Unusual Cystic Hypopharyngeal Mass in a Child with Obstructive Symptoms

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Abstract We present a rare case of a cystic posterior pharyngeal wall mass in a 5 year old child in whom prior surgery without biopsy led to considerable delay in both diagnosis and definitive treatment. The child presented with recurrent aspiration pneumonia, dysphagia and obstructed breathing from birth. Transoral wide excisional biopsy established the diagnosis of foregut duplication cyst. Only seven reported cases have been described in the English language literature till date.

Keywords Congenital cyst · Enterocystoma · Hypopharynx

Introduction

Obstructive mass lesions in the pharynx in children pose a dilemma, particularly if they have been present since birth. Foregut duplication cyst is one such rare congenital anomaly in which portions of the alimentary system that are derived embryologically from the foregut during development, become entrapped in an ectopic location. These cysts contain heterotopic islands of gastrointestinal mucosa. The heterotopic mucosa is most often gastric, but occasionally intestinal, colonic or pancreatic [1]. The duplication of this mucosa is what gives this entity the name.

Most foregut duplication cysts occur in the thorax or abdomen. Their appearance in the head and neck region, especially in the hypopharynx is particularly rare [2]. Till date, only 7 definite foregut duplication cysts of the

hypopharynx have been described in the literature [3]. The present case highlights the diagnostic dilemma faced and the successful management of a child with this rare congenital malformation.

Case Report

A 5 year old boy hailing from a town in North India was referred with a history of noisy breathing, weak voice, recurrent lower respiratory infections and dysphagia. At birth, the child was a full term normal delivery with normal APGAR scores. He presented on the 20th day of life to the local pediatrician with recurrent apneic episodes and febrile seizures for which he was commenced on anticonvulsants. By 1 month of life he began having recurrent episodes of aspiration pneumonia and failure to thrive.

At the age of 14 months the condition worsened to such a degree that he needed antibiotic treatment every 15 days. He was referred first to a local ENT surgeon who reported a smooth, soft, pale, mucosa-covered mass arising from the posterior pharyngeal wall at the level of the epiglottis. Deep wide-bore needle aspiration yielded 6 ml of dark brown colored fluid. Cytological analysis of the fluid showed a few lymphocytes with neutrophils and cells with elongated, oval nuclei, suggestive of endothelial cells, in a background of proteinaceous material. This was reported by the pathologist as being consistent with a diagnosis of lymphangioma.

Following aspiration, symptoms were temporarily alleviated for a few months but recurred. At the age of 2 years, the patient was referred to a bigger centre in the same region where marsupialisation of the cyst by radiofrequency ablation of the roof of the mass (without biopsy) was performed. The lesion was described as a posterior pharyngeal wall cyst. The child developed aspiration pneumonia in the

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postoperative period. After recovery, he had temporary relief of symptoms for about a month. Thereafter, he developed bouts of aspiration pneumonia as before.

Three years later, because of the persistent symptoms of recurrent aspiration and dysphagia, the child was referred to our hospital which is a multispecialty, tertiary care, referral centre. He was malnourished and had noisy, stertorous breathing. There was no cyanosis. Fibre optic endoscopy confirmed the presence of a large, lobulated, mucosa-covered mass arising from the posterior pharyngeal wall, overlying and obscuring a view of the larynx. MRI scanning of the neck (Fig. 1) showed an irregular lesion measuring $29 \times 13 \times 30$ mm in size arising from the posterior pharyngeal wall which had a high signal in T1 and T2 weighted images. Contrast enhanced scans showed no evidence of hypervascularity. The tumor was bulkiest at the level of epiglottis, extending superiorly 8 mm above the level of epiglottis, inferiorly within a few millimeters of the post-cricoid region and laterally into the lateral pharyngeal wall. The mass was confined to the mucosa and submucosa with no extension into the muscle layer. There was no cyst like structure with fluid.

Transoral excision biopsy of the mass by placing the child in the tonsillar position was planned and performed. The mass was excised in toto and the raw area covered by mucosal flaps which were reapproximated with absorbable sutures. Histopathological examination showed a lesion composed of fibrocollagenous tissue lined partly by stratified squamous epithelium and partly by gastric body type epithelium with some of the fragments containing muscularis propria and muscularis mucosa (Fig. 2). A diagnosis of marsupialised foregut duplication cyst was made.

X-ray of the soft tissues of the neck, lateral view, done 2 weeks postoperatively showed significant improvement of

Fig. 1 T1 weighted MRI scan of the neck with contrast showing an irregular, lobulated, enhancing mass arising from the mucosa and submucosa of the posterior pharyngeal wall obstructing airway

Fig. 2 Lesion composed of fibrocollagenous tissue lined partly by stratified squamous epithelium and partly by gastric body type epithelium containing muscularis propria and muscularis mucosa (H&E staining $\times 50$)

the airway and regression of the mass (Fig. 3a, b). The child was decannulated and feeding well. At 1 year's follow up the child was found to have gained weight and had no evidence of residual lesion clinically.

Discussion

Duplication cysts of the alimentary canal occur with a frequency of about 1 in 4,500 [2]. Head and neck location of these cysts is rare. When the pharynx is involved, the tongue is the most common site and the hypopharynx the least common [1].

All of the previously reported cases of foregut duplication cyst except one have presented with stridor and respiratory obstruction [4]. Common differential diagnoses of retropharyngeal lesions present from birth or early infancy include teratoma, ectopic thyroid, aberrant thymus and vascular malformation.

Although this child had a congenital lesion which was very symptomatic, there was considerable delay in making a definitive diagnosis and providing the optimal treatment. Similarly, in many of the cases reported, repeated procedures have been required in children with foregut duplication cysts, as the initial treatment had not included complete excision and biopsy [2–5].

The diagnosis of foregut duplication cyst is based on a triad of: (1) Cyst covered by a smooth muscle coat (2) Presence of epithelium derived from the foregut (3) Attachment to a portion of the foregut. In the present case, all three criteria were fulfilled. Ectopic gastric mucosa is also known as “gastric choristoma.” This may occur in either a solid (also called “gastric heterotopia”) or cystic (also called

Fig. 3 **a** and **b** Preoperative (**a**) and postoperative (**b**) X-rays of the soft tissues of the neck lateral view showing improvement in the airway

“enterocystoma”) form [4]. The present case is, hence, an enterocystoma.

Various theories have been proposed regarding their origin. During normal embryonic development, the primordial notochord which is derived from ectoderm and later forms the neural tube, fuses with the endoderm which later forms the foregut. The notochord later separates from the endoderm and the neural tube and foregut continue to develop independently.

According to the split notochord theory by Bentley and Smith [6], abnormal adherence of endoderm (which differentiates into the foregut) and ectoderm (which forms the neural tube) and herniation of the same through a sagittally split notochord dorsally may lead to a neurenteric fistula. If the fistula is not closed by subsequent differential growth of nearby tissues, a variety of abnormalities including a duplication cyst may result.

Other theories proposed include that of Veeneklass [7] who suggested that if the separation of notochord and foregut did not occur, the adherent part of the esophagus could be separated from the developing esophagus and form a cyst. This would result in a foregut duplication cyst. In an experimental study on Adriamycin-injected rats, Qi et al. [8] were able to demonstrate that failure of the foregut to detach from the notochord at the normal time may contribute to development of these cysts, giving credence to Veeneklass’s theory.

Conclusion

Foregut duplication cyst is a rare congenital anomaly characterized by the presence of ectopic gastrointestinal mucosa

in a cystic lesion which chiefly occurs in the thorax and abdomen. We report an unusual presentation of foregut duplication cyst which involved the hypopharynx and contained gastric mucosa.

The present report highlights the need to consider foregut duplication cyst as a rare possibility for a child presenting with an obstructive cystic lesion of the posterior pharyngeal wall. Early surgical excision is the treatment of choice.

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