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Case Report

Teratoma of larynx: case report

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ABSTRACT

Teratomas are embryonal neoplasm arises from totipotent germ cells. They are having tissues from all the three blastodermic layers. There are various locations of congenital teratoma. Naso-oropharyngeal site teratoma are either sessile or pedunculated. We describe a rare case of laryngeal teratoma in a five years old patient presented with change in voice and breathing difficulty. On flexible laryngoscopy, it appeared like supraglottic cyst but on CT scan it was confirmed as teratoma. Pre-operative tracheostomy and transoral carbon dioxide laser assisted excision done. Histopathological examination showed osteoid trabeculae, chondroid tissue with loose myxoid islands and adipose tissue. No recurrence of tumor on 18 months follow-up. This is the first case report of pediatric larynx teratoma reported in present century.

Keywords: Teratoma, Head Neck, Larynx

INTRODUCTION

Teratomas are benign neoplasms composed of multiple tissues that are foreign to the part of the body they arise from.[1] The term teratoma arises from the Greek word 'teraton' or monster, and its highest form consist of duplicated foetal parts. The overall incidence of teratoma is 1 in 4000 births with no gender predominance and is associated with an 18% risk of other congenital malformations.[2,3]

Commonest sites of the teratomas are sacrococcygeal region, gonads, and the mediastinum. Only 5% of all teratomas occur in the head and neck region, predominantly in the neck, nasopharynx, face, and orbit. Incidence of cervical teratoma ranges from 2.3 to 9.3 of the paediatric teratomas, roughly equating to 1 per 40,000 births.[4] Sign and symptoms of head and neck teratomas depend on their mass effects.

Contrast enhanced CT scan (CECT) of head neck remains main investigation to know the vascular involvement and content inside the tumor.

CASE REPORT

Five years old boy presented with hoarseness of voice for more than two years and noisy breathing during sleep for past 4 months. The child's father complained about very noisy breathing with chest retraction during sleep. The patient first underwent flexible nasopharyngo-laryngoscopy and followed by a CECT neck. After the probable diagnosis of teratoma larynx on CECT, the child was further investigated for planned surgery under general anesthesia. Other specific investigations like thyroid function tests and alfa fetoprotein were also been done. Consent for the tracheostomy was taken and surgery was done under general anesthesia.

Planned tracheostomy was done under total intravenous anesthesia with HHFNC (Heated high flow nasal cannula). Anesthesia was delivered through the tracheostomy and transoral laser (carbon dioxide- Lumenis Acublade-Tel Aviv) assisted excision was done. Tumour removed en block, a remnant of the mucosal flap of aryepiglottic fold sutured by 4-0 round body vicryl. Post-operatively child

was kept on nasogastric feeding for two days and allowed a semisolid oral diet thereafter.

Planned decannulation is done after the 12th post-operative day. The patient was followed in the outpatient department with flexible laryngoscopy at three, six, and twelve months period. On flexible laryngoscopy, a smooth mass arising from right aryepiglottic fold occupying right hemilarynx covering the right side of the true cord and partly in right pyriform sinus (Figure 1). It resembled a huge supraglottic cyst. CECT neck revealed an oval well defined heterogeneous density lesion of the size of 2.2 cm craniocaudal×2.2 cm transverse×2.1 cm anteroposterior in the right side of the hypopharynx (Figure 2). The mass shown with internal fatty density, bony density and soft tissue density areas. The lesion caused narrowing of the hypopharyngeal/supraglottic airway.

Pre-operative biopsy or fine-needle aspiration cytology (FNAC) was not done due to compromised airway, age of the child, site, and smoothness of the tumor mass. Tracheostomy helped in good exposure, better hemostatic control, and easier excision using CO_2 laser (Figure 3 and 4).

Mass was firm to hard with well-defined borders. It was excised en block and sent for histopathology. Patient was regularly followed for 18 months with no recurrence (Figure 5).

Post-operative histopathology showed a circumscribed mass displaying several small irregular lobules of preserved osteoid trabeculae merging abruptly with small to large globules of cellular chondroid tissue.

The rest of intervening stroma showed dense fibrosis with loose myxoid islands and large lobules of adipose tissue. Areas were displaying smooth muscle bundles. No epithelial cell origin was demonstrable (Figure 6 and 7).

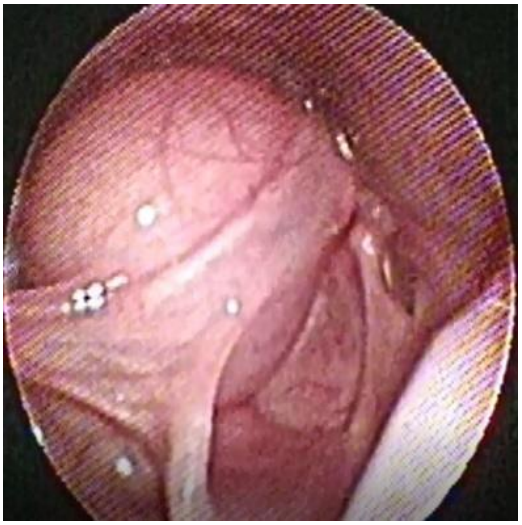

Figure 1: Endoscopic view-smooth mass from right aryepiglottic fold.

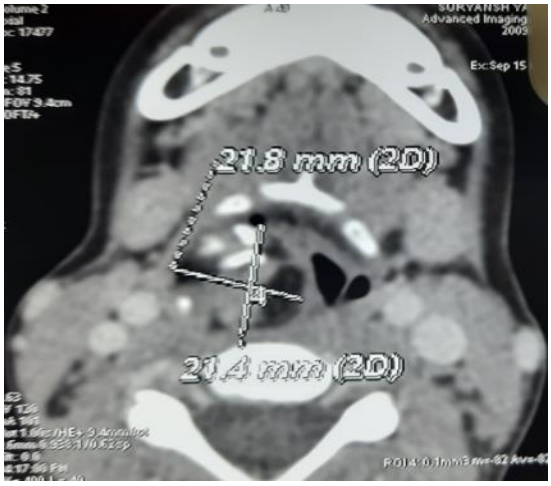

Figure 2: CECT- well defined heterogenous density lesion, size of 2.2cm craniocaudal×2.2 cm transverse×2.1 cm.

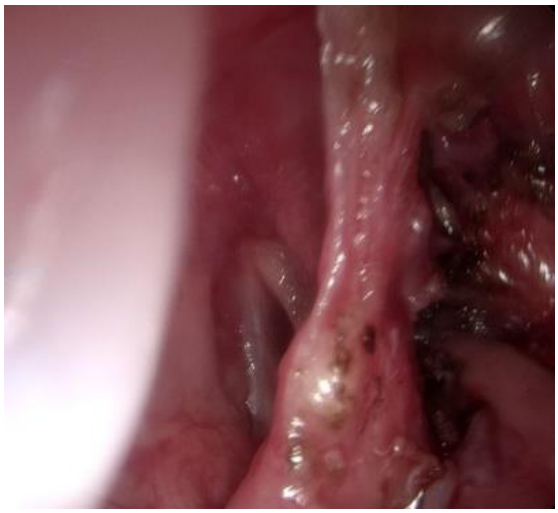

Figure 3: Per-operative tumour laser assisted excision.

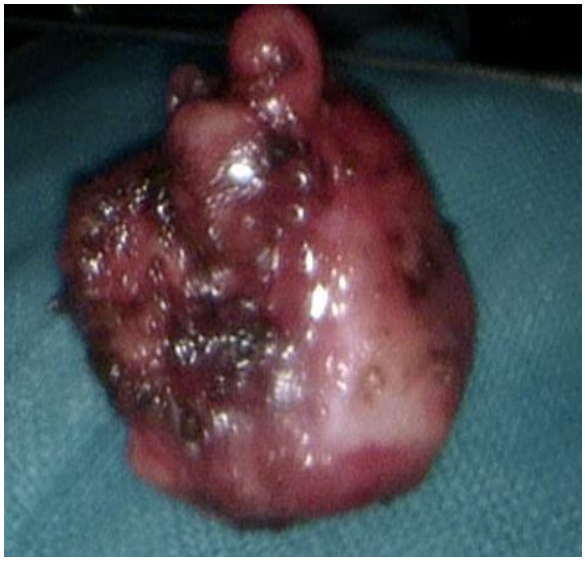

Figure 4: En-block excision of tumour.

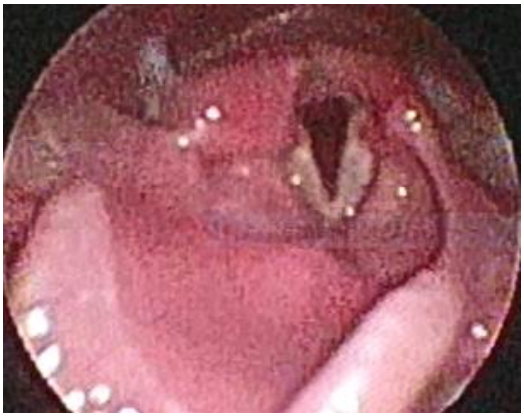

Figure 5: Post-operative endoscopy after 1 year.

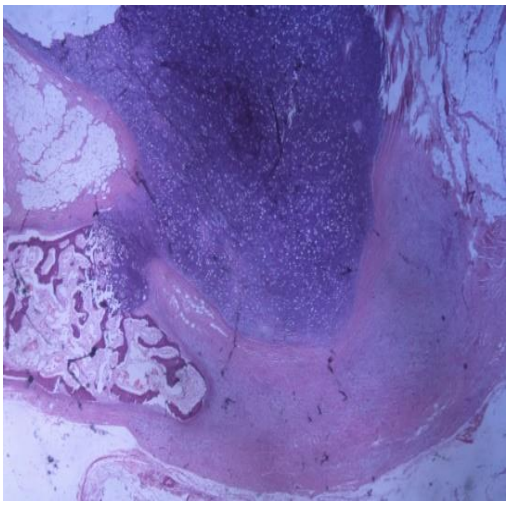

Figure 6: HPE-adipose chondroidosteoid (20X).

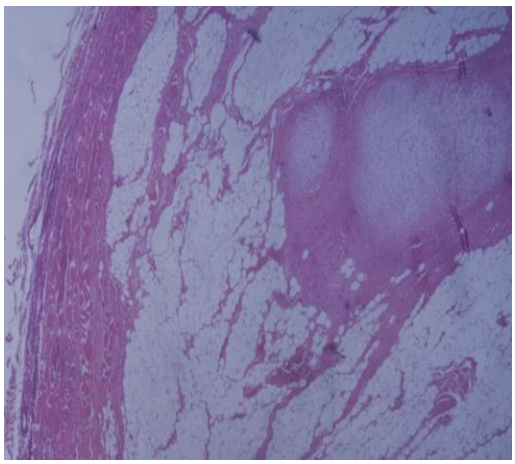

Figure 7: Adipose rich (20X).

DISCUSSION

Teratomas are embryonal neoplasms that arise when totipotent germ cells escape the developmental control of primary organizers and give rise to more or less organoid masses in which tissues are derived from all three blastodermic layers (ectoderm, endoderm, and mesoderm) can be identified. In literature there only a few anecdotal reports of teratoma of the larynx.

Albert E Bulson (1925) first reported throat teratoma in a newly born who was unable to breathe and had an occasional paroxysm of suffocation. Respiratory distress was relieved by holding the baby upside down and patting it on the back. A pedunculated mass was removed by snare and sent for histopathology. Histopathology showed natural skin with its normal layers with connective tissue in its center.[5]

The earliest description of a teratoma larynx was reported by Johnson and Young (1953) in a 50-years old white man who presented with difficulty in breathing for a decade.[6] Mirror examination showed a smooth swelling arising from the right false cord and aryepiglottic fold. The tumor was excised by an external approach after elective tracheostomy. Gross appearance of the tumor resembled a fibrolipoma but histopathology of tumor revealed an admixture of fat cells, fibrous tissue, islands of glandular tissue with cystic spaces. Scattered smooth muscle bundles and few areas of osteoid tissue. Clinical course, site, and histopathology resembled the present case except for the age of presentation and endoscopic surgical management.

There was another case report of immature teratoma of the larynx by Cannon et al (1987) in 32-years old woman presented with 3 months history of hoarseness of Voice.[7] Histopathology from a papillary lesion of the vocal fold showed primitive cells, uniform nuclei, scant cytoplasm, irregular distribution of melanin and mucoid matrix resemble fetal cartilage.

Above two cases were reported in the adult age group, while the present case was less than 6-years old child.

Imaging is of paramount importance for pediatric supraglottic tumors to learn the content and vascularity of the tumor. Besides CECT, other radiologic investigations like plain X-ray Neck will show calcified areas within soft tissue shadow in case of teratoma. Ultrasonography in such cases will generate images with mixed echogenicity with multiloculated cystic or sold areas. Radiological differential diagnoses of teratoma are lymphangioma, venous malformations with phleboliths, dermoid, neurenteric cysts, Thornwalds cyst, and basal meningocele. Pre-operative thyroid function tests were done in present case as cervical teratomas frequently contain thyroid tissue within and post-surgical hypothyroidism can result.[8,9] Paediatric cervical teratomas do not turn malignant, however raised serum alfa fetoprotein levels are good markers for malignancy.

There are various locations of congenital teratomas. Cervical teratomas can present as stridor due to extrinsic compression. Nasopharyngeal teratoma is sessile or pedunculated and protrudes through the nose or oropharynx. A large oronasopharyngeal teratoma with disfiguring mass in the neonate is known as epignathus.

They are attached to palate/skull base with or without intracranial component.[10] Pediatric teratomas are diagnosed early due to symptoms like respiratory distress, facial disfigurement, and orbital involvement. Teratomas in children are often congenital and they rarely convert into malignancy.

Teratomas commonly are classified using the Gonzalez-Crussi grading system: 0 or mature (benign); 1 or immature, probably benign; 2 or immature, possibly malignant (cancerous); and 3 or frankly malignant.[11] Laryngeal teratoma in this study is of mature benign type (0-grade).

Teratomas are often associated with polyhydramnios and other congenital malformations. Antenatal diagnosis helps in proper planning for cesarean delivery and airway management in such cases. There was no significant antenatal history in the present case.

Other germ cell tumor which resembles teratoma are hamartoma, Choristoma, and dermoid. Choristoma, a form of a heterotopic tumor with masses of normal tissues is found in abnormal locations. Hamartoma is a disorganized overgrowth of tissues in their normal locations. Dermoid are also germ cell tumours with both ectoderm and mesodermal components.

Waklu (2000) reported seven cases of head neck teratomas in five years. These were two intraoral teratomas, both arising from the oropharynx without any history of breathing difficulty.[12]

CONCLUSION

There were only two case reports in the twentieth century on adult teratoma larynx. This is the first case report of Paediatric teratoma of the Larynx in the present century with its distinct supraglottic location and huge size. The diagnosis was confirmed by a CECT scan and postoperative histopathological examination. Surgical planning by first securing the airway and laser-assisted en-block excision was done.

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