Case report:

Rare presentation of cervical lymphangioma in adult

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Abstract:
Lymphangiomas or Cystic hygromas are relatively uncommon congenital malformations of lymphatic system, usually reported in first 5 years of life. Presentation in adult is rare. We report a case of adult cervical lymphangioma and discuss the presentation, diagnosis and management.

Keywords: Lymphangioma, cystic hygroma, lymphatic system

Case report

16-year-old girl came with history of left sided neck mass of 1-month duration, which was not associated with pain, discharge. No H/O fever. No H/O previous head and neck surgery or neck trauma. On examination smooth surfaced, cystic and non-tender swelling of 6 X 3 cm was present in left posterior triangle of neck. Ultrasound examination revealed cystic lesion measuring 6.5 X 3.6 cm seen in subcutaneous plane, lateral to sternocleidomastoid muscle that showed thin septae within it. No cervical lymphadenopathy. Both lobes of thyroid were normal in size and echo texture. Aspiration cytology demonstrated lymphoid cells, but no malignant cells. Posted for excision under general anesthesia.

Intraoperatively revealed cystic lesion of size 10 X 6 cm extending beneath the sternocleidomastoid and was adherent to the underlying nerves. Hence complete resection was not possible. Cyst is partially excised leaving a small-adhered cyst wall. The remnant cyst is obliterated with absorbable sutures. Postoperative period was uneventful. Histopathological examination confirmed it as lymphangioma. No recurrence till date.

Discussion

Lymphangiomas are malformation of lymphatic system that result from abnormal development of lymphatic vessels and are thought to originate from sequestered lymphatic sacs that do not communicate with peripheral draining channels. Majority appears in head and neck region in that 75% located in neck usually located in posterior triangle, with up to 10% extends down into mediastinum. Other common sites include axilla, chest wall, shoulder, abdominal wall, retro peritoneum, pelvis and thigh. This lesion occurs in regions with a predominance of fascial planes and lax tissue such as neck where it can expand and create large cystic spaces. Depending on cyst size these may bmacro and micro cystic or mixed. Macro cystic lymphangiomas will be seen mostly in infrahyoid region and microcystic ones, in suprahyoid region.

In congenital cases, the obvious sign is mass, which can be present at birth or noticed later with in first 5 years of life. Patient may be completely asymptomatic or may present with respiratory obstruction, dysphagia, feeding problems, associated infection, abscess, and sepsis. In adults diagnosis can be more
difficult to determine and depends on a high index of suspicion. The differential diagnoses include branchial cyst, thyroglossal cyst, dermoid cyst, lipoma, occult thyroid, and haemangioma. To date there have been fewer than 150 case reports of adult cervicofacial lymphangiomas in literature and optimum management is yet to be cleared. In a large retrospective study by Kennedy et al in 46 patients with cystic lymphangioma found 4 possible cases with acquired causes. Such causes can include trauma, infection, neoplasia or they can be idiopathic or iatrogenic. In another 2 adult acquired case series of cervical lymphangioma reports in literature, an asymptomatic mass or an unexpected intraoperative finding was the most common presentation of cervical lymphangioma. Although cervical lymphangioma tends to enlarge progressively over months, relatively rapid increase in size has also been described as was seen in our patient. In another case series Thirty-two patients with cervical lymphangioma were treated at the Mayo Clinic. In that study also, rapid enlargement over a short period of time has frequently been reported. In adults sudden appearance is mostly related to history of trauma or acute infectious disease in upper respiratory tract. But in our case there was no history of trauma and upper respiratory tract infection, we may consider the etiology as idiopathic (unknown).

CT or MRI aid in diagnosis and define the extent of the lesion and the surrounding vital structures. MRI in particular gives a clear definition of structures, which is useful in planning the surgery. But due to lack of MRI in our hospital, it was not done. FNAC is essential to confirm and to rule out any other pathology including metastatic thyroid malignancies. Most acceptable management approach to cervical lymphangioma is complete surgical excision. In some cases spontaneous regression was reported. Surgical success can be improved by preoperative scanning. In 19% to 33% of cases postoperative complications (infections, fistula, anti aesthetic scars etc.) were observed. A high recurrence of about 50% was seen in patients who underwent incomplete excision due to adhesions. Such recurrences generally take place within 1 year and they often associated with upper respiratory tract infections. Therefore, a 2-year post-operative follow up is advised.

Other treatment modalities are aspiration, radiation, intralesional injection of sclerotic agents (fibrin-sealants, triamcinolone, bleomycin, hydrocolloid dental impression material and OK-432. Most studies of aspiration and sclerosing agent used OK432 (Picibanil) as sclerosant. OK432 (Picibanil) is derived from low virulence strain of streptococcus pyogenes of human origin. It has been used successfully in macro cystic lymphangiomas and inpatients who has high risk from anesthesia. The pathway of action in cystic hygroma was probably cellular and cytokine mediated. Toxic neutrophilia and giant macrophages were seen probably due to inflammatory reaction due to OK-432. IL6 levels in Lymphangioma fluid remained high in post OK-432 injection. Elevated cytokines increase the permeability of vascular endothelium with in cystic hygroma resulting in increased migration of lymphocytes. Helper T cells, Killer T- cells, macrophages and neutrophils activated by OK-432 induce a localized inflammatory reaction, which increases the permeability of endothelial
cells lining the dilated lymphatics, which leads to involution of cystic hygroma.  

**Conclusion**  
In conclusion cervical lymphangioma is rare in adults. High index of suspicion is needed for diagnosis. Hence cervical lymphangioma should be in differential diagnosis of neck masses in adults. Of the various surgical and non-surgical methods described complete surgical excision is to be the treatment of choice wherever possible.

**References**  
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