CERVICAL LYMPHANGIOMA IN ADULTS- A RARE ENTITY IN ADULT NECK SWELLINGS

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HOW TO CITE THIS ARTICLE: Daniel R, Rajkumar AB, Sadasivam P, et al. Cervical lymphangioma in adults- a rare entity in adult neck swellings. J. Evolution Med. Dent. Sci. 2018;7(27):3144-3147, DOI: 10.14260/jemds/2018/707

PRESENTATION OF CASE

Cystic hygromas/lymphangiomas are extremely rare malformations in adults. They are usually seen in infants and children under 2 years of age. En bloc resection is difficult due to the adhesive characteristics of the tumours. Inadequate surgical intervention often leads to recurrent disease. We report herein the case of a cystic hygroma/lymphangioma that presented as an uncommon mass on the cervical region in an adult.

A 24-year-old male presented with a swelling on the left side of the neck since childhood, noticed to have increased in size for the past six months. It was a solitary 4 cms x 3 cms, soft, non-tender, ovoid swelling at the left submandibular region with a well-defined margin with smooth surface and the overlying skin was normal. There was no intra-oral extension on bimanual palpation. The routine blood investigations are done and were within normal limits. FNAC and CT neck were done. Patient underwent wide excision, where the intraoperative findings showed a superficial cystic swelling with few loculi over the muscle layer of the mylohyoid and the anterior belly of the digastric with few finger-like extensions between the muscle planes. The swelling was excised in toto. Histopathology reports revealed multilocular cystic swelling lined by fibrous stroma and on microscopy was multiple lymph spaces lined by lymphatic epithelium with a diagnosis of lymphangioma of the neck.

DISCUSSION OF MANAGEMENT

Cystic hygromas/lymphangiomas are rare congenital malformations of the lymphatic system. Most are seen in the head and neck region (75 - 80%) and usually affect children under 2 years of age.¹ They are quite rare in adults.² The aetiology of hygromas in adults is controversial, but they are thought to be due to proliferation of lymphoid vessels in response to head and neck trauma and/or infection.³

'Financial or Other Competing Interest': None.

Submission 23-05-2018, Peer Review 17-06-2018,

Acceptance 22-06-2018, Published 02-07-2018.

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DOI: 10.14260/jemds/2018/707

Lymphatic tissue can also improperly anastomose with capillaries, veins and arteries.⁴

Microscopically, lymphangioma is characterised by large, dilated lymphatic vessels in a fibrotic or loose stromal background. Focal areas of papillary endothelial proliferation were described aside by the classic histological picture.⁵ The presence of lymphoid aggregates, even lymphoid follicles may be confusing when existing in large amounts and needs to be distinguished from atypical lymphoid proliferations. The main histopathologic differential diagnosis of cystic hygroma is cavernous haemangioma, in which blood filled large cystic spaces were described to be similar to lymphangioma. The lining endothelium of cavernous haemangioma shows positivity with pancytokeratin and factor VIII immunohistochemically. The presence of lymphatic spaces with thin walls containing fibrous tissue, smooth muscle and lymphoid aggregates favours the diagnosis of lymphangioma.⁶ In general, lymphangioma is accepted as a benign tumour with no malignant transformation and curable by excision.⁷

Lymphangiomas can manifest anywhere in the body, but are most commonly seen in cervicofacial regions (Especially in posterior cervical triangle), axilla, mediastinum, groin and oral cavity.⁸⁻¹⁰ More than 95% of lymphangiomas are situated in the head, neck, axilla and extremities.⁸⁻¹⁰ Although, common in childhood, occurrence in adults is quite uncommon and fewer than 150 reports of adult cervicofacial cystic hygromas have been reported in the literature.^{11,12} It presents a diagnostic challenge in adults as compared to children and definitive diagnosis is usually based on post-operative histology.¹¹ Here, we present the case of a 38-year-old male patient with cystic hygroma in the cervical region.

A 24-year-old male presented with a swelling on the left side of the neck since childhood, noticed to have increased in size for the past six months and there was an increase in size on eating. Patient had no pain over swelling, shortness of breath, dysphagia, dysphonia and no other swelling in body. There was no history of previous surgeries.

CLINICAL DIAGNOSIS

On clinical examination, a solitary 4 cms x 3 cms soft, non-tender ovoid swelling was seen at the left submandibular region. The swelling had a well-defined margin with smooth surface and the overlying skin was normal (Figure 1). There was no intra-oral extension on bimanual palpation.

Figure 1. Neck Swelling in the Patient

The routine blood investigations were normal, and the CT-scan revealed a cystic swelling in the left submandibular region with a suggestive diagnosis of a retention cyst in the submandibular gland (Figure 2). FNAC done later only showed serosanguineous fluid.

Figure 2. The CT Scan Pictures (Transverse and Sagittal Section) revealing the Tumour with the extent marked by White Double-Sided Arrows

Figure 3. Cystic Lymphangioma

Figure 4

After taking informed written consent, patient had been taken for surgery. A wide excision was made under general anaesthesia. A transverse incision is made over the swelling parallel to the mandible, where the intraoperative findings was that of superficial cystic swelling with few loculi over the muscle layer of the mylohyoid and the anterior belly of the digastric with few finger-like extensions between the muscle planes. Cystic swelling was circumferentially dissected and removed in toto (Figure 4).

Figure 4: Intraoperative Pictures (Short arrow on right pointing to dissected portion of the posterior belly of left digastric muscle. Long arrow on left pointing to the cystic tumour lymphangioma).

Figure 5

PATHOLOGICAL DISCUSSION

Care has been taken during dissection to prevent injury to vital structure and the histopathology report gross was a multilocular cystic swelling lined by fibrous stroma and microscopy was multiple lymph spaces lined by lymphatic epithelium with a diagnosis of lymphangioma of the neck.

Figure 5: Histopathological findings of lymphangioma (Long arrow- Cystic areas with lymphoid aggregates; Short arrow- Blood vessels).

Figure 6

DISCUSSION OF MANAGEMENT

Cystic hygromas/lymphangiomas are thought to be developmental abnormalities associated with a failure in the embryological connection between lymphoid vessels and venous system and generally not accepted as true tumours.¹³

Three types of cystic hygroma/lymphangioma can be distinguished. The capillary form is usually asymptomatic with small sizes. Although, cavernous and cystic lymphangiomas show the same histological pattern, cystic lesions are usually larger and symptomatic.¹⁴ Although, more than 90% of lymphatic malformations are clearly congenital, later presentations were due to trauma, infection, neoplasms or iatrogenic injuries.¹⁵ The pathophysiology of adult lymphangiomas is not clearly understood, but these lesions may occur secondary to induction of dormant rests of embryonic lymphatic tissue that are stimulated to differentiate and grow.

Robert P¹⁶ reported 32 cases of cervical lymphangioma treated at Mayo Clinic between 1950 and 1982. He reported that lesions were seen in all decades of adult life and equally in men and women. Most lesions presented as a rapidly enlarging asymptomatic mass. The anterior triangle of the neck was involved nearly as often as the posterior triangle and right-sided locations predominated (72%). Histopathologic differentiation of the lesions into simplex, cavernous and cystic lymphangiomas was not helpful in predicting clinical behaviour. Surgical excision is the treatment of choice. The lesions may intimately involve the carotid sheath or adjacent nerves. Recurrence in 21% of the 28 patients on whom follow-up data were available mostly represented incomplete excision.¹⁵ Primary treatment for lymphangioma is total surgical excision.¹⁷ The treatment of choice for cystic hygroma is surgical excision without which there is a danger of infection and fistula formation. Saxena et al² described that cystic hygroma might infiltrate into adjacent structures, which can lead to recurrence in 10%-15% of cases if complete excision is not done.

Other treatment modalities have been reported such as marsupialisation, injection of sclerosing agents and steroids, diathermy and radiotherapy in unresectable or persistent cases.^{18,19} Injection of sclerosing agents into the lesion such as OK-432 (a lyophilised mixture of group A streptococcus pyogenes of human origin), 50% dextrose, triamcinolone, bleomycin, fibrin sealant and hydrocolloid impression material has had some success. For recurrent cystic hygroma,

intravenous cyclophosphamide and intracystic injection of OK-432 have been advocated.²⁰

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