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Glandular hamartoma is an extremely rare congenital malformation of the larynx. Presenting symptoms result from airways obstruction and may include slowly rising respiratory distress, stridor, changes in voice, eating and activity levels.

Management consists in local mass excision with a good functional result and prognosis. Recurrences are usually associated with incomplete removal. We present a 3-month-old infant with a history of stridor and respiratory distress caused by a firm 0.4cm wide and 1.4cm long mass arising in the supraglottic region and detected with endoscopic approach.

The lesion was excised endoscopically with an uneventful postoperative course and an excellent long-term prognosis. Histopathologically the lesion consisted of mature tissues with abnormal growth and disorganized architecture, chiefly composed of mature glandular structures, smooth muscular fibers, mature fat, surrounded by fibrous stroma and covered by typical squamous epithelium.

The aim of our report is to underline how this condition must be considered by physicians, paediatricians and anaesthetists as an important cause of airway obstruction.

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