized and later established that the incidence rate of Pulmonary Hamartomas is 0.25 %, whereas it is yet not known for hamartomas which arise from other organ sites [14].

To dive deeper into the classification of Vascular Hamartomas, we found that there are several distinct types, such as cavernous angiomas, telangiectasias, and venous or arteriovenous malformations. Said hamartomas are primarily congenital in origin and they develop simultaneously with the remaining body tissues. As soon as patients reach their adult size and once the growth phase is complete, hamartomas won't proliferate to include further soft tissues than the ones they originated from except when outside factors play a role, such as edema, different infections, traumatic events, or inflammations [15].

Classically, Vascular Hamartomas are asymptomatic even when the etiology is congenital. Furthermore, they might stay silent until different stages of adolescence or adulthood. Hamartomas are labeled as such due to the presence of abnormally proliferating distinct soft tissue classes, such as collagen fibers, fibrous tissues, adipose tissues, nerve bundles, and skeletal muscle fibers. These are chiefly located on the perimeters of this benign lesion. It doesn't usually possess a tissue capsule, and this yields distinct sizes of vascular masses. The unique abnormal combination of such tissues is exhibited in the native tissue of the organ of origin [14,16].

When it comes to the pathophysiology and rate of growth of Vascular Hamartomas, the published literature asserts that they could either proliferate sluggishly, swiftly, or remain of the same size. Nevertheless, Vascular Hamartomas possess the capability to swiftly hypertrophy out of proportion, in addition to the ability to spontaneously regress. Each special nature of each anatomical site's vascular formation is directly proportionate to the rate of growth of Vascular Hamartomas [17].

From a preoperative diagnostic standpoint, physicians are prime supporters of the utilization of ultrasound. This is due to its relatively cheap cost, its widespread availability, and its capability to diagnose Vascular Hamartomas. On the other hand, there multiple opinions amongst the scientific community who emphasize the necessity to use more complex diagnostic methods, such as an MRI [18].

From a microscopic standpoint, the definitive diagnosis is evinced via means of histopathology. Noteworthy histological findings consist of stable proliferative cells, slow or rapid cellular turnover, normal or thin basement membrane, ordinary morphology of mast cells, and a leveled endothelium [11].

There are multiple proposed therapeutic modalities that have been attempted so far. Those include cryotherapy, injection of sclerosing substances, irradiation, and utter surgical resection [19].

To detail those methods even further, treatment of Hamartomas, specifically Vascular Hamartomas include tattooing, attentive observation, and cosmetic reconstruction after surgical resection. Notwithstanding, there are miscellaneous more novel therapeutic tactics offered and documented in the literature. These include systemic steroid administration, arterial embolization with thrombin-gelfoam appliance, and laser-focused therapy [17,20,21].

All that taken into consideration, Vascular Hamartomas with limited

size are best treated via full and utter surgical excision [22].

4. Conclusion

To come across a Vascular Hamartoma in the neck region of an adolescent patient, especially a Middle Eastern female is a tremendously rare occurrence.

It is ponderously scarce to witness such a diagnosis in young female. The resultant misdiagnoses can frequently transpire and hence, yield a delay in treatment and create more complex repercussions for patients. As demonstrated in our review of the published literature, it is abundantly difficult to find ample information about a case with a pathology like our patient's. Furthermore, the incidence and prevalence rates of Vascular Hamartoma aren't even known. This highlights the need to further study this disease and document it so that we can build future relevant epidemiological studies. This allows us to set-up research protocols and studies to provide surgeons with the necessary information to choose the suitable techniques and establish screening tools to timely manage such a rare vascular abnormality.

Ethics approval and consent to participate

Institutional review board approval is not required for deidentified single case reports or histories based on institutional policies.

Consent of patient

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Availability of data and materials

The datasets generated during and/or analyzed during the current study are not publicly available because the Data were obtained from the hospital computer-based in-house system. Data are available from the corresponding author upon reasonable request.

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Omar Al Laham.

CRediT authorship contribution statement

OA: Conceptualization, resources, who wrote, original drafted, edited, visualized, validated, and literature reviewed the manuscript.

JS, AA, JA: Supervision, project administration, resources, literature review, and review of the manuscript.

IJ: 1st surgical assistant in the operations performed, supervision, project administration, validation, resources, and review of the manuscript.

AA: General Surgery consultant who performed and supervised the surgical procedure, in addition to supervision, project administration, and review of the manuscript.

OA: The corresponding author who submitted the paper for publication.

All authors read and approved the final manuscript.

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Declaration of competing interest

The authors declare that they have no competing interests.

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