An Unusual Case Report of Non-Syndromic Bilateral Branchial Cyst

Nor Azirah Salahuddin[1], Saravana Selvi Sanmugam[2], Azreen Zaira Abu Bakar[2], Siti Asmat Md Arepen[1], Nour Hanan Daniah Mohd Bakhit[1], Ahmad Anwaar bin Muhammad Saifullah[1] and Nor Eyzawiah Hassan[1*]

[1]Otorhinolaryngology Head and Neck Surgery, Surgical-based Department, Faculty of Medicine and Health Sciences, Universiti Sains Islam Malaysia

[2]Department of Otorhinolaryngology, Hospital Kuala Lumpur, Ministry Of Health, Malaysia

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Corresponding author: eyzawiah@usim.edu.my

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Abstract

Branchial cleft cyst is a common cause of soft tissue swelling in the neck of young adults. They generally occur unilaterally at the lateral aspect of the neck. Bilateral branchial cysts are rare and may have familial associations. Fine needle aspiration cytology (FNAC) is a good diagnostic tool for branchial lesion, assisted by concurrent radiological modalities. Surgical excision is the gold standard treatment for branchial anomalies. We report a rare case of non-syndromic bilateral branchial cyst in a 23-year-old lady who presented with chronic bilateral, progressively enlarging painless neck swellings. Complete surgical excision of the bilateral cyst was done and histopathological examination (HPE) confirmed the diagnosis. Bilateral branchial cysts may be familial in nature and even part of a larger clinical syndrome. In patients with branchial cleft anomalies with a complaint of auricular deformity or a similar history and findings in other family members, additional examination should be done to find the possibility of branchio-oto-renal (BOR) syndrome. Precise diagnosis and treatment of branchial cysts is crucial to prevent misdiagnosis which may lead to unnecessary and distressing interventions and management. Delayed diagnosis may be complicated by infection, fistula, and even malignant degeneration. For best outcome, early and complete surgical excision is recommended.

Keywords: Branchial cleft cyst, Branchial arches, Branchial anomalies, Bilateral, Congenital

Introduction

Branchial cysts are commonly unilateral, slow growing swelling at lateral neck resulting from persisting branchial apparatus, with second branchial arch anomalies being the most common.[1,2] It is the second commonest congenital head and neck lesions after thyroglossal duct cysts, and clinically apparent in late childhood or early adulthood, with higher incidence rates in males than females.[3–5] We report a rare case of bilateral branchial cysts in a young lady.

Case presentation

A 23-year-old lady presented with bilateral painless neck swelling for one year which were gradually enlarging. There was no history of pain, fever, skin changes, discharge or swelling elsewhere in the body. Aside from the swelling, there are no other significant symptoms from ear, nose and throat (ENT). She denies any B-symptoms, hyper- or hypothyroid symptoms, or contact with tuberculosis patient. There was no similar problem or malignancy in the

family. She also denied a personal or family history of renal anomalies or hearing loss. She is a non-smoker and teetotaller.

Upon neck examination, there were bilateral soft cystic masses at level II anterior to sternocleidomastoid muscle (SCM). Left sided mass was measuring 4 x 6 cm and the right sided mass was measuring 2 x 3 cm. Both swellings have well-defined margin, smooth surface, non-tender with normal overlying skin (Figure 1). Other ENT assessments including the nasopharnygolaryngoscopy examination were unremarkable.

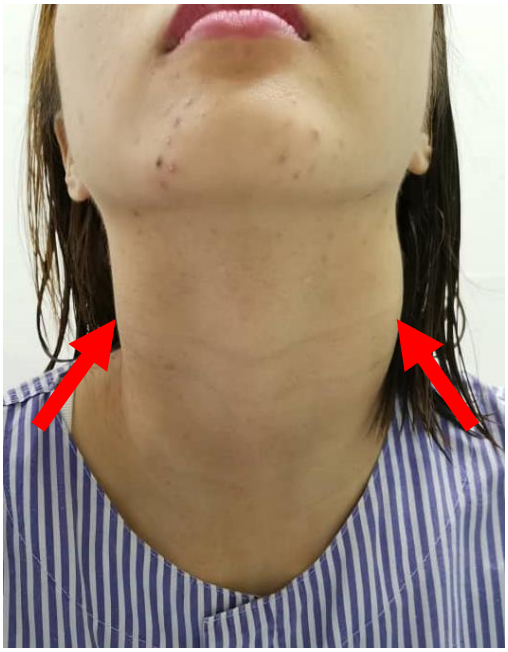

Figure 1: Showed clinical finding of bilateral well defined soft cystic neck swelling at level II-III, anterior to SCM (arrows)

Fine needle aspiration cytology (FNAC) of the mass showed hypocellular smear with foamy macrophages, cholesterol crystals, inflammatory cells and proteinaceous fluid, no epithelial, atypical or malignant cells seen; which was consistent with a cyst. Tuberculosis workups were negative. Neck ultrasonography (USG) revealed bilateral well defined hypoechoic masses with echogenic debris within. Neck computerized tomography (CT) scan showed two well defined cystic lesions with no extension seen medial to sternocleidomastoid muscles and lateral to carotid space; 3.3 x 2.7x 4.8 on the right and 4.0 x 2.2 x 6.8 cm on the left (Figure 2) , which confirmed the ultrasound findings and ruled out features of head and neck malignancy. These results were highly suggestive of bilateral type II branchial cleft cysts. In addition, abdominal and renal USG revealed normal findings, thus excluding the possibility of branchio-oto-renal (BOR) syndrome .

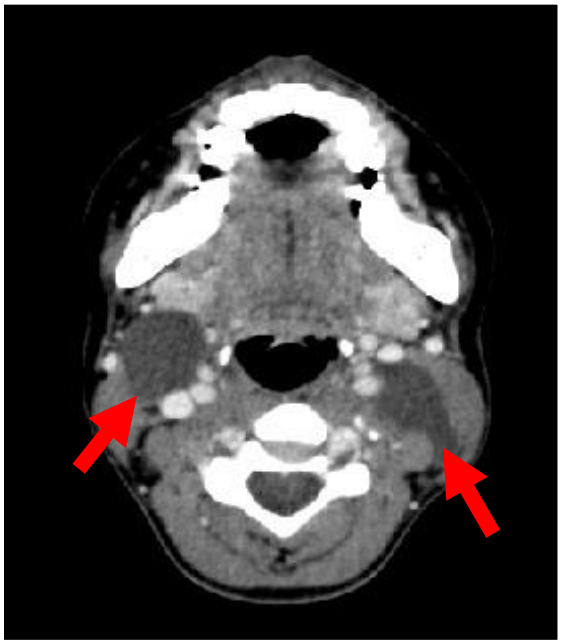

Figure 2: CT scan finding on axial view showed bilateral neck swelling, right swelling is anterior to SCM and the left swelling is deep to SCM (arrows)

Complete bilateral surgical excision of the branchial cysts was done on the same setting. During the procedure, the excision was done ~~for~~ one side at a time. Bilateral skin crease transverse incision were made at lateral upper 1/3rd of the neck. Subplatysmal flap was raised and the SCM retracted laterally to expose the lesion. The right lesion extended superiorly to the right post belly of digastric, laterally to lateral border of SCM, medially to laryngeal framework and inferiorly to level of thyroid cartilage. The cyst was sitting on right IJV, however it was not attached to it. The left lesion extended superiorly to posterior belly of digastric, laterally to 1cm lateral to lateral border of SCM, medially to the carotid space and inferiorly to left inferior belly of omohyoid. We were having some difficulties for the left lesion as it was bigger and located posterior to SCM and closely related to the left carotid artery posteriorly. However we managed to dissect it carefully without any complication or vascular injury. Intraoperatively, the size measuring 3 x 4cm for right side and 7 x 4 cm on left side. It contained straw coloured fluid upon aspiration ~~of the cysts,~~ and no sinus extension to the pharynx. Bilateral drain was inserted to prevention any postoperative collection and were off on day 3 of excision. Postoperatively, she recovered well and HPE showed cyst lined by squamous epithelial cell with stroma contain numerous lymphoid tissues; establishing the diagnosis.

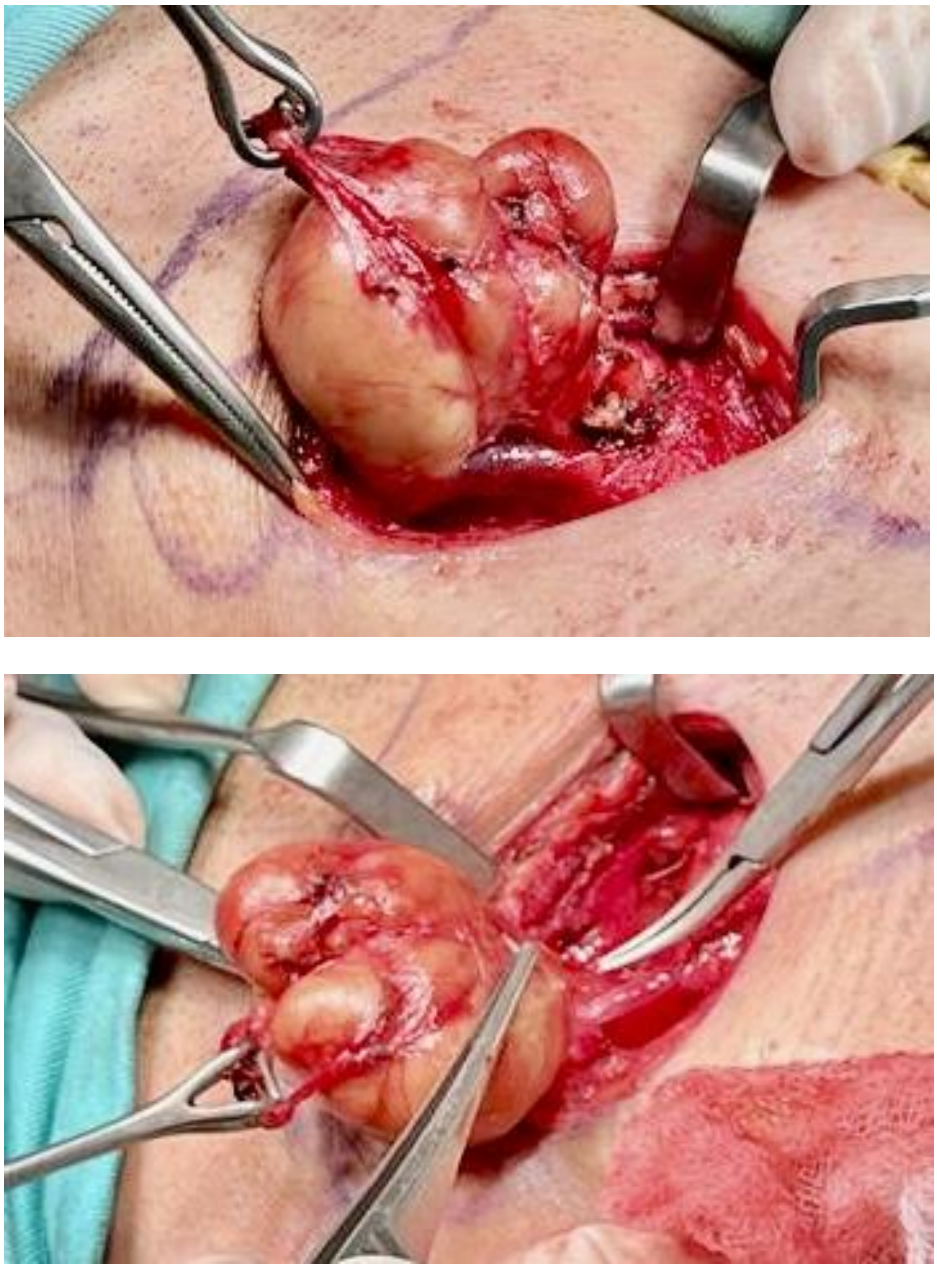

Figure 3.1 and 3.2: Intraoperative finding of right branchial cyst. Yellowish colour lesion with capsule. Surface marking were done at lower border of mandible and along the SCM.

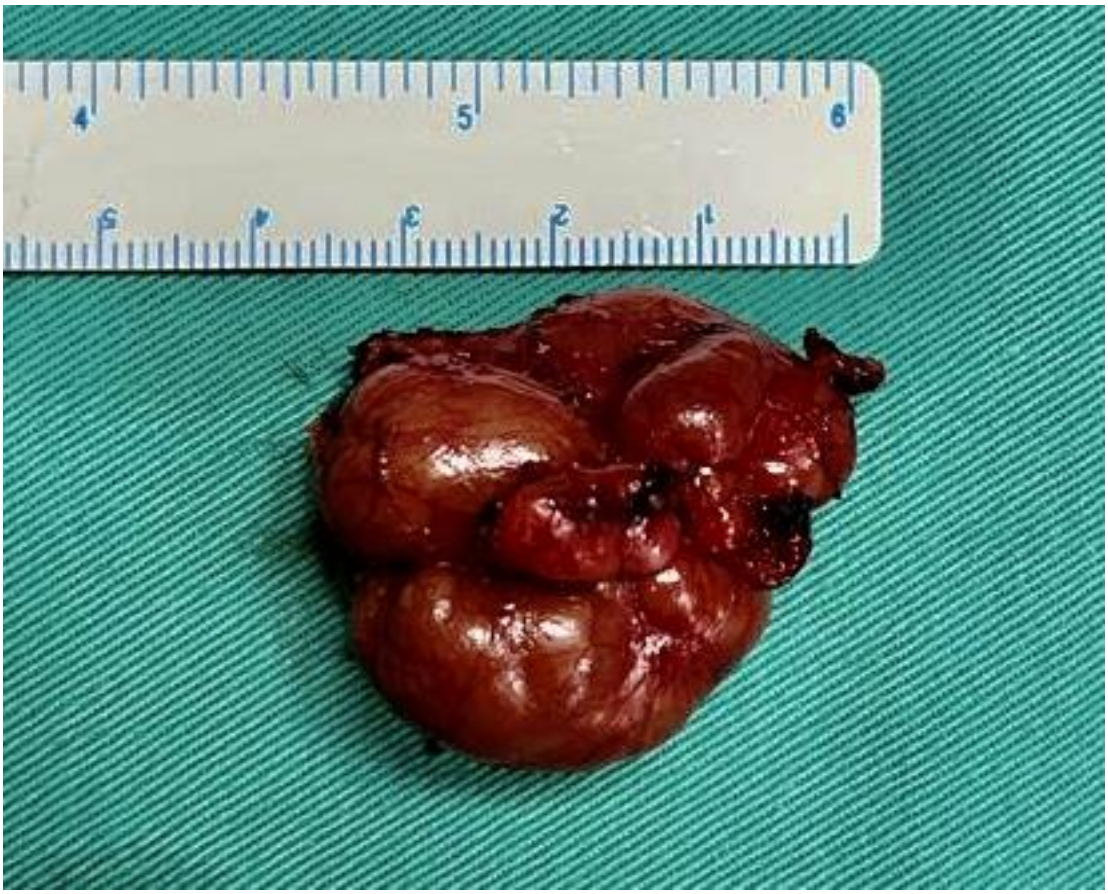

Figure 4: The specimen of right branchial cyst measuring 3x4 cm.

Discussion

Neck masses are common in clinical practice and affecting all age groups. It can be subacute, acute, or chronic, benign or malignant and congenital or acquired in nature. This patient had a chronic bilateral neck swelling. Differential diagnosis should be according to patients' age group and character of the swelling. Differential diagnoses for chronic neck masses include thyroid pathology, branchial cysts, laryngocele, thyroglossal duct cysts and dermoid cysts.[6] Although the patient is still young, malignant causes should be rule out such as lymphoma, papillary thyroid carcinoma and secondary metastasis which is not rare (10.7% (3/28)).[7] For malignant causes, it is important to consider metastatic cystic squamous cell carcinoma to a lymph node from an oropharyngeal HPV-associated squamous cell carcinoma, which may comprise 3% of the cases.[8]

Branchial cyst commonly presents as a single, painless neck mass in a child or young adult, with mostly distributed on left side, followed by right side. Only 2-3% of the cases are bilateral and rarely multiple.[1,2] Although congenital, branchial cyst is a slow growing lesion that may take years to develop and manifest, with variable size and location. Second branchial cyst has been previously categorized into four different sub-types following its anatomical position.[9] This patient has a type II branchial cyst[9] which lies anterior to the SCM, and adjacent and lateral to the carotid sheath.

Bilateral lesion may have familial association with autosomal dominant inheritance.[10] It may also be associated with the autosomal dominant BOR syndrome characterized by branchiogenic anomalies, hearing loss, and congenital anomalies of the kidney and urinary tract.[11] It is confirmed with genetic tests. As branchiogenic anomalies were present in approximately half of BOR patients[12], family history of hearing impairment, renal anomalies or urinary symptoms

should be asked and if present, renal ultrasound, ear examination and genetic tests should be done to detect BOR syndrome.[12] However this patient had no familial history nor other symptoms of BOR.

Patient with branchial cyst typically presented with painless neck swelling. Bilateral branchial cyst may also presented with symptoms similar to solitary branchial cyst such as local compressive symptoms like dysphagia, dysphonia, dyspnea, and stridor.[13] The cysts may become infected and painful following an upper respiratory tract infection, with suppuration, fistula formation, and discharging sinus.[14] Infected branchial cyst may also develop into a life threatening retropharyngeal abscess.[15]

Branchial cyst is diagnosed through history, clinical examination and supported by the diagnostic/radiological modalities including ultrasound (USG), plain and/or contrast enhanced computed tomography (CT) scan, magnetic resonance imaging (MRI) and FNAC.[15] Patients presenting with lateral neck cystic swelling with or without episodes of recurrent neck abscess should raise suspicion for branchial anomalies. The diagnosis of branchial cysts may seem straightforward with high diagnostic accuracy, as demonstrated by previous case series.[4,16] In contrast, other case series by Zaifullah et al have shown a disappointing failure to make an initial correct diagnosis of branchial cyst, which correct clinical diagnosis was made only in 41.6% (5/12) of the cases.[2] Poor diagnostic accuracy was also reported by Daou where only 41.2% (14/34) were correctly diagnosed on presentation.[17] The poor diagnostic accuracy was mainly due to misdiagnoses as neck abscess, neoplastic lymphadenopathy, cold abscess, neoplasms and thyroid pathology.[2]

FNAC is recommended prior to surgical intervention as it is diagnostic and therapeutic, where it relieves the symptoms and temporarily shrinks the cyst. Cytology typically demonstrates benign epithelial cells with cholesterol crystals and lymphoid aggregates.[17] Similarly, our case showed cyst lined by squamous epithelial cell with stroma containing cholesterol crystals and numerous lymphoid tissues, establishing the diagnosis of branchial cyst.

In the primary health care setting, USG may be used for initial imaging to differentiate cystic from solid lesions, measure size, and differentiate high-flow from low-flow vascular malformations. It is non-invasive, rapid, low-cost with no ionizing radiation, although the extent of the lesion may not be determined accurately.[6] CT or MRI scan is needed in a case where the cyst is extensive or in an unusual site in order to delineate its extension and anatomical relation to adjacent structures before definitive surgical excision.[1] Appropriate radiological imaging facilitates complete excision and prevents recurrences.

Surgical excision is best performed in non-infected neck to minimize risks of bleeding and postoperative wound infection.[3.] Although presented as bilateral lesion, the excision can be done simultaneously in the same setting. There was no additional morbidity and it can reduce the number of hospital admission, surgery, hospital visit for follow up as well as medical cost saving for this patient. Re-excision of branchial cysts following previous improper surgery has a high recurrence rate of up to 20%, while complete excision with no prior surgery has a low recurrence rate of 3–4%.[17] Recurrent cases may be complicated by fibrosis from previous surgery which further increases the likelihood of incomplete excision.[18]

There is also a risk of malignant transformation of branchial cyst; primary branchiogenic carcinoma. Even though it is extremely rare, it must be excluded prior to surgical intervention as it entails a different course of treatment.[19] Al-Mufarrej et al. found malignant degeneration in 4 out of 421 cases, or 1%, in older patients (over 45 years old) with branchial anomalies.

Conclusions

Neck mass poses diagnostic challenges if not properly addressed and investigated. Careful and meticulous examinations and investigations are crucial before embarking on any surgical treatment. Numerous differential diagnoses is possible and will depend on the patient's age, the location of the mass, the detail characteristic of the mass as well as the associated symptoms exhibited by the patients. Branchial cysts are congenital neck lesions commonly presented as unilateral neck swelling during early adulthood. The presence of bilateral branchial cysts should raise suspicion of a larger clinical syndrome which should be probed further which includes meticulous personal and family history. Early detection and reaching correct diagnosis are the major factors influencing the

prognosis and avoiding possible complications. Primary health care practitioners play an important role as gatekeepers to detect the possibility of branchial anomalies and arrange for referral to secondary care.

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