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(RESEARCH ARTICLE)

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Cystic lymphangioma of the neck in adults: A report of four cases

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Abstract

Background: Cystic lymphangioma is a rare malformation characterized by a cystic structure deriving from detachment of lymph sacs from venous drainage systems. It is the result of the dilatation of capillary and sinusoidal lymphatic vessels still connected to the lymphatic network.

Case presentation: In the following article, we describe the clinical and pathological features of cystic lymphangioma of the neck in four adults who underwent a surgical removal of the tumor with a well postoperative progress.

Discussion and conclusion: Contrarily to children, the diagnosis of cystic lymphangioma in adults is exceptional. Patients with cystic lymphangioma of the neck are often asymptomatic; However, the lesion growth may cause respiratory disorders, dysphagia, or vascular and neurological compression syndromes. There are multiple modalities of treatment however the surgical complete removal remain the treatment of choice.

Keywords: Cystic lymphangioma; Neck mass; Adult; Neck surgery

1. Introduction

Cystic lymphangioma is a rare malformation infrequently seen in adults. The etiology of cystic hygromas in adults is controversial, but they are thought to be the result of lymphoid vessels proliferation, in response to head and neck trauma and/or infection. We report in this study three cases of cystic lymphangioma of neck who underwent a surgical removal of the mass.

2. Case presentation

2.1. Case presentation 1

We report the case of a 35-year-old female patient with no pathological history, who presented with a slow growing mass on the right side of her neck. The patient did not report any associated symptoms like dysphagia, dyspnea and did not have a recent history of trauma or upper respiratory tract infection.

On medical examination, the swelling was ovoid, measuring 6 cm with well-defined borders, soft, fluctuant in nature and noncompressible. Located in the sub clavicular fossa, the lesion was extending superiorly to the middle part of the neck, inferiorly to 1 cm upper to the clavicle and laterally to 2 cm to the sternum [Figure 1].

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The CT scan performed detected a big, regular-surfaced, cystic formation with a fluid content and an unmodified shape after intravenous contrast injection.

This formation was meeting the jugulo-carotid vessels medially and anteriorly, the paravertebral region medially and internally and the sternocleidomastoid muscle laterally. However, the lateral cervical lymph nodes were normal. [Figure 1].

The mass was surgically excised, and the specimen was a soft and large cystic sac (6 cm × 3 cm in diameter) with external pebbly surface. [Figure 1].

The postoperative course was uneventful, and the patient was discharged on the 5th postoperative day in good general condition.

At the date, the patient is still without signs of recurrence two years after surgery.

2.2. Case presentation 2

A 53-year-old woman with diabetes on metformin, presented to the Surgery department with a slowly growing mass on the left side of her neck that had been evolving for a year. She reported mild pain when moving her head and left arm, with no other symptoms such as dysphagia, dyspnea, torticollis, or monoplegia.

Physical examination revealed a single, cystic, fluctuant, mobile and bilobed swelling measuring 12 × 8 cm in the left part of the neck extending from region I to the thoracic inlet. [Figure 2].

The CT scan performed detected a big, regular-surfaced, cystic formation with a fluid content and an unmodified shape after intravenous contrast injection.

This formation extended from the region I of the neck to the thoracic cavity deep behind the clavicle, coming into contact with the jugulo-carotid vessels medially and anteriorly, the paravertebral region medially and internally and the sternocleidomastoid muscle laterally. [Figure 2].

Radical extirpation was indicated, and the polycystic formation was removed. [Figure 2].

The postoperative course was uneventful, and the patient was discharged on the 10th postoperative day in good general condition.

At the date, the patient is still without signs of recurrence two years after surgery.

2.3. Case presentation 3

B.A is an 18-year-old young woman with no pathological history. She consulted at the Surgery department for a cystic swelling on the floor of the oral cavity extending gradually to the right submandibular region over 2 years.

The medical examination located a diffuse swelling on the right submandibular region of size approximately 4 cm ×3 cm, without significant change in surface texture or color. Intraoral hard tissue examination revealed an upward expansion of the mucous membrane of the floor of the oral cavity due to the pressure of the mass, with no reduction of tongue movements nor alteration of sensory or motor functions.

Based on the history and clinical features, a diagnosis of lymphangioma was firstly supposed. [Figure 3].

An MRI confirmed the cystic nature of the mass located at the floor of the oral cavity and extended below the right mandibular angle, measuring 71 × 40 in diameter. It was well limited, with a thin wall and unmodified after contrast. [Figure 3].

The patient underwent a right cervicotomy which allowing a complete removal of the mass.

The patient progressed well postoperatively. Regular monitoring did not reveal any recurrence over a one-year follow-up.

2.4. Case presentation 4

We report the case of a 25-year-old female patient with no pathological history, consulting for a slow growing mass on the left side of her neck. No associated symptoms as dysphagia, dyspnea or recent history of trauma or upper respiratory tract infection were reported.

Physical examination revealed an ovoid swelling, measuring 5 cm with well-defined borders, soft, fluctuant in nature and noncompressible. Located in the sub clavicular fossa, the lesion was extending superiorly to the posterior part of the neck. [Figure 4].

The CT scan performed revealed a regular cystic formation with a fluid content and an unmodified shape after intravenous contrast injection measuring 6 ×6 × 6,3 cm.

This formation was insinuated between the sterno-cleido-mastoid muscle and the scalene muscle. the mass represses with the internal jugular vein. However, the lateral cervical lymph nodes were normal.

The MRI confirmed the cystic nature of the mass located at the sub-clavicular fossa and extending posteriorly. it is a well limited mass unmodified after contrast and measuring 71 × 75 × 63 mm in diameter. [Figure 4].

The mass was surgically excised, and the specimen was a soft and large cystic sac (8,5 cm × 6 cm in diameter) with external pebbly surface.

The postoperative course was uneventful, and the patient was discharged on the 5th postoperative day in good general condition.

At the date, the patient is still without signs of recurrence one years after surgery.

3. Discussion

Lymphangiomas are classified as lymphangioma simplex, cavernous lymphangioma and cystic hygroma.[1] Cystic lymphangioma corresponds ordinarily to a congenital lesion ; it is a benign proliferation of lymphoid tissue that fails to communicate normally with lymphatic channel leading to hamartomas .[2] The most common sites are in the posterior triangle of the neck (75%), axilla (20%), mediastinum (5%), groin, retroperitoneal space and pelvis. It occurs frequently in infancy 80% of cases within the first two years but seldom presents *de novo in* adulthood.[3] In adults, lymphangiomas may occur either spontaneously or in response to infection or trauma.

Lesion on radiographic feature may occupy one or more of deep spaces of the neck, in between normal structures.

On ultrasound the mass is a multilocular with internal septs of varying thickness and cystic contents, anechoic or hyperechoic if debris, Hight lipid concentration, hemorrhage or infection. [4]

On CT scan we find a homogeneous and cystic formation with rarely intrinsic septs without displacement or compression of adjacent tissues. [4]

On MRI a cystic image with variable signal in T1 sequence and Hight signal in T2 sequence. [4]

They are classified as macrocytic under the level of mylohyoid muscle, located mainly within anterior or posterior cervical triangle, microcystic above mylohyoid muscle in the oral cavity , the tong , submandibular fossa , or parotid region , and mixte . [5]

Majority of cervical lymphangiomas in adulthood are asymptomatic, and present as a painless mass that enlarges progressively. The lesion is soft, nontender, and transilluminant. [6, 7]

Fine needle aspiration cytology may be a good diagnostic tool to confirm the cystic lymphangioma. It shows small and round lymphocytes with intermingling histiocytes without mitoses or atypical cells. Histopathologically, endothelial-lined lymphatic spaces are seen with intervening fibrous tissue and lymphoid aggregates. [6]; In our cases; the CT scan and MRI helped establish the diagnosis without resort to cytology.

The primary treatment modality is surgery, however the use of other treatment modalities has been advocated these include: sclerotherapy, radiation therapy, cryotherapy, electrocautery, sclerotherapy, steroid administration, embolization, and ligation, laser surgery with Nd-YAG, CO2 12 and radiofrequency tissue ablation technique. Surgical excision is the treatment of choice and there is a 15% recurrence rate if the lesion is not fully excised. A two-year postsurgical follow-up is recommended to detect recurrence. [8 ;9]

The indications for surgical treatment are the functional limitations, intolerable symptoms, and altered aesthetics. A balance between the surgery and the risk of potential morbidity from operating are to be discussed case by case.

Surgical difficulties remain important, it is due to proximity of vital structures and poorly demarcated margin of the lesion. [10]

Total excision is important for prognosis and provides tumor recurrence, however it is seldom possible because of the proximity to normal structures that must be preserved. Surgical complications are cranial nerve or vascular injuries, infection, or hemorrhage.

In addition to ablative surgery, microcystic disease involving the tongue can be treated with radiofrequency ablation, laser, and cautery to address lesions on the surface. [11]

Complex cystic hygroma recures in 10 to 27% of cases even after apparent complete excision and 50 to 100% in partial excision. [12]

Sclerotherapy has emerged in recent years as an acceptable alternative to surgery in select lesions. It is noted to be most beneficial in patients who have macrocystis lymphatic disease, but microcystic lesions may also derive benefit from therapy [13]. Under ultrasound guidance, the cysts are aspirated and then injected with a sclerosant such as hypertonic saline, sodium tetradecyl sulfate, absolute ethanol, bleomycin, or Streptococcus pyogenes–derived OK-432. Admission to hospital for overnight observation is recommended if there is a risk of airway swelling secondary to the manipulation of the lesion. [11]

The most common adverse effect of sclerotherapy includes fever, bleeding, and neuropathy, prolonged swelling requiring intubation or tracheostomy.

Electrocoagulation requires the use of an electrode mounted on a hand piece that emits low-power, high-frequency, high-voltage electrical that leads to protein denaturation. One report demonstrates multiple treatments were undertaken resulting in a smoother appearance of a cutaneous lymphangioma but did not eradicate it.[14]

Embolization has been reported in the treatment of cystic lymphatic malformation via arterial embolization, although challenges lie with the multiple feeding vessels and narrow arteries in the neck leading to technical difficulty in cannulation of the vessel. [15]

The mechanism of radiotherapy in treatment of lymphangioma is thought to induce lymphatic endothelial proliferation and oedema that leads to obstruction of dilated vessels [16], Within the head and neck, in rare situations, low-dose radiation treatment with 30 Gy has been reported to be given in one report of a residual cystic lymphangioma with no recurrence at 3 years follow up. [17]

Cryotherapy is a method of deliberate tissue destruction by the application of extreme cold, it can use liquid nitrogen spray directly onto the lesion or probes and nitrous oxide.

Complete resection was possible in all four cases with no evidence of recurrence, and the diagnosis was confirmed by histopathological examination of the surgical specimen.

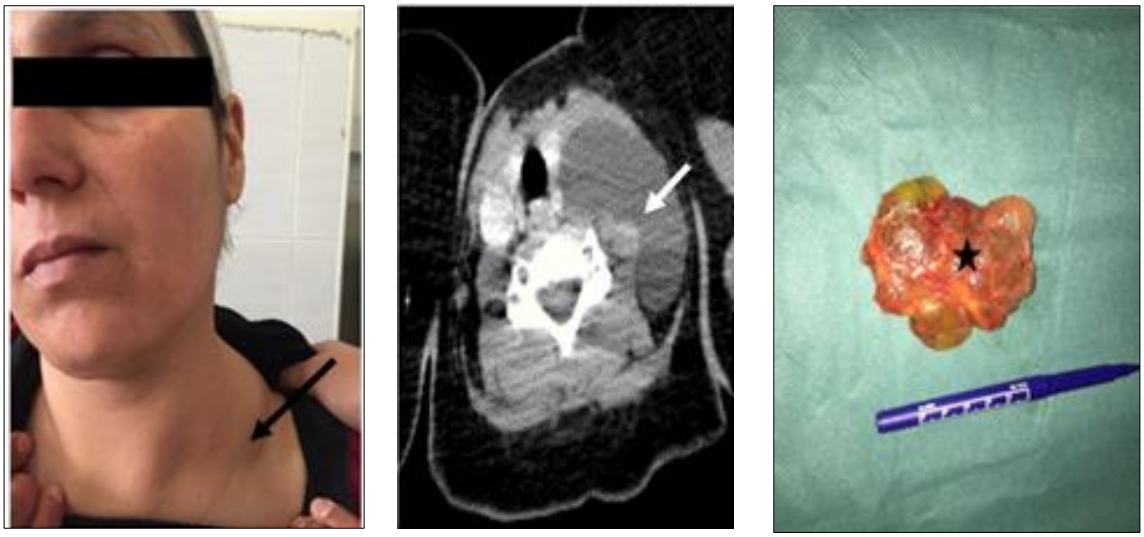

Figure 1 Case 1: 35- year-old female presenting with a left swelling on the posterior triangle of the neck and the sub clavicular fossa (black arrow), Axial CT scan showing an extended cystic formation in the left part of the neck (white arrow) , surgical specimen (★)

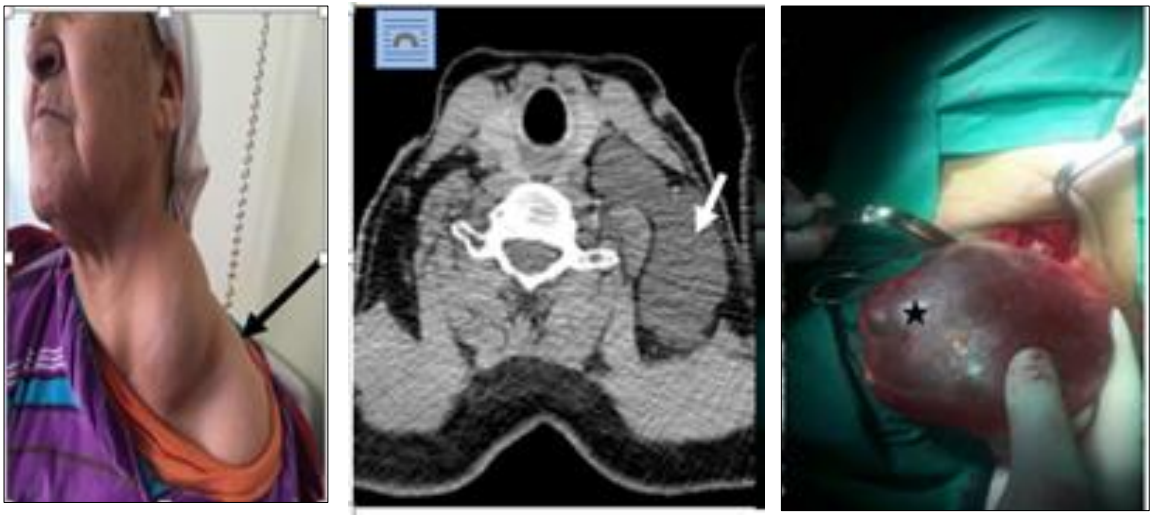

Figure 2 Case 2: 53-year-old female presenting with a left mass on the posterior triangle of the neck extending to the thoracic cavity (black arrow), Axial CT scan showing an extended cystic formation in the left part of the neck (white arrow) ,removed mass (intra-operative) (★)

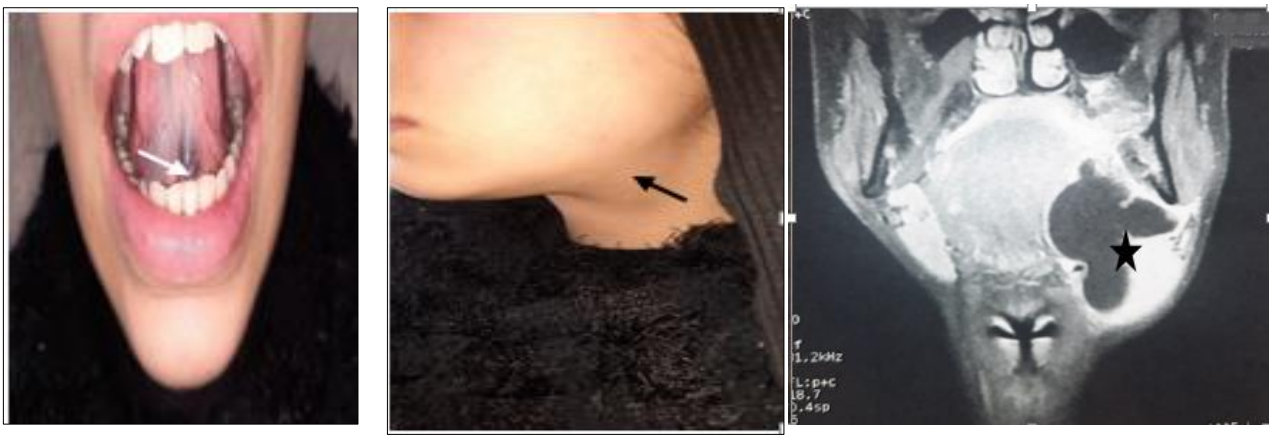

Figure 3 Case 3 18-year-old female presenting with a left swelling on the submandibular fossa (black arrow) extending to the floor of oral cavity (white arrow), Coronal MRI showing the extended cystic formation from the neck to the oral cavity floor (★)

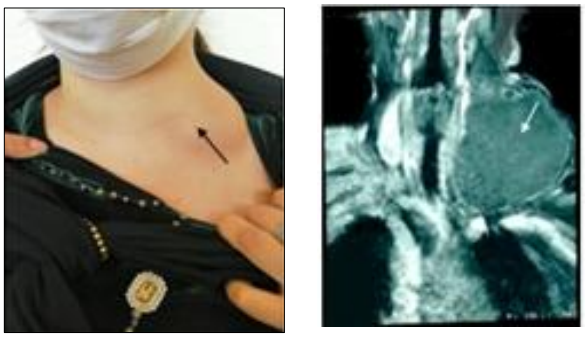

Figure 4 *Case 4: 25- year-old female presenting with a left swelling on the sub clavicular fossa (black arrow) , MRI showing an extended cystic formation in the left part of the neck (white arrow)

4. Conclusion

Cystic lymphangioma of the neck occurs frequently in infancy but seldom presents *de novo in* adulthood. The primary treatment is complete surgical excision to avoid recurrences but other treatment modalities, essentially sclerotherapy, are an acceptable alternative to surgery in select lesions.

Compliance with ethical standards

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Disclosure of conflict of interest

The authors declare that they have no competing of interest Statement of ethical approval.

Statement of ethical approval

The present research work does not contain any studies performed on animals/humans subjects by any of the authors

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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