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PEDIATRIC NECK MASS THYMOPHARYNGEAL DUCT CYST

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Abstract:
Neck masses in children are common issues. Differential diagnosis includes infectious, congenital and neoplastic. In this article, the researchers have presented a case presentation of an unusual neck mass. A 7-year-old boy presented with a history of left anterolateral neck mass noticed with a clinical impression of venolymphatic malformation versus branchial cleft cyst. The 1-year back radiological evaluation revealed a picture of thymopharyngeal duct cyst.
Thymus develops in the neck from the 3rd pharyngeal pouch and descends to anterior superior mediastinum forming a tract, thymopharyngeal stalk that obliterates embryologically.
Failure obliteration results in thymic remnant and thymopharyngeal duct cyst, which can be presented in childhood as lateral neck mass. The thymopharyngeal duct cyst can be best managed by complete surgical excision.
Keywords: *Neck masses, Venolymphatic malformation, Thymopharyngeal duct, Thymic remnant.*

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INTRODUCTION:

Neck masses are common in children and infectious, congenital and neoplastic are common differential diagnostic features associated with this condition. In contrast to adults, the majority of neck masses in children are categorized as benign inflammatory and congenital lesions. The management of these neck masses in the children are also different from adults. However, one significant point to be considered is that management is challenging and depends on the history and physical examination. The radiological evaluation may be helpful, but some cases may need a surgical biopsy to confirm the diagnosis.

AIM OF THE STUDY:

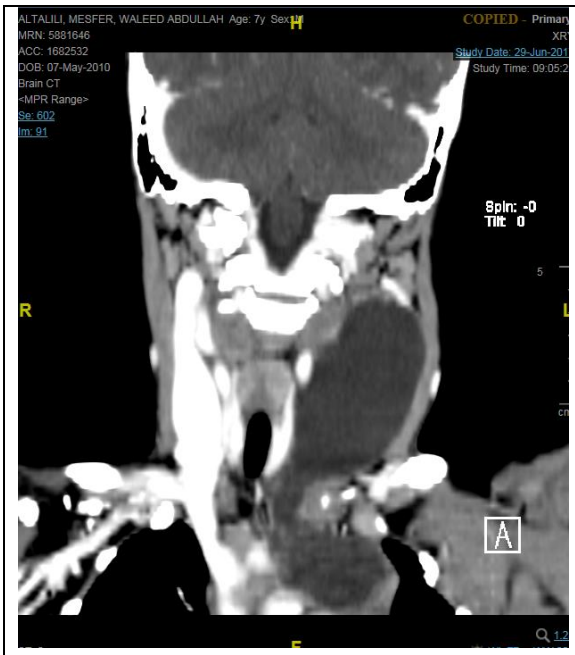
This research article is a case study based presentation of the neck mass in children. In this article, the researchers have presented a case presentation of an unusual neck mass in a child with an initial impression of thymopharyngeal duct cyst. The earlier diagnosis was carried out radiologically and confirmed by surgical histopathology.

CASE PRESENTATION:

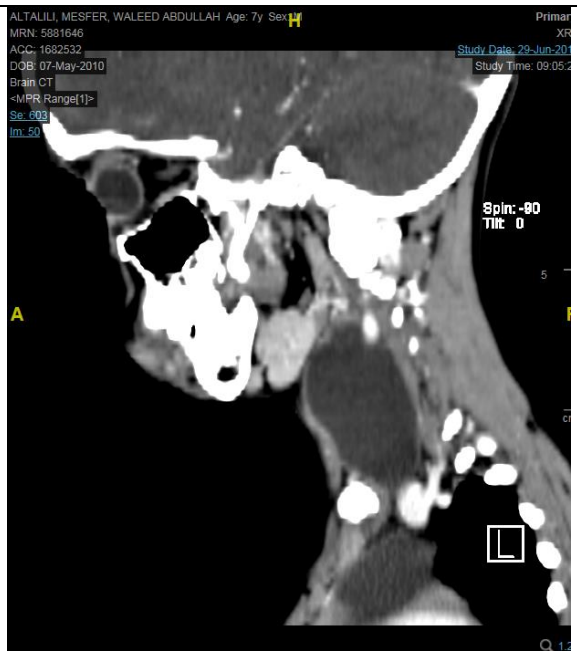
A 7 years old boy was presented to otolaryngology outpatient service with a 1-year history of a painless left lateral neck mass. The neck mass was increasing in size in the last 3 months causing disfigurement with no compression symptoms. He had no history of fever or recent infection. He had not experienced weight loss or night sweating. In addition, no history of recent travel or contact with sick people was reported. Clinical examination revealed soft non-tender diffuse compressible left neck mass measuring about 11 x 5 cm. Flexible nasolaryngoscopy showed no pharyngeal mass effect.

Figure 1: Case Presentation

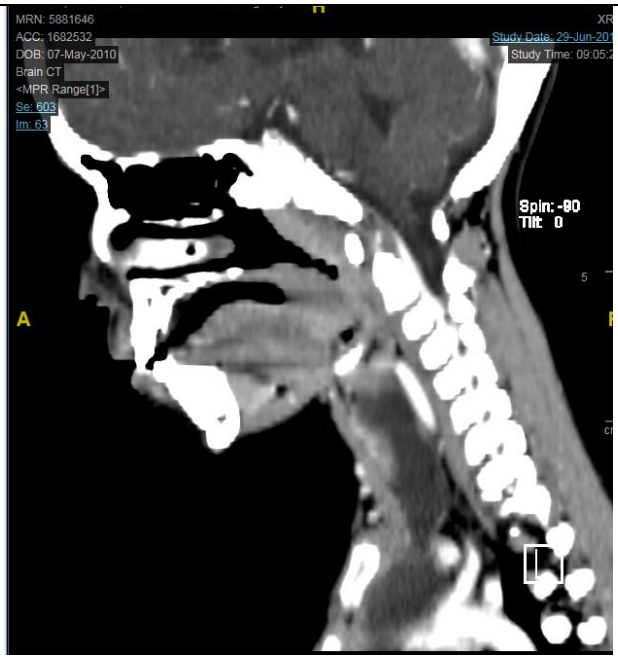
Working diagnosis for a lateral neck mass in children includes:
-Congenital (Branchial cleft anomalies, Laryngocele, Lymphatic or vascular malformation, Thymic cyst).
-Neoplastic (Lymphoma, Sarcoma, Carotid a body tumour)
CT neck with contrast was requested and showed cystic non-enhancing left neck mass extending to upper mediastinum. CT neck with contrast, coronal view showed a hypodense cystic mass in the left side of the neck with extension to upper mediastinum. While the sagittal view has shown cervical and chest component of hypopharyngeal duct cyst.



CT neck with contrast, coronal view showed a hypodense cystic mass in the left side of the neck with extension to upper mediastinum



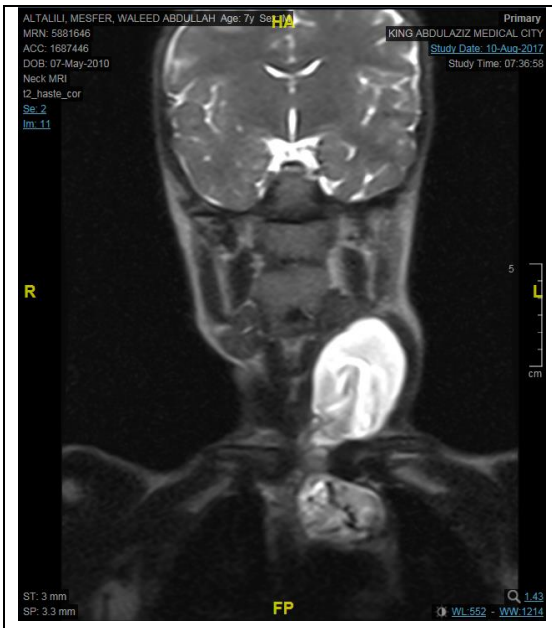
CT neck, sagittal view showing cervical and chest component of hypopharyngeal duct cyst.



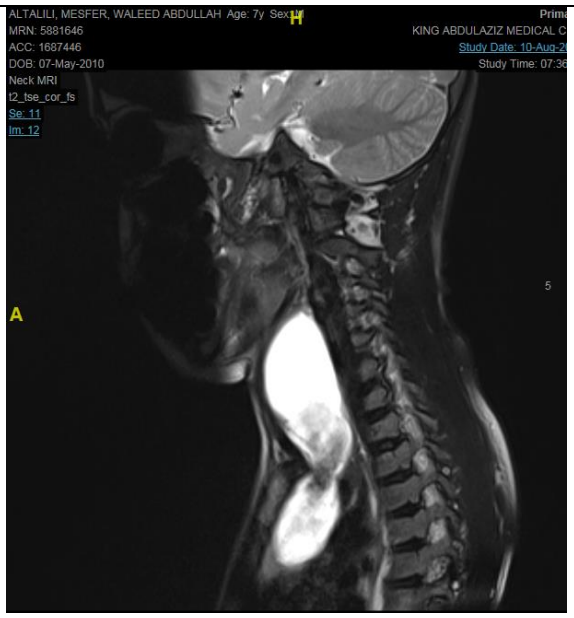
CT neck, sagittal view showing cervical and chest component of hypopharyngeal duct cyst.

Figure 2: CT neck

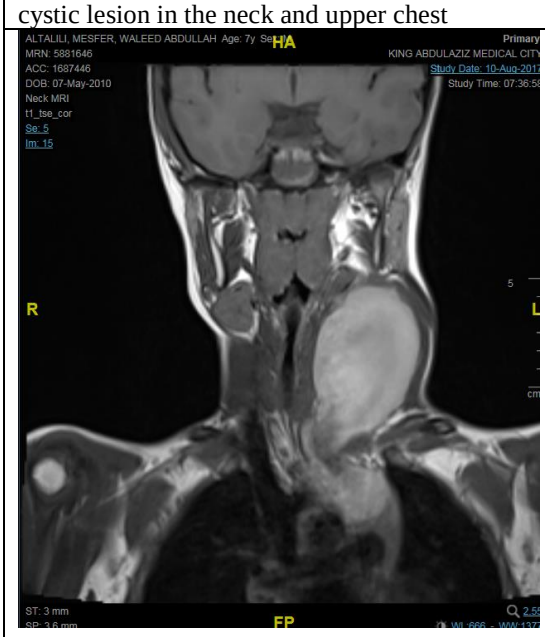
MRI neck showed left cystic lesion originating from the anterior mediastinum to the lower left cervical region, high signal intensity on T1 and T2 representing the proteinaceous component within no septation.



MRI neck, coronal view showed hyperintensity cystic lesion in the neck and upper chest



MRI neck, sagittal view showing communication between cervical and thoracic lesion.



MRI neck coronal view showed hyperintensity lesion in T1

Figure 3: MRI Neck

The lesion measures 11 x 5 x 5 cm and is seen lateral to the carotid sheath and medial to the SCM with mild mass effect on the left thyroid gland the findings are in keeping with thymopharyngeal duct cyst, which extends from thymus to the cervical level. No other lesions were seen.

After the diagnosis and confirmation, surgical excision was planned and pediatric surgery consultation was done for possible thoracotomy in the case of failed neck approach excision. Intraoperative, transverse neck incision was made on skin crease and dissection was made until reaching the cystic mass anterior to SCM. The dissection continued inferiorly and cystic mass separated carefully from carotid sheath posteriorly. Gentle pulling of cystic mass with finger dissection reveal total removal of the cyst with part of thymus excised and sent for histopathology.

Figure 4: Surgical Procedure

The postoperative management of the patient was also ensured. The patient underwent pediatric ICU for close observation and was then shifted to the ward and discharged home the next day. Outpatient follows up was maintained and, a patient seen in the clinic with no complaint and healed surgical wound flexible scope done showed no compression and mobile vocal folds bilateral. Histopathology of the patient in the gross section and the microscopic section was also carried out. Gross section: consist of a single piece of cystic like tissue, measures 14 x 4 x 1.2 cm, the outer surface is tan and glistening. Upon sectioning, the cystic and solid areas were seen.

Microscopic Description

Sections examined shown cystic spaces lined by low cuboidal and respiratory epithelium in other foci. The unremarkable thymic tissue is identified. The underlying fibrous stroma has shown acute and chronic inflammatory cell infiltrates. Lymphoid follicles and foci of cholesterol granuloma are noted.

DISCUSSION:

Neck masses in children are common and differential diagnosis included infectious, congenital and neoplastic. In contrast to adults, the majority of neck masses in children are categorized as benign inflammatory and congenital lesions. Management of the patient was challenging and in this regard, the history and physical examination of the patient were carried out. The radiological evaluation may be helpful, but some cases may need a surgical biopsy to confirm the diagnosis. The literature review has also shown a few scattered similar cases.

Hsieh et al. (2003) carried out a 20-year-study on 331 pediatric patients presenting with cystic neck masses, only one case was diagnosed as a thymic cyst (frequency 0.30%) [1]. This has revealed that thymic cyst is a rare disease and is only presented by only a few children in thousands. Cigliano et al. (2007) also carried out the study to evaluate the frequency of thymic cyst in case of patients with cystic neck masses. They carried out a research during a period of 16 years in three major institutes, only nine cases of thymic cysts were reported [2]. Both of these studies have provided evidence that thymic cyst is a rare disease. Khariwala et al. (2001) carried out the research on the cervical preservations of thymic anomalies in children, while De Caluwe (2002) also conducted the research on the cervical thymic cyst. Both of these studies found that cervical thymic cyst is the rare lesion, which is often misdiagnosed as the branchial cyst. Until 1978, fewer than seven cases were reported in the literature, a number that seemed to rise to 150 cases in 2004 [2, 3, 4]. Komura et al. estimated this number has increased to more than 80 cases and the management of the patient is imperative to avoid this complication after the surgical procedure [5].

Thymus develops in the neck from the 3rd pharyngeal pouch and descends to anterior superior mediastinum forming a tract, thymopharyngeal stalk that obliterates embryologically. Failure obliteration resulted in thymic remnant and thymopharyngeal duct cyst, which can be presented in childhood as lateral neck mass. Cervical remnants of thymic tissue have been found in up to 30% of infants studied at autopsy [6].

CONCLUSION:

The thymopharyngeal cyst is a rare disease presenting as neck mass in children. It is a benign disease, which is mostly presented with no symptoms other than neck mass. Further evaluation for airway compressive symptoms should be addressed. Imaging of choice is MRI with contrast to assess surrounding structures, consistency and extension of mass and to rule out vascular malformation. The thymopharyngeal duct cyst best managed by complete surgical excision with careful dissection to great vessels, vagus nerve and important surrounding structures are effective to prevent complications.

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