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In Toto Excision of Lymplangioma - A Challenge

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ABSTRACT

Cystic Lymphangiomas are malformations in the lymphatic system that are uncommon and usually presents within the first two years of life. They are extremely rare in adults with a very few reported cases. They are usually located in the head and neck, especially in the posterior triangle of the neck, presenting as a painless fluctuant transilluminent mass. In adult trauma and upper respiratory tract infection has been suggested as a trigger for its onset. It can clinically mimic a variety of other conditions, hence imaging and histopathology is crucial to its diagnosis. The treatment of choice remains complete surgical resection, though other alternative modalities like Sclerotherapy are available. However, due to its location and extent a complete surgical removal is often impossible. We report a case series (4 cases) of cystic hygroma, all above the pediatric age group, its varied presentation; the diagnostic as well as interpretive challenges faced and tackled in its management.

KEYWORDS

Cystic hygroma; Adult nymphangioma; Cystic neck swelling

INTRODUCTION

Cystic swellings in the neck have a wide range of differentials ranging from benign to malignant; common to rare conditions. Cystic hygroma is a rare differential in adults, hence posing a diagnostic as well as a surgical challenge for the clinician [1-3]. It is believed to arise from early sequestration of the embryonic lymphatic channels. Most of such cases are diagnosed before 2 years of age owing to main period of lymphatic growth [3]; However its prevalence in adults is such that, literature, even in the beginning of the twenty first century suggests less than a hundred reported cases [4]. We are reporting a case series of four patients above the pediatric age group, with cystic hygroma of variable extension who had presented to us in a period of 6 months and the challenges in its surgical removal because of its location and extent with due importance in removing the mass in to-do with preservation of vital structures close to its vicinity.

Case report

Case 1

A 32-year-old male presented with right sided neck swelling since 1 year, insidious in onset, progressively increasing in size (Figure 1a). On examination, there was right sided supraclavicular single swelling 10 cm × 10 cm × 8 cm, fluctuant and transilluminant (Figure 1d).

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CT scan was suggestive of a large well defined non enhancing cystic lesion in right posterior cervical space (Figure 1b). Fine needle aspiration cytology (FNAC) report was suggestive of Lymphangioma.

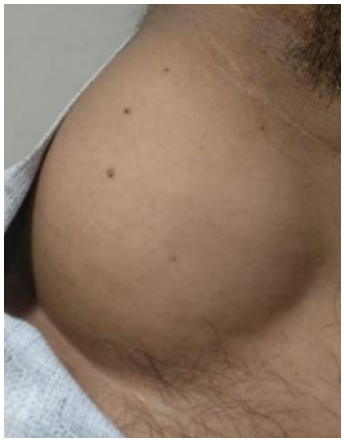

Figure 1(a): Showing right sided neck swelling.

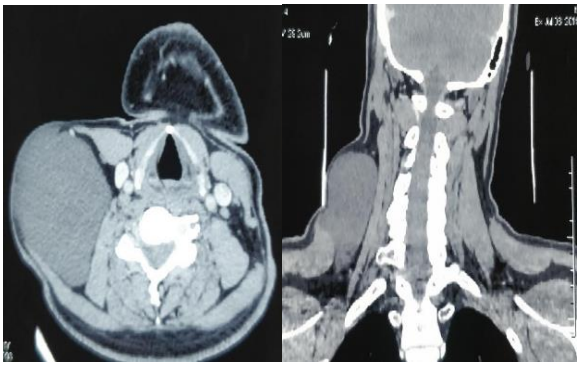

Figure 1(b): CT scan showing a large well defined non enhancing cystic lesion in right posterior cervical space.

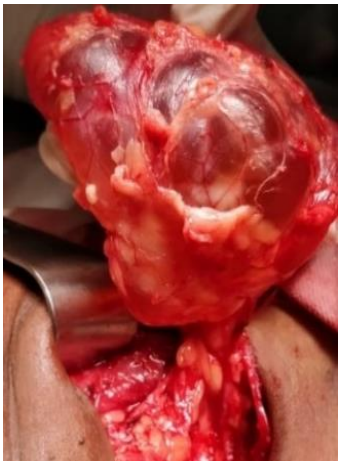

Figure 1(c): Showing intraoperatively Mass extending in the posterior triangle going just below.

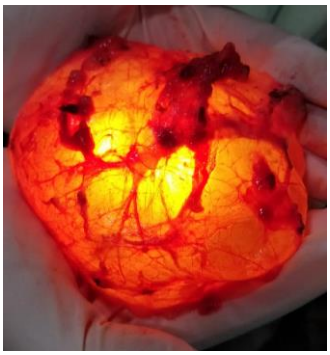

Figure 1(d): Showing right sided supraclavicular single swelling fluctuant and transilluminant.

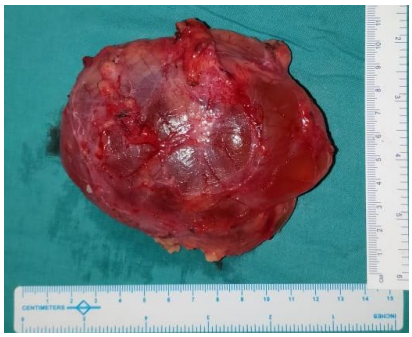

Figure 1(e): Showing dissected and removed in-toto Mass.

Intraoperatively Mass has seen extending in the posterior triangle going just below the clavicle, medially to the carotid and internal jugular vein, laterally till the Trapezius, posteriorly till the constrictor muscles (Figure 1c). Mass dissected and removed in-toto (Figure 1e). Corrugated drain placed till 10 days and removed. Intraoperative and postoperative period was uneventful.

Case 2

The clavicle, medially to the carotid and internal jugular vein, laterally till the Trapezius, posteriorly till the constrictor muscles. A 14-year-old male came with complaints of Right sided anterior neck swelling, insidious in onset, gradually progressing in size, painless, size 8 cm × 12 cm soft in consistency extending from mentum till 1 finger breadth above the clavicle, laterally till the posterior border of sternocleidomastoid muscle medially till midline (Figure 2a), Transilluminant, transmitted pulsations present.

CT scan was suggestive of cystic lesion arising from the lateral wall of nasopharynx extending to a right parapharyngeal area, abutting right Common carotid artery, medially to thyroid suggestive of cystic hygroma (Figure 2b). Fine needle aspiration cytology (FNAC) report was suggestive of a right side cystic lesion.

Surgical excision was done. Mass dissected from all around, separated from the anterior border of the sternocleidomastoid, inferiorly extending just above the clavicle (Figure 2d), superiorly extending uptill pharynx (Figure 2c). Mass excised after separation from the submandibular gland (Figure 2f).

The extension into pharynx was marsupialised and walls sutured to the adjacent structures (Figure 2 (e). Corrugated drain inserted and was removed on postoperative day 14 . Glove drain reinserted on POD16, in view of the collection, Neomycin installed. Symptomatically improved, hence discharged after a week following drain removal. Patient followed up every month postoperatively for five months. He recovered without any morbidity.

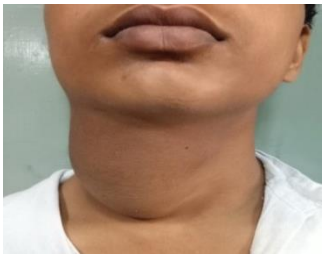

Figure 2(a): Showing Right sided anterior cystic neck swelling.

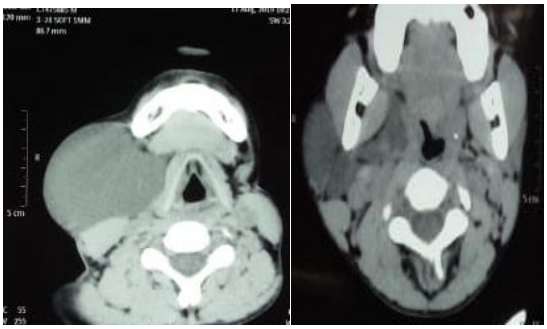

Figure 2(b): CT scan showing of cystic lesion arising from lateral wall of nasopharynx extending to right parapharyngeal area, abutting right common carotid artery, medially to thyroid.

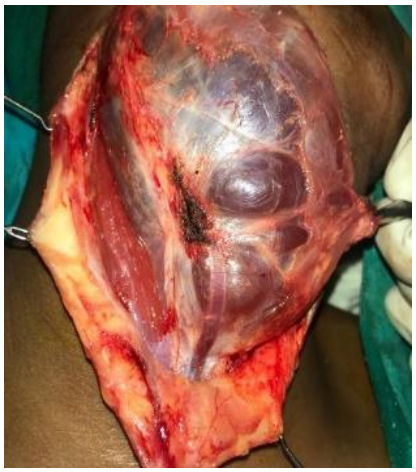

Figure 2(c): Showing Surgical excision of mass extending uptill superiorly pharynx.

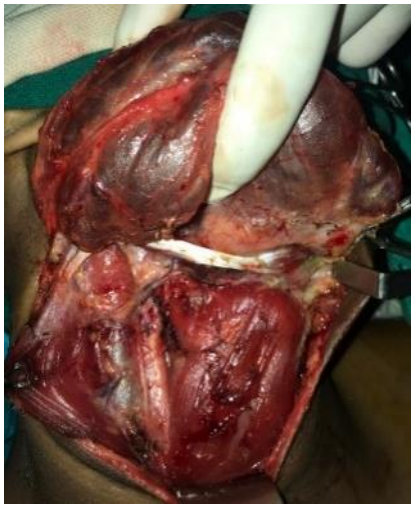

Figure 2(d): Mass dissected from all around, separated from the anterior border of the sternocleidomastoid, inferiorly extending just above the clavicle.

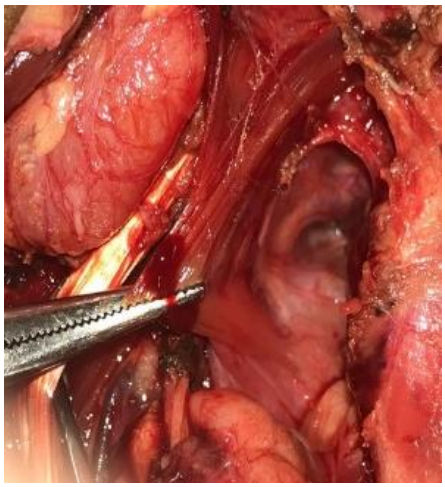

Figure 2(e): Showing extension into pharynx was marsupialised and walls sutured to the adjacent structures.

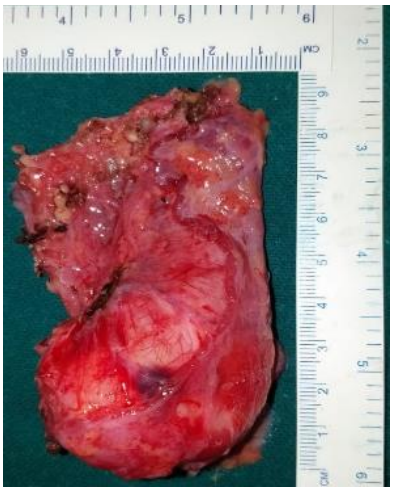

Figure 2(f): Showing Mass excised after separation from the submandibular gland.

Case 3

A 24-year-old male presented with right sided neck swelling since 10 to 12 months, insidious in onset, progressing to chest and axilla (Figure 3a). On examination two separate swelling, one in right supraclavicular region soft, 8 cm × 6 cm, non tender, fluctuant, crossing midline and extending below the clavicle. Another swelling 10 cm × 6 cm soft to firm in consistency, in the right axilla, transilluminant with engorged veins. CT Scan and MRI done, suggestive of well defined thin walled non enhancing cystic lesion 12.4 cm × 6.6 cm × 2.9 cm in right lower posterior cervical space, extending into infraclavicular space and axilla (Figure 3 (b).

The surgical resection was done with the assistance of Cardiothoracic team. Intraoperatively Neck crease incision taken over supraclavicular region, separate vertical incision taken in the axillary region. Large cystic swelling seen extending infraclavicularly towards the right axilla (Figure 3 (c). Mass separated from surrounding structures, including right Internal Jugular vein, carotid artery and subclavian vessels, brachial plexus, axillary vessels. Complete excision of mass done and delivered through supraclavicular incision in toto (Figure 3 (d). Brachial plexus injury identified during supraclavicular dissection through a neck incision. Exploration done by a plastic surgeon and incision extended to join auxiliary and neck incision. Cut ends of the branches from upper trunk identified (branch to suprascapular) and primary epineural repair were done. Post operatively advised shoulder immobilization in the adducted position with elbow physiotherapy for two weeks. Postoperative Histopathology was consistent with cystic hygroma. Patient followed for 6 months postoperatively. There was no recurrence of swelling and no gross restriction of shoulder movements.

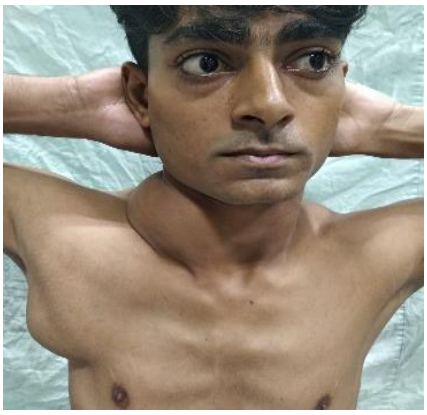

Figure 3(a): Showing right sided neck swelling since progressing to chest and axilla.

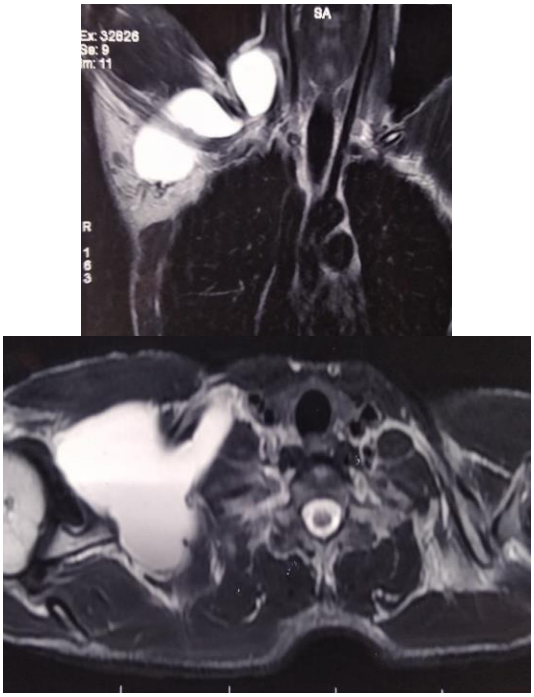

Figure 3(b): CT scan and MRI showing non enhancing cystic lesion in right lower posterior cervical space, extending into infraclavicular space and axilla.

Figure 3(c): Showing intraoperatively Neck crease incision taken over supraclavicular region, separate vertical incision taken in axillary region & large cystic swelling seen extending infraclavicular region.

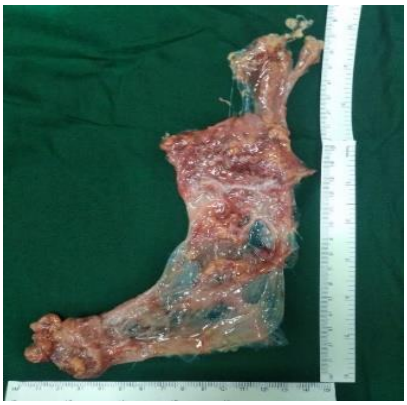

Figure 3(d): Showing Complete excision of mass done and delivered through supraclavicular Incision in toto.

Case 4

A 30-year- old male presented with right sided neck, swelling of 7-8 months duration, insidious in onset, gradually increasing. On examination, there was swelling 7 cm × 5 cm × 3 cm in the right submandibular region, non tender, and fluctuant (Figure 4a).

Fine needle aspiration cytology was suggestive of a benign cystic lesion consistent with lymphangioma, CT scans showed well defined homogenous isodense mildly enhancing soft tissue lesion 6 cm × 2.4 cm × 2.4 cm within the right submandibular region, lateral to submandibular gland abutting and displacing gland posteriorly (Figure 4b), Suggestive of the lymphatic collection/cyst. USG was suggestive of a Collection/cyst measuring 38 mm × 37 mm in the right submandibular region, Lateral to submandibular gland abutting and displacing gland posteriorly (Figure 4b), suggestive of the lymphatic collection/cyst. USG was suggestive of a Collection/cyst measuring 38 mm × 37 mm in the right submandibular region extending to the floor of mouth. Intraoperatively neck incision given 2 finger's breadth below the angle of mandible over the maximum bulge of swelling (Figure 4c), large cystic swelling extending toward the oral cavity (suspecting- plunging ranula, Figure 4d).

Complete surgical excision of neck mass done (Figure 4 (e) with marsupilisation in the floor of mouth. Glove drain placed. Oral Glycopyrrolate given postoperatively. Wound healed well. Histopathology report confirmed lymphangioma. And no recurrence at 6 months postoperative follow up.

Figure 4(a): Showing cystic swelling in the right submandibular region.

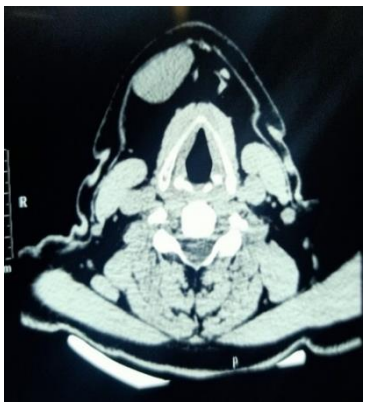

Figure 4(b): CT scan showed well defined homogenous isodense mildly enhancing soft tissue lesion within right submandibular region, lateral to submandibular gland abutting and displacing gland posteriorly.

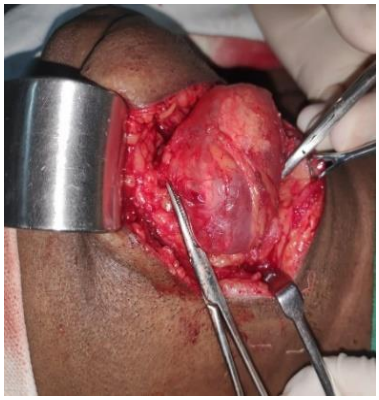

Figure 4(c): Showing Intraoperatively neck incision given 2 finger breadths below angle of Mandible over maximum bulge of swelling.

Figure 4(d): Showing large cystic swelling extending toward oral cavity, suspecting plunging ranula.

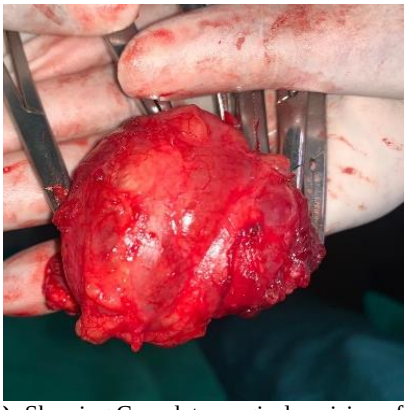

Figure 4(e): Showing Complete surgical excision of neck mass done with marsupilisation in the floor of mouth.

DISCUSSION

There are many differentials for cystic lesions of Neck ranging from bronchial cyst, thyroid cyst, thyroglossal cyst, dermoid or epidermoid cyst, to metastatic squamous cell carcinoma. Lymphangiomas are considered rare and occur mainly in childhood [5]. Because of its very low incidence, most surgeons are unable to gain much personal experience with this intriguing lesion [6]. Lymphangiomas are a benign congenital tumor of lymphatic origin, with cystic spaces lined with true endothelium, classified as microcystic (capillary lymphangiomas), macrocystic (cavernous lymphangiomas) and cystic hygromas [7, 8]. Cystic hygromas were first described by Redenbacker in 1828. The name "Cystic Hygroma" was devised by Werner in 1834 [9-11]. They are composed of cysts and sinuses, containing eosinophilic acellular lymph fluid. The most widely accepted theory about the development of cystic lymphangioma is that they arise from sequestrations of the primitive embryonic lymph sacs [5]. However, the etiology in the adult population is controversial. Some authors attribute adult lymphangioma to delayed proliferation of the congenital or acquired lymphoid rests following trauma or preceding respiratory infection [6], However, there was no history of the same in our patients. These relatively uncommon malformations present as a painless mass cystic to palpation, transilluminant generally involving the head and neck region [12]. As within all our cases. All our cases were males above the age of twelve, although most of the studies suggest no gender predilection.The most common documented site is the neck. Particularly in the posterior triangle [4]. All our four cases involved different regions of head and neck. Two cases involved the posterior triangle. In case 3 the neck lesion extended in the subclavicular plane to the axilla and chest. And in the other two cases it involved the anterior triangle; however case 2 had extension till nasopharynx.

In children, cervical lesions can cause dysphagia and airway obstruction, however, this is rare in adults [13]. There were no obstructive symptoms in any of our patients. Lymphangiomas may remain static or involutes, but in some cases, they may increase in size, especially after internal hemorrhage or infection, can grow rapidly potentially leading to life-threatening airway compromise or obstruction. They also have a tendency to infiltrate into and around muscles, vital nerves and vessels [6].

The diagnosis is usually made on clinical grounds, but CT and MRI scanning will more accurately determine the size, the exact anatomical location, its relationship with important structures and aid the surgical planning [14]. The CT scan was done in all our cases. MRI done in the case which had extended to exile. It is essential to evaluate the intrathoracic extension of cystic hygroma, which might be seen in 10% of cases, and this

is done by USG/CT/Magnetic Resonant Imaging (MRI) [15]. There is no consensus in literature concerning the use of Fine needle aspiration cytology (FNAC) to diagnose these lesions, However, there are a few reported cases where FNAC proved to be an important diagnostic tool [16]. FNAC done in all the cases. Suggestive of lymphangioma except in case 2 which was just reported as a cystic lesion. FNAC together with imaging proved to be beneficial. Despite all the advanced imaging techniques, the diagnosis of adult lymphangiomas remains a challenge. A correct diagnosis is ensured only by histopathological examination of the surgical specimen.

There can also be interpreted surprises. As in one of our cases (case 4) there was sublingual extended of the lesion, hence mimicking a plunging ranula intraoperatively, however later proved histopathologically as lymphangioma. Such rare instances where cases of sublingual lymphangioma mimicking a ranula have also been reported [17].

Though many methods of treatment like aspiration, sclerotherapy have been described over the years. Surgical excision remains the treatment of choice, but it is challenging and it should be, therefore, best undertaken by experienced surgeons in specialist centers. The goal should be complete excision with good cosmesis [9]. Although benign, histology must be undertaken for affirmative diagnosis. In adult patients, this neoplasm can even switch to squamous cell carcinoma [8, 18], Hence the need for its early intervention.

Surgery can be helped by the injection of tissue blue into the lymphatic spaces [19], Though All our cases underwent appropriate surgical excision without the injection of tissue blue. Total tumor removal is the method of choice if it can be achieved. Subtotal removal is defensible in extensive tumor [18]. Partial removal of lymphangiomas is proved unreliable and is considered adequate only for lymphangiomas involving the tongue, pharynx, and larynx. In two of our cases, total removal was not possible, in case 2 and 4 parts extending into the pharynx and to the floor of mouth was marsupialized respectively.

DE series had proposed a staging system which includes laterality ("Uni" or "Bi") in addition to relating to the hyoid ("infra" or "supra") as a reliable way of predicting outcome in patients with lymphatic malformations of the head and neck.

All our 4 cases were unilateral. Two cases which extended to suprahyoid region required marsupialisation [20]. Intraoperatively all the cases had a variable extension and different ease of surgery. One case had an intraoperative complication of nerve injury (suprascapular nerve) which was identified in the table, and repaired by plastic surgeons. The patient had undergone physiotherapy sessions. Complete surgical removal with no recurrence till date along with normal shoulder function was achieved with the help of multidisciplinary teamwork. Postoperatively, all our cases received antibiotics and in case 3 which extended to floor of mouth, oral Glycopyrrolate given postoperatively in order to reduce salivation thus preventing the orocutaneous fistula formation. There are reported cases of wound infections, respiratory infection post excision hence antibiotic prophylaxis is necessary.

CONCLUSION

Lymphangioma being an uncommon pathology in adults needs specific emphasis. Though there is no gender predilection described in literature, all our cases were males. Our case series also helps in concluding that head and neck lymphangiomas may arise spontaneously in adults, even without a significant history of trauma or exertion. Though classically described as being common in the posterior triangle, our cases had varied anatomy ranging from a small mass confined to the posterior or even the anterior neck triangle to a one that can even extend to the pharynx or up till the axilla. Histopathology is crucial to its diagnosis as it can mimic a variety of conditions. Though complete surgical excision is the treatment of choice, the surgical accessibility to its complete removal might not always be possible due to its anatomical location, and it often remains a challenge for the surgeon to remove the mass intoto preserving the vital structures around.

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