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Cyst In Mediastinum – A Rare Case Report

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ABSTRACT

Background: *Cystic lesions in mediastinum are rare (10-27%). They are believed to originate as a result of developmental anomalies of the foregut. Generally found in young adults (average age, 31 years).*

Case Description: *Clinical presentation: 55 year old female presented with dry cough for 6 months, breathlessness for 3 months, progressively increasing for past 10 days presented as a acute emergency in the casualty. Radiological imaging revealed a non-enhancing cystic lesion at the upper two third of right hemithorax. Thoracotomy was performed and intra-operatively a multiloculated encapsulated tumor adherent to lung and lateral aspect of trachea, containing straw colored fluid was noted. Cyst was removed in piecemeal and exact origin was not made out.*

Gross: *specimen was received in multiple fragments with grey white external surface and irregularly thickened yellowish cut surface.*

Microscopy: *showed a thick fibrocollagenous wall discontinuously lined by single layer of ciliated coloumnar epithelium with abundant inflammatory infiltrate of foamy histiocytes, lymphocytes and spindle cells- fibroblastic proliferation, lung parenchymal attachment with no evidence of peripheral thymic tissue hence diagnosed as Branchial cyst.*

Discussion: *Branchial cleft cysts are located on the lateral side of neck, mediastinum is involved if it arises from fifth to sixth cleft. Theoretically, the existence of fifth or sixth branchial cleft and its anomalies in mediastinum are possible in humans.*

Conclusion: *Branchial cyst in mediastinum is rare with only 3 cases being reported in the literature till now. The key to uncover the mystery of Mediastinal cyst is only by histological means.*

Keywords: *Branchial cyst, Mediastinal cyst.*

INTRODUCTION

Reports of Mediastinal branchial cyst are extremely rare. Though they are congenital, average age of presentation is 31 yrs (young adults).Three cases of mediastinal branchial cleft cysts have been reported in the literature. One such case is being reported in a 55 yr old female at SRM Medical College Hospital Kattangulathur. It is reported for its rarity and unusual presentation.

CASE REPORT

Clinical Presentation

A 55 year old female presented with dry cough for 6 months, breathlessness for 3 months, progress-ively increasing for past 10 days. X-ray Fig 1(a) revealed a well defined mediastinal mass adjacent to right cardiac border involving the Right upper & middle hemi thorax, trachea & heart pushed to left, costo and cardiophrenic angles were free.

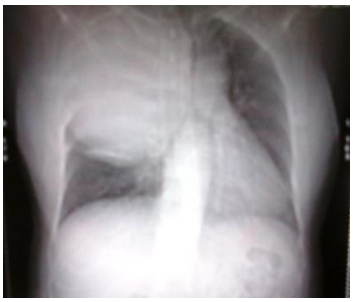

Fig 1(a)

Chest CT Fig 1(b) revealed a non-enhancing cystic lesion located at the upper two third of Right hemi thorax, the heart was found to be deviated to the left, and there was a well defined lobulated thick walled cyst with no evidence of an abnormal enhancing lesion within the cyst.

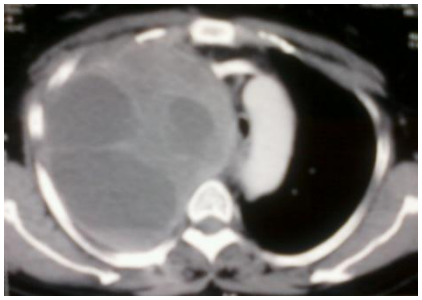

Fig 1(b)

A diagnostic thoracotomy was performed to obtain an excision biopsy. Intraoperatively awell encapsulated tumor adherent to lung & lateral aspect of trachea containing straw colored & hemorrhagic fluid at several loculation was noted. Cyst was removed in piecemeal due to high vascularity.

Macroscopic Appearance

Gross specimen was received in two containers, the largest specimen was fragmented and was reminiscent of a collapsed cyst measured about 10 x 8 cm. The external surface was grayish white and smooth Fig 2(a).

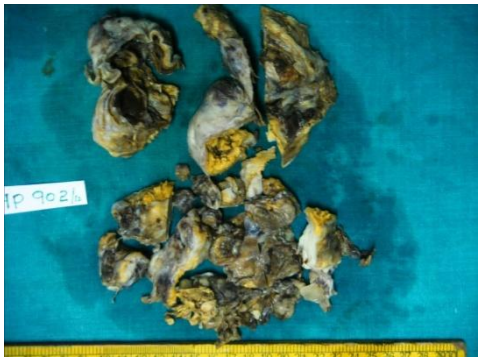

Fig 2(a)

The cut surface was irregularly thickened, wall measuring 0.5 – 1 cm with yellowish patches. The inner surface is ragged, grayish brown and friable Fig 2(b).

Fig 2(b)

Microscopic appearance

Sections from cyst showed a thick fibro collagenous wall discontinuously lined by single layer of ciliated columnar epithelium Fig3(a), with moderate to abundant inflammatory infiltrate composed of foamy histiocytes Fig3(b), lympho-cytes and spindle cells- fibroblastic proliferation Fig3(c). The thick fibrous capsule showed vascular channels, and there is no evidence of peripheral thymic tissue or hassal's corpuscles and hence diagnosed as Branchial cyst.

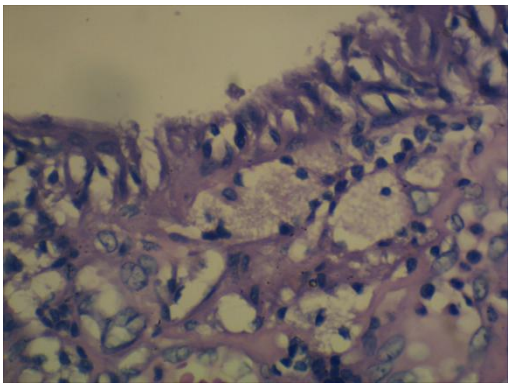

Fig 3(a)

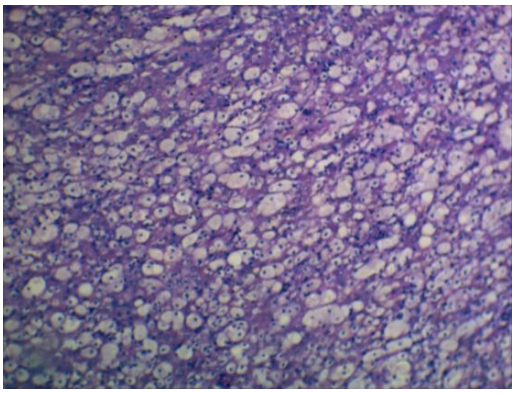

Fig 3 (b)

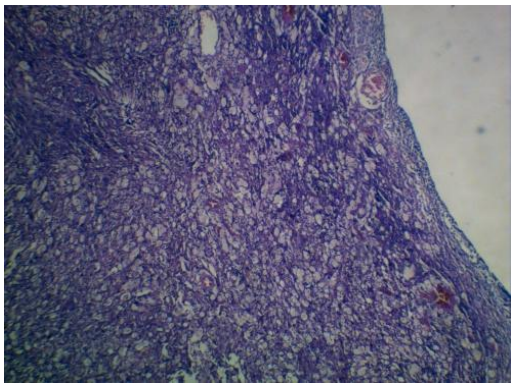

Fig 3(c)

DISCUSSION

Mediastinal cystic lesions are relatively uncommon and comprises about 10-15% of the lesions [(1)]. The commonly encountered cystic lesion include Parathyroid cyst, Thymic cyst, Bronchogenic cyst, Pericardial cyst, Enteric cyst, Cystichygroma/lymphangioma and Cystic meningocoele[(2)]. The diagnosis is usually made by examining a patient's clinical history and manifestations, as well as the anatomic position or certain features seen with the CT and pathological findings.

The term "branchial apparatus" refers to the embryologic precursors that develop into the tissue of the neck. Many developmental anomalies of the branchial apparatus have been identified: cysts, fistulas, sinuses, ectopic glands, and malformations of head and neck structures. When a branchial cleft is not properly involuted, a branchial cleft cyst forms. The most common anatomical site for the occurrence of branchial cleft cysts is in the cervical area, generally anterior to the sternomastoid muscle in the upper or middle portion of the neck [(3)]. Anomaly of the second branchial cleft accounts for approximately 95% of all branchial cleft cyst [(3)].

Usually the Branchial cyst presents in younger age group in the cervical region. In this case the age of presentation is 55 and the site is mediasinum making this a rare case scenario. William et al. suggests that mediastinal branchial cleft cysts originated from fourth or fifth branchial cleft .Because the fourth, fifth, and sixth branchial clefts are located below the fourth aortic arch artery during the embryologic stage, the cyst would course below the fourth branchial artery or its derivation namely, the aortic arch or the right subclavian artery [(4-6)].

The radiologic feature of the mediastinal branchial cleft cysts exhibit no specific findings to distinguish them from other anterior mediastinal cysts, which include thymic or bronchogenic cysts [(7,8)]. CT finding in this case shows a huge space occupying cyst, encompassing the right hemithorax (superior, anterior, middle) and adherent to lung. If the cyst is restricted to a single mediastinal compartment, it would have presented earlier. But in this case it is not restricted to a single compartment attributing towards delayed presentation.

The patient presented as acute emergency with history of breathless for past ten days with possible cause of cyst compressing the bronchi.

Pathologically it is important to distinguish a branchial cleft cyst from a thymic cyst because the thymic cyst would be lined byciliated columnar epithelium. The correct diagnosis of thymic cysts can be established by the detection of thymic tissue and Hassall's corpuscles within the cyst

The classical branchial cleft cyst is characterized histologically by an epithelial lining which is typically squamous but may be ciliated columnar. Underlying the epithelium is abundant lymphoid tissue with germinal centers. The same histological picture is seen in this case. In addition, the presence of xanthomatous response and fibroblastic proliferation in this case, forms the tell tale evidence of chronicity of the cyst

Very rarely only had such Branchial cysts been reported in the literature[(4,5)]. Existence of this entity came in literature only after a case report in 1969. And after the proof of existence, only one case have been reported making this the third case reported so far. The two cases reported in 1969 and in 2007 were also in the fifth decade, as in this case making case scenario intresting requiring further work up neither to analyse the cause of presentation at fifth decade or it being incidental. In limited case reports it was found that there was

no sexual predliction though many more cases are required to prove. Further a large branchial cyst asymptomatic for years together and suddenly being symptomatic is quite interesting and make this case unique as previously reported cases were diagnosed incidentally. In this case the sudden increase in the symptom may be due to the lesion compressing the bronchus as evidenced by the CT images.

Post operatively patient was followed up for three years and she is free of any symptoms.

CONCLUSION

This case is being reported for its rarity as only two cases have been reported so far and for its unusual presentation. This case is also presented to emphasise that a congenital lesion can be asymptomatic for many years to present only in elderly individuals as evidenced by other case reports too, and also to suggest that, all space occupying mass lesions need not always be of malignant origin. It is also presented to highlight that acute breathlessness can also be due to a long standing congenital cyst and this prospective should also be considered by the clinicians.

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