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Case Report

Bilateral branchial cleft cysts with primary papillary thyroid carcinoma concealed within left branchial cleft cyst: An unusual presentation

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Abstract

Branchial cleft cysts are lateral cystic neck lesions, usually presenting in 2nd to 4th decade of life, due to incomplete obliteration of the branchial apparatus. Bilateral presentation is an infrequent occurrence. Sometimes they may harbour normal thyroid tissue but rarely present with primary papillary thyroid carcinoma without thyroid gland involvement.

We report a case of 23-year-old female presenting with gradually increasing bilateral painless neck swellings. Radiological examination suggested plunging ranula in bilateral sublingual space abutting each other in the midline with enhancing irregular soft tissue density in the left ranula suggesting papillary neoplasm. The thyroid gland was normal in size, shape and signal intensity. FNAC was suggestive of a bilateral cystic lesion with intracystic papillary neoplasm on the left side. HPE confirmed a bilateral BCC with primary left intracystic papillary carcinoma thyroid with extra cystic tumour deposits.

This case report highlights the importance of clinical, radiological, FNAC, and HPE for correct diagnosis and further management.

Keywords: Lateral neck lesion, Branchial cleft cyst, Papillary carcinoma of the thyroid.

Key Messages: Branchial cleft cysts can harbour malignancies and thorough clinic-pathological-radiological approach is a useful in differentiating it from its close mimics.

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1. Introduction

A branchial cleft cyst (BCC) is a developmental cyst that usually presents later in life. They account for 25% of cysts, originating from embryonic remnants.[1] Bilateral presentation is an infrequent occurrence. Usually, they are asymptomatic but sometimes may present with compressive symptoms due to their large size or when infected.[1] Sometimes they may harbor normal thyroid tissue but rarely present with primary papillary thyroid carcinoma without thyroid gland involvement.[2-5]

Only 8 cases of primary Papillary thyroid carcinoma (PTC) in BCC have been reported in the literature.[2] This is the 9th case which highlights the importance of clinical, cytological, radiological & histopathological examination for correct diagnosis and further management.

2. Case History

A 23-year-old female presented with gradually increasing bilateral painless neck swelling for three years. Physical examination revealed right cystic swelling measuring 3x3 cm in the anterior triangle, Left swelling was cystic in the submandibular extending to midline & downwards anterior to sternocleidomastoid 6x6 cm. There were no compression signs or symptoms. Fine needle aspiration cytology (FNAC) yielded 10 ml of thin fluid from the left swelling and 5 ml from right, and the lesions partially collapsed after aspiration. The cytospin preparation from the left side fluid showed numerous papilliform epithelial cell clusters with peripheral scalloping borders, with a mild degree of anisonucleosis. Occasional cell cluster with vacuolated cytoplasm was also seen. Few metaplastic squamous cells were identified in the background of a large number of foamy macrophages &

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RBCs. Fluid from the right side showed squamoid metaplastic cells and a large number of vacuolated macrophages in a hemorrhagic background. The features were suggestive of the bilateral cystic lesion with intracystic papillary neoplasm left side. (**Figure 1**)

Radiological examination suggested plunging bilateral ranulae abutting each other in midline. Enhancing irregular soft tissue density was noted in the inferior medial left Ranula. Papillary neoplasm is suggested within the ranulas**.** The thyroid gland was normal in morphology and enhancement. Enlarged bilateral lymph nodes? Metastasis. cytohistopathological correlation was suggested. (**Figure 2**) Thyroid function tests were within normal limits.

The patient was operated on after the initial workup. A cyst measuring 6x6cm, on the left side, extending from the floor of the mouth to the suprahyoid region below was removed along with the left adherent submandibular gland. Another cyst was identified in the right side extending midline underneath the anterior belly of the digastric and above till the floor of mouth and laterally extending to the right submandibular gland, was excised.

The specimen received on gross examination showed a salivary gland with an adherent cut-open cyst, fibrofatty tissue bits and hyoid bone. Salivary gland measured 6x4.5x2.2cm, and the cyst measured 5x3 cm. Wall thickness varied from 0.3cm to 0.9 cm. The right-sided cyst was received measuring 2x1.8x0.4cm.

Multiple sections taken from the left submandibular cyst show a thick fibrous wall lined by Low attenuated cuboidal epithelium along with peripheral lymphoid collections and intracystic tumour with follicular & papillary configuration. The cells were cuboidal to columnar with glassy nuclei & infrequent mitosis. The cells were positive for TTF on IHC (Gene Ab Monoclonal Rabbit Anti-body). Coarse psammomatous calcification was also noted. Multiple nodular tumour deposits were seen in the cyst wall as well as the adjacent fibromusculoadipose tissue. No normal ectopic thyroid tissue was seen anywhere in the cyst wall. Section from the adherent salivary gland did not show any malignancy. Lymph nodes did not show any tumour deposits. Sections from the right neck swelling show a cystic structure with a fibrous wall lined by low cuboidal epithelium with focal squamous metaplasia & peripheral lymphoid tissue. There was no evidence of any malignancy. No ectopic thyroid tissue was identified. (**Figure 3**) Findings was suggestive of a bilateral branchial cleft cyst (BCC) with primary left intracystic (PTC) with intracystic tumour deposits. The patient was examined for Branchio-Oto-Renal Syndome and no evidence for the same was found. The patient is on regular follow up and there is no evidence of any recurrence.

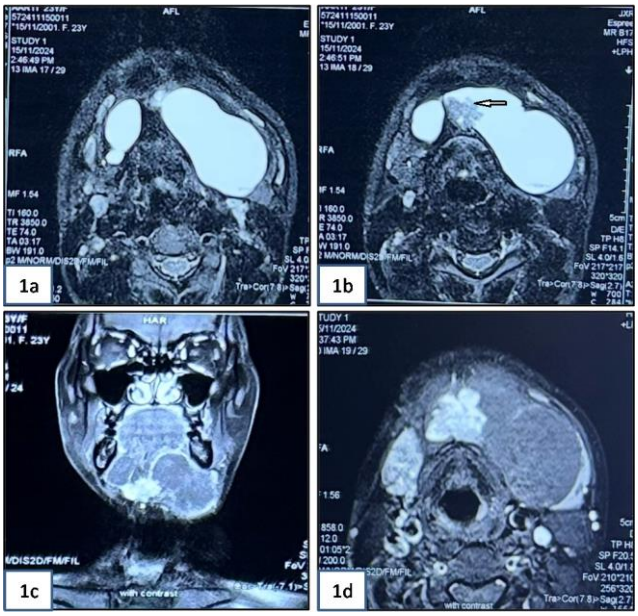

Figure 1: (a, b): Coronal and axial sections of contrast enhanced MRI neck shows bilateral plunging ranula in the sublingual space with enhancement of the mass lesion along infero-medial aspect of left ranula seen extending across midline into the right side cyst. (**c, d**): T2 weighted axial images of neck MRI shows bilateral plunging ranula in the sublingual space anterior to the carotids with an irregular soft tissue intensity mass lesion along infero-medial aspect of left ranula (arrow).

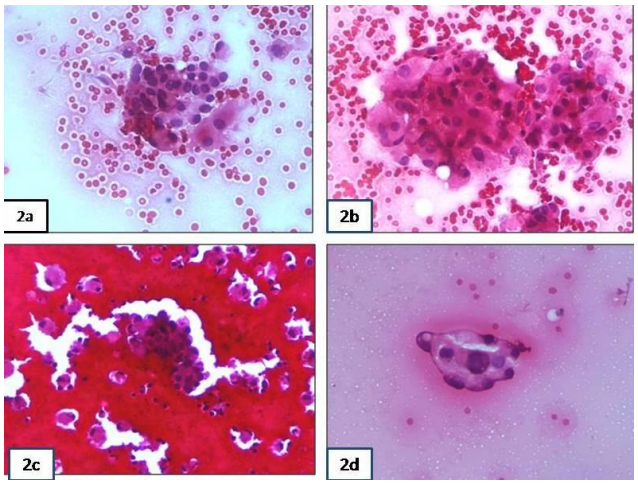

Figure 2: (**a**): Smears from the right side fluid showing cluster of cuboidal epithelial cells with squamous metaplasia. (H&E stain, x 400); (**b**): Smear from the left side fluid show metaplastic squamous cell admixed with cells with foamy cytoplasm, (**c**): Papillaroid epithelial cell clusters in a haemorrhagic background; (**d**): Papillaroid epithelial cell cluster with vacuolated cytoplasm

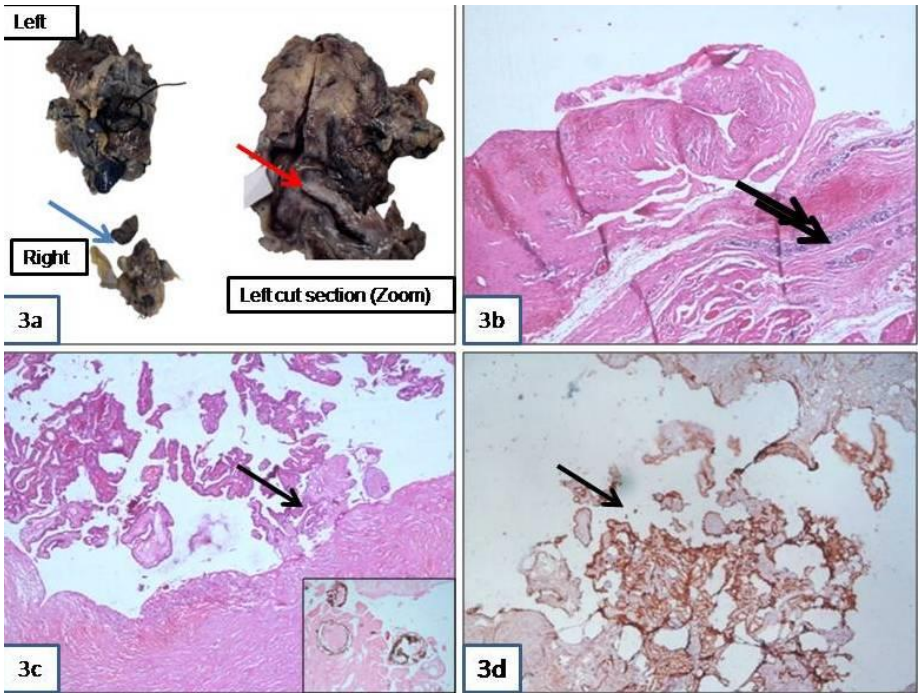

Figure 3: (a): Gross photograph showing left salivary gland with adherent cyst cut section showing thick cyst wall (arrow) & right side cyst (arrow head). (**b**): Histopathological image shows Thick fibrous wall with lymphoid collection in the wall. (H&E stain, x 40). (**c**): Histopathological image shows Intracystic tumour with papillary configuration with psammomatous calcification in inset. (H&E stain, x 400). (**d**): IHC (TTF) showing strong expression (x 400)

Table 1: Congenital cystic neck lesions

S. No.	Type of cyst	Age	Origin	Location/ Presentation
1	Thyroglossal duct cyst	90% before 10 yrs of age	Along the residual tract left by the thyroid gland during descent.	Midline at the level of hyoid bone 15-50%, suprahyoid 20-25%, infrahyoid 25-65%. Embedded i the infrahyoid strap muscles.
2	Branchial cleft cyst	20-40 years	Theories*	Mostly unilateral rare bilateral (familial) usually anterior to sternocleidomastoid. 60% left side,
***According to the theories for origin of BCC**				
a. *Failure of branchial pouch apparatus*				
First branchial cleft cyst		10-30 yrs (5-25%)	First branchial arch	Type I: near external auditory canal or in parotid gland or at angle of mandible Type II: associated with submandiublar gland OR in the anterior triangle of neck
Second branchial cleft cyst		20-40 yrs common (40-95%)	Second branchial arch	Along anterior border of upper third of sternocleidomastoid anywhere from skin of
Third branchial cleft cyst		Rare (2-8%)	Third branchial arch	Deep inside the sternocleidomastoid, may be with fistulous tract into larynx
Fourth branchial cleft cyst		Rare (1%)	Fouth branchial arch	Arises from lateral neck and parallels recurrent laryngeal nerve. Various locations
b. *Inclusion theory* - Epithelial tissue from upper aerodigestive tract or landular tissue enters a cervical lymph node through lymphatic channels, it stimulates degeneration resulting in the formation of a lateral cervical cyst.				
c. *Cystic degeneration* following migration into lymph nodes				
d. *Acquired theory*				
3	Cystic Hygroma	Found at birth or discovered later	Due to malformation of the lymphatic system.	Commonly occur in posterior triangle of the neck.

3. Discussion

Cystic masses of the neck can be congenital or acquired. Acquired lesions can be inflammatory or neoplastic. The congenital neck lesion can be either of thyroglossal duct (TD) or of Branchial cleft cysts (BCC) origin. (**Table 1**)

BCCs are the most common lateral cystic neck lesion, presumably due to incomplete obliteration of the branchial apparatus and TD are single midline neck swellings extending from the foramen cecum in the tongue base to the suprasternal region usually at or below the level of hyoid bone that moves with deglutition. Bilaterality is not a feature of TDC. A dermoid cyst may present as a midline cyst.[1] Other acquired cystic neck lesions include: Ranula, abscess, tubercular adenitis, primary carcinoma in aberrant thyroid, or metastatic lesions.[6] Ranula is a benign, submental fluid-filled cyst that forms anteriorly under the tongue due to a damaged salivary gland or blocked duct. The plunging ranula located in the submandibular region, extending downward is also suspected clinicoradiologically as in this case.[1] BCC is diagnosed using medical history, clinical appearance, diagnostic CT or MRI, FNAC & confirmed on HPE. They are congenital but usually present in late childhood & early adulthood.[1] BCC has been classified into 4 types (**Table 1**) Type 1 typically develops at the junction between the upper third & lower third of the sternocleidomastoid anteriorly and the posterior margin may about the submandibular gland as in our case.[6] Similar presentations have also been reported by other authors.[1,4,5] Bilateral BCC is rare & may have a familial predilection. Our case had no family history.

FNAC from branchial cleft cyst has yielded mixed results. Dispersed macrophages admixed with few lymphoid cells, in a hemorrhagic proteinaceous background have been reported.[2,7] Wokii et al reported the presence of mature squamous cells, as was also seen in the right branchial cyst aspirate.[8] The malignant component of PTC was usually missed on FNAC.[2,8] However papillary clusters were identified in our case from the left cyst, similar to findings of Cho et al.[4] The presence of vacuolated cell clusters in papillary configuration from cyst in submandibular location can also raise the possibility of intracystic papillary carcinoma of salivary gland origin, which can be ruled out on radiological examination.[6,9] The role of cytology cannot be overlooked.

The percentage of branchial cleft cysts that develop into carcinoma is rare, ranging from 4–22%. 66% were PTC and 33% were tonsillar squamous cell carcinoma.[10]

The first case of PTC arising in ectopic thyroid tissue within a branchial cyst was reported by Balasubramaniam et al. in 1992.[11]

Sidhu *et al*. proposed the criteria for diagnosing PTC in BCC as the presence of an epithelial lining layer with subepithelial lymphoid tissue, normal thyroid tissue in the wall, & absence of PTC in the thyroid.[7,12] All the above criteria were met with in our case except for the presence of normal thyroid component in the wall, similar to the findings of other authors.[3-5] It is postulated that all the normal thyroid tissue could be replaced by PTC.[4] The background lymphoid tissue was also not identified by Khwaja et al, unlike our case.[5] Psammoma bodies can be identified in the cyst wall or the papillae, as an additional feature, similar to our findings.[7] The presence of papillaroid clusters with vacuolated cells, is unusual, as seen in our case on FNAC, which has also been reported in PTC.[13] IHC (TTF & TG) can be confirmatory as seen in our case too.[4,5,7]

4. Conclusion

Branchial cleft cysts can harbour malignancies and a thorough clinic-pathological-radiological approach is useful in differentiating it from its close mimics. Presence of papillary neoplasm in the cystic neck masses demands a thorough investigation. It includes ruling out primary in the thyroid, as well as origin in the thyroglossal duct. Bilaterality rules out thyroglosal duct origin.

5. Conflict of Interest

None.

6. Source of Funding

None.

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