

Different Presentations of Branchial Cysts: A Case Series

Anju S^{1*}, Sankar S²

¹Junior Resident, ²Professor and Head of the Department Govt. Medical College, Kottayam, Kerala, India

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*Corresponding author: Anju S

Abstract

Among the branchial cysts, those arising from second arch, which appear in the mid neck are the most common. It is followed by cysts arising from the third and fourth arch, which appears in the lower neck, supraclavicular or suprasternal location. But branchial cysts presenting as a mediastinal mass is very rare. Here we present four different clinical presentations of branchial cysts with their gross morphology and microscopic findings.

Keywords: Branchial arches, branchial cysts, mediastinal mass.

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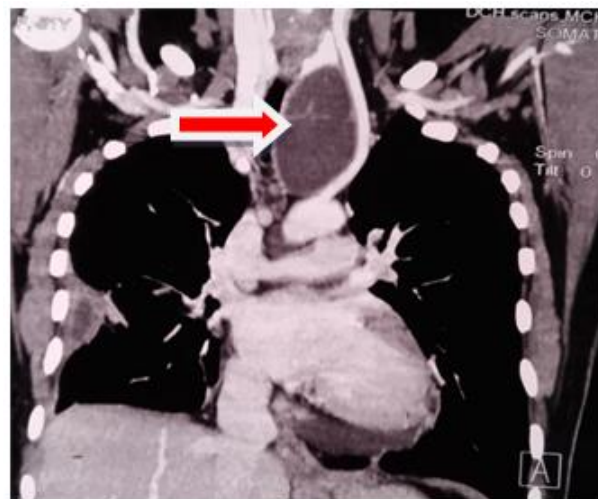
INTRODUCTION

The development of head and neck structures commence with the formation of branchial apparatus during the 4th week of gestation. There are 6 paired branchial arches. Each branchial arch consists of a core of mesenchyme covered externally by the ectoderm and internally by the endoderm. The 5th arch disappears and the 6th arch is rudimentary. Many congenital anomalies and cysts of the head and neck region are attributed to the aberrant development from these structures. Among these cysts those from the second branchial arch are the most common with a frequency of 90-95%. This is followed by cysts from the 3rd and 4th arch with a frequency of 2-8%. Those related to first arch appear in the preauricular area and those related to second arch appear anterior to the sternocleidomastoid muscle in the mid neck. Those related to third and fourth arch appears in the lower neck, in the suprasternal/supraclavicular location. Mediastinal location is very uncommon. The branchial anomalies can be sinuses, pouches or cysts. Branchial cysts presenting as a mediastinal mass is very rare. There are only very few documented cases of the fourth branchial arch anomaly, especially that involving the mediastinum.

CASE 1

A 51 year old female presented with fever and productive cough of 1 week duration. She was admitted in the general medicine department and treated with a diagnosis of pneumonia. She was on antibiotics for 2 weeks following which she got relieved of her symptoms. As part of the investigations for the same, CT thorax was taken. CT scan revealed a superior

mediastinal cystic mass with thin septations, which had a mass effect on trachea and esophagus along with atelectatic changes in the middle and lower lobe of the right lung.

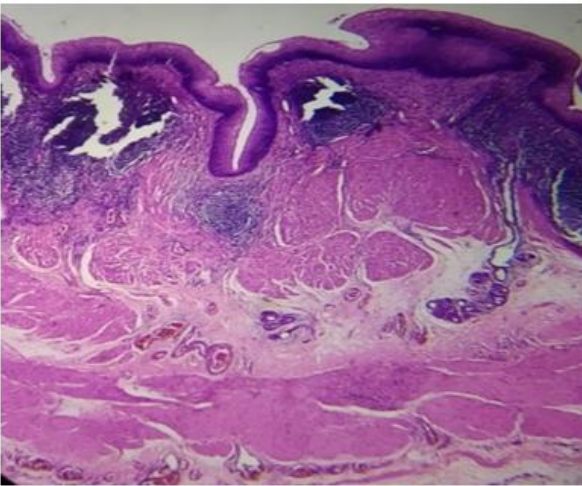


CT Thorax showing a well-defined lesion of size 4.7x3.7x8.5cm in the superior mediastinum

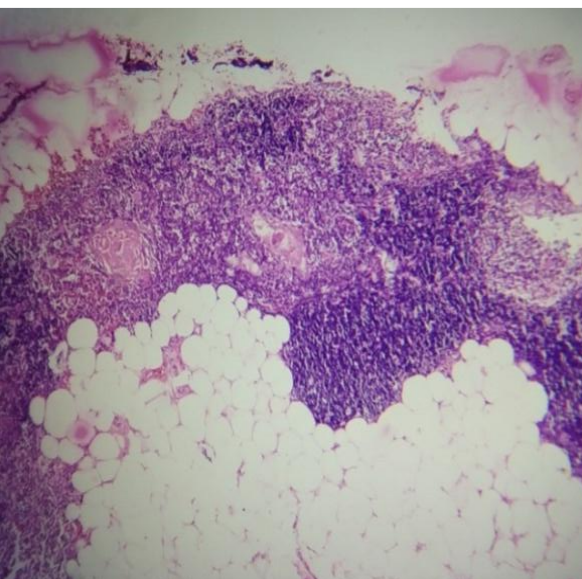
A midsternal thoracotomy was carried out which revealed a cystic mass which was excised. It was weighing 50gm, measuring 5x2.5x1.5cm. Cutting through the mass extruded creamy material and showed a biloculated cyst separated by a thick fibrous septa. Inner wall was smooth with tiny whitish specks.



Biloculated cyst separated by thick fibrous septa



H&E 100X: Section from cyst wall shows hyperkeratotic stratified squamous epithelium with a fibromuscular wall containing lymphoid follicles



H&E 100X: Section shows adjacent thymic tissue

H&E stained paraffin sections showed a cyst lined by hyperkeratotic stratified squamous epithelium with fibromuscular wall containing lymphoid follicles. Areas of calcification and congested blood vessels also were seen in the wall of the cyst. Adjacent compressed thymic tissue was noted. A diagnosis of branchial cyst related to thymus possibly arising from 3rd or 4th branchial pouch was made. The patient is remaining healthy and free of symptoms after surgery

CASE 2

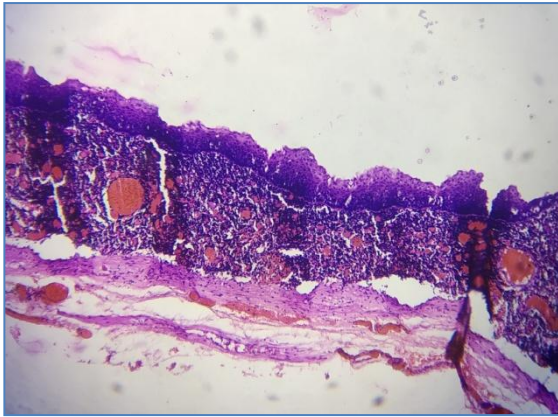
A 21 year old male presented with swelling, right side of neck since 6months, which gradually increased in size. On examination there was a swelling in the right side of neck, in the anterior border of sternocleidomastoid, measuring 5x4x2cm, which was soft in consistency with restricted mobility. MRI revealed a well-defined thin walled cystic lesion on right side of neck deep to the sternocleidomastoid muscle, suggestive of branchial cyst. It was excised and sent for histopathology.

We received a cystic mass weighing 20gm, measuring 4.5x3.5x2cm. Cutsection extruded brown coloured fluid and showed a unilocular cyst with smooth inner wall with focal solid area measuring 1.5x1.0x0.8cm.

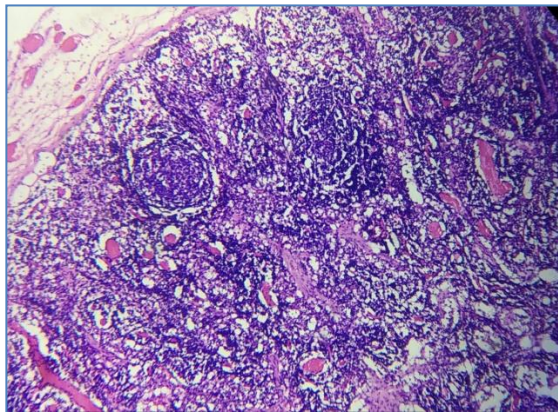


Uniloculated cystic mass with smooth inner wall with focal solid area

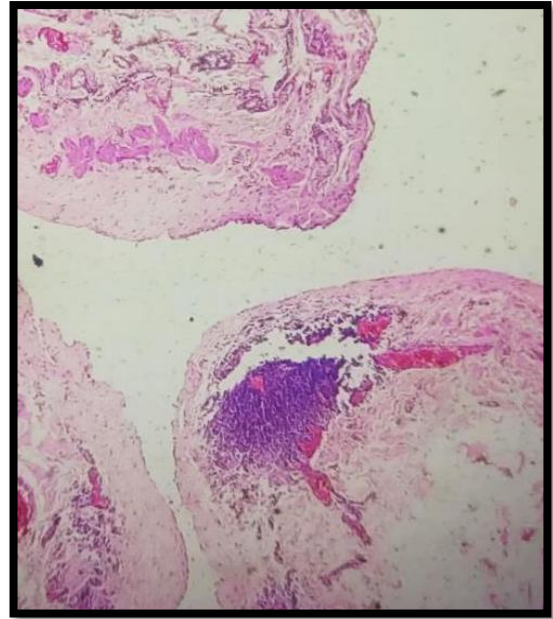
H&E stained paraffin sections show a cyst lined by stratified squamous epithelium and the fibrocollagenous wall shows lymphoid tissue with germinal centres along with congested blood vessels and hemorrhage. Section from the solid area also showed fibrocollagenous tissue with lymphoid follicles and hemorrhage. A diagnosis of branchial cleft cyst arising from second branchial pouch was made.



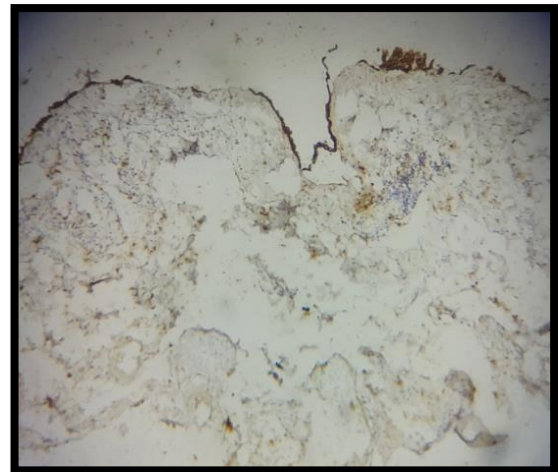
400x: cyst lined by stratified squamous epithelium



400 xs: cystwall showing lymphoid follicles and congested blood vessels



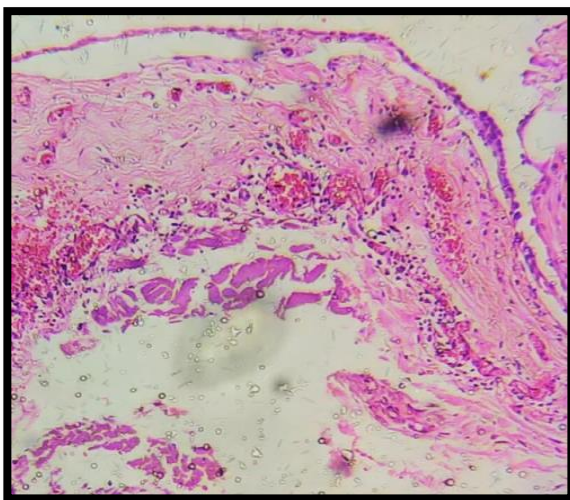
H&E, 400 x-Cyst walls shows lymphoid follicles, smooth muscle and congested blood vessels



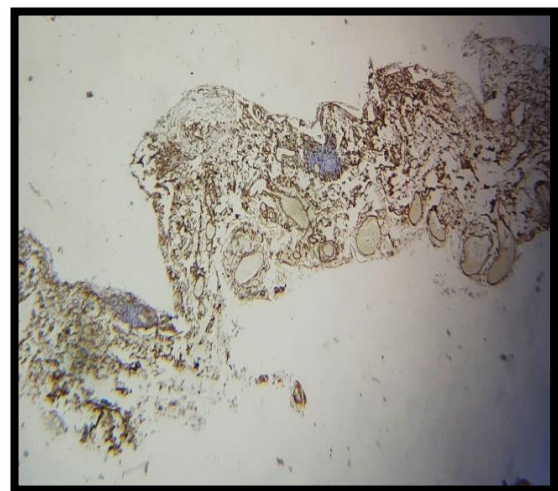
IHC, 400 xs- The lining cells show positivity for CK

CASE 3

A 35 year old man presented with difficulty while swallowing for last 7 months, especially for spicy and hot food. Clinical examination findings were within normal limits, but USG revealed a well-defined cystic mass in the right paratracheal region .It was excised and sent for histopathology. The specimen was received in multiple pieces which were part of a cystic lesion together measuring 5x 3x 1cm.No solid area seen.



H&E, 400x-Cyst wall lined by flattened cuboidal epithelium



IHC, 400 xs: The lining cells are negative for CD34 with positive internal control.

H&E stained paraffin sections showed fragments of a cyst wall lined by flattened cuboidal epithelium, wall of which shows lymphoid follicles, smooth muscle and congested blood vessels. Immunohistochemistry was done to distinguish from lymphangioma using the markers CD 34 and CK. The lining cells were positive for CK and negative for CD34 with positive internal control. Hence it was diagnosed as branchial cyst.

CASE 4

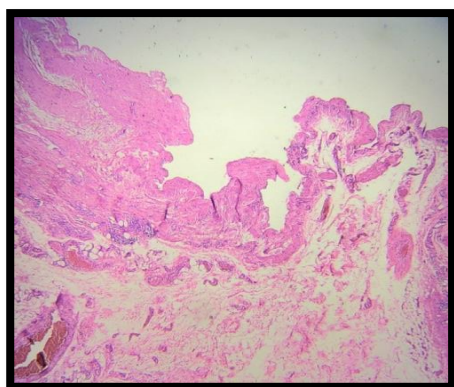
A 55 year old man presented with difficulty in swallowing for last 2 months. Clinical examinations were within normal limits. CT scan revealed a well-defined lesion in the right paratracheal region in the antero superior mediastinum measuring 7.6x7x6.3cm.

Excision was done for the same. We received a collapsed cyst measuring 7x5x5cm. Cutsection showed a biloculated cyst. Inner wall appeared shaggy. No solid area seen.

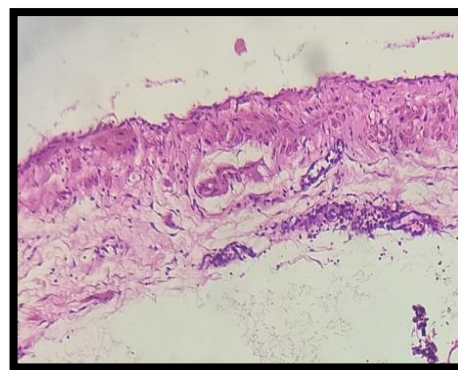
H&E stained paraffin sections showed fibrofatty tissue with a cystic lesion lined by flattened epithelium. Cyst wall shows smooth muscle and focal lymphoid aggregates. No thymic tissue identified. It was diagnosed as benign cystic lesion suggestive of branchial cyst.



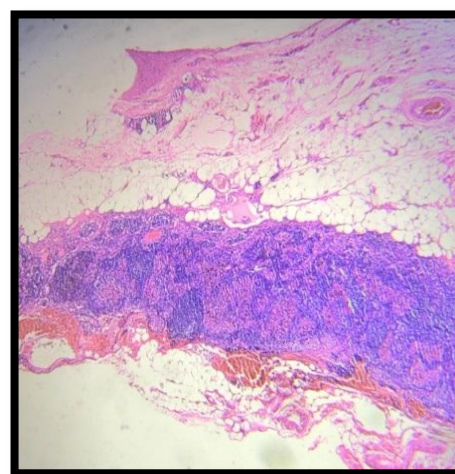
Biloculated cyst with shaggy inner wall



100x-cyst lined by flattened epithelium and wall showing smooth muscle



400x-cyst lined by flattened epithelium



Cyst wall showing lymphoid aggregates

DISCUSSION

This report describes four different presentations of branchial cleft cysts, arising from second, third and fourth branchial arches. Branchial cysts arising from second arch presenting as neck swelling are the most common among branchial cleft cysts.

Mediastinal cysts are relatively uncommon, accounting for 10 to 15% of radiologically detected masses at this site. Several tissue types can be seen in such lesions. Sometimes neoplasms that undergo cystic degeneration also may present as a mediastinal cyst. Its unusual presentation in the mediastinum causes a delay in the diagnosis.

Some mediastinal cysts may contain more than one of these constituents, relating to the fact that many intrathoracic cysts are congenital and also to the close proximity in which the embryonic foregut, pleuropericardial membranes and branchial pouches are formed during early morphogenesis.

Branchial cysts may rarely arise within the thyroid gland. 33% of patients with congenital third branchial arch anomalies and 45% with fourth branchial arch abnormalities present with acute infectious thyroiditis.

Patients usually present between 20 to 40 years of age. 20% of cases are seen in children. 2% to 3% of cases are bilateral. Branchial cleft cysts are 10 times more common than fistulas.

The cysts are lined by stratified squamous epithelium/columnar/flattened cuboidal epithelium. Cysts derived from branchial cleft have squamous epithelial lining while cysts derived from branchial pouch have respiratory epithelial lining. But repeated infections may cause squamous metaplasia. The cyst wall shows lymphoid tissue, smooth muscle bundles and rarely sebaceous/mucinous glands. Sometimes the cysts may get infected also.

Differential diagnosis is

- First branchial cleft cyst: epidermoid cyst, dermoid cyst, cystic sebaceous lymphadenoma
- Second branchial cleft cyst: lateral thyroglossal duct cyst

- Third branchial cleft cyst: papillary carcinoma with cystic change.
- Fourth branchial cleft cyst: thymic cyst.

During the surgery the cyst should be followed into the neck and excised completely in case of mediastinal lesions. It will minimise the chance of recurrence, which is usually high following incision and drainage of third and fourth branchial cyst. Recurrence rate is around 5% and depends on completeness of resection. There are no reported cases of malignancies arising from the branchial cysts.

CONCLUSION

Branchial cysts should be a differential diagnoses in case of neck and mediastinal cysts. Though branchial cysts arising from second arch is common, rarely it can also arise from third or fourth branchial arches. Complete resection will minimise the recurrence rate.

| Case No | Age/Sex | Clinical presentation | Site of the cyst | Size | Lining of the cyst in histology | Finding in the cyst wall | Possible arch of origin |
|---------|-----------|--------------------------|--|---------------|---|------------------------------------|------------------------------------|
| Case 1 | 51/female | Pneumonia | Superior mediastinum | 5x2.5x1.5 cm | Hyperkeratotic stratified squamous epithelium | Smooth muscle, lymphoid tissue | 3 rd or 4 th |
| Case 2 | 21/male | Swelling neck | Anterior border of sternocleidomastoid | 5x4x2 cm | Stratified squamous epithelium | Lymphoid tissue | 2 nd |
| Case 3 | 35/male | Difficulty in swallowing | Right paratracheal region | 5x3x1cm | Flattened cuboidal epithelium | Smooth muscle, lymphoid follicle | 3 rd or 4 th |
| Case 4 | 55/male | Difficulty in swallowing | Right paratracheal region | 7.6x7.6x6.3cm | Flattened cuboidal epithelium | Smooth muscle, lymphoid aggregates | 3 rd or 4 th |

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