

## RARE BRANCHIAL ARCH ANOMALIES

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### ABSTRACT

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#### AIM

Amongst the branchial arch anomalies third arch anomaly occurs rarely and more so the fourth arch anomalies. We present our experience with cases of rare branchial arch anomalies.

#### PATIENTS AND METHODS

From June 2006 to January 2016, cases having their external opening in the lower third of sternocleidomastoid muscle with the tract going through thyroid gland and directing to pyriform sinus (PFS) or cysts with internal opening in the PFS were studied.

#### RESULTS

No fourth arch anomaly was encountered. One cyst with internal opening which later on formed a fistula, three fistulae from beginning and two sinuses were encountered. The main stay of diagnosis was the fistula in the PFS and the tract lying posterior to the internal carotid artery. Simple excision technique with a small incision around the external opening was done. There was no recurrence.

#### CONCLUSION

Third arch fistula is not very rare as it was thought. Internal fistula is found in most of the cases. Though radiological investigations are helpful, fistulae can be diagnosed clinically and during operation. Extensive operation of the neck, mediastinum and pharynx is not required.

#### KEYWORDS

Third Branchial Fistula, Rare Branchial Fistulae, Neck Sinuses.

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**INTRODUCTION:** Cyst and sinuses of the neck are an important area in the paediatric surgical practice. Among the branchial anomalies second branchial arch anomalies encountered in more than 90% cases. First arch anomaly is next to that and third and fourth arch anomalies occur rarely.<sup>1</sup> Second arch anomalies can be managed relatively easily. Rare branchial arch anomalies may present in unusual way and often difficult to manage unless the treating surgeon has the proper knowledge of this pathology. The classical description of third and fourth branchial cleft remnants have been theorized that the tracts take an extensive course behind the carotid artery and open in pyriform sinuses. Third arch remnant rarely and fourth arch remnant never take the theoretically described course. These may take a variable course and can be classified on the basis of its internal opening in pyriform sinus (PFS) or approaching towards it.<sup>2</sup> Moreover instead of going through the aortic bifurcations these go to PFS lying posterior to the internal carotid artery.<sup>3</sup> The cases with third arch anomalies encountered by us have been analysed here.

**MATERIAL AND METHODS:** This is a retrospective analysis of patients diagnosed to have the sinuses, fistulae and cysts of third arch origin. We have not encountered any classical fourth arch anomaly. Patients with external opening in the lower third of sternocleidomastoid muscle with the tract going through thyroid gland and directing to pyriform sinus (PFS) or cysts with internal opening in the PFS is included in the series. These cases did not traverse through the carotid bifurcations as found in the second arch remnants. Indeterminate cases and first and second arch anomalies are not included in the series. Study period was from June 2006 to January 2016.

Contrast X-ray was done in three patients and MRI scan was done in one patient. While patients were under general anaesthesia, to evaluate the internal opening laryngoscopic examinations were done in three patients and upper GI video endoscopy was done in one patient.

All the patients were operated under general anaesthesia. At the beginning of the operation a thick nylon suture or a thin feeding tube was inserted through the external opening. Excision of the tract was done by making a small incision around the neck opening and dissection completed staying close to wall. No effort is made to look the surrounding structures. Closure was done in layers without trying to close the pharyngeal mucosa.

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**RESULTS AND OBSERVATIONS:** During this nine year seven months period we encountered six cases of third arch anomalies. Out of these four patients were male and two were female. Age ranged from 1½ months to 6yrs (Table 1).

Cases	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Age at presentation	6m	1yr	6yr	3yr	1½ m	11m

**Table 1: Showing age distributions of the patients at presentation**

Presenting features in five patients were the presence of an opening in the neck since birth. The locations of the external openings were in the lower third of anterior aspect

Cases	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
External opening	Lower 1/3	Supra clavicular	Lower 1/3	Just above the clavicle	Just above the clavicle	Middle after its rupture
Discharges	Milk	Serous	Serous	Sweat like	Mucoid	Whitish initially, mucoid later on.

**Table 2: Locations of external openings and their discharges**

All the branchial fistulae were on the left side.

Out of the three cases where contrast X-ray of the tract was done, two of them showed fistulous opening to PFS (Figure 1). Other one was blind ending. MRI scan was done in one case who presented with a neck swelling (Figure 2). In four cases laryngoscopic examination was done after intubation to determine any visible internal opening. In two cases laryngoscopy was repeated at the end of the dissection of the tract. Pharyngeal walls were found to be indented at left PFSs on pulling the tract. But there were no fistulous communication present with the pharynxes. Fiberoptic upper GI endoscope was used in one case (Figure 3). A case where tract was blind ending and of short length neither endoscopy nor contrast x-ray was done. The diagnosis was made by supra clavicular location of its external opening, location of the tract behind the internal carotid artery and its extension up to the thyroid gland.

Under general anaesthesia probing of the tract was done from the neck (Figure 4) and a thick nylon thread was kept in it. Excision was done staying close to the tract (Figure 5). Wound closed in layers. Pharyngeal mucosa was not closed. Histopathological examinations of the excised tissues were done. Most of the tissues showed pseudo stratified columnar epithelium lining with lymphocytic infiltration on the wall (Figure 6). Two cases had hyperplastic stratified squamous epithelium with lymphocytic infiltration on the wall (Figure 7). Out of these one had metaplastic squamous stratified epithelium with some exocrine mucus secreting gland on the wall (Figure 8).

Oral feeding was allowed in all the cases after the patient fully recovered from the anaesthesia.

Patients were discharged on second post-operative day. On follow-up on seventh day the wounds were inspected and dressings were removed. Skin stitches removed in two cases where non absorbable sutures were used. There were no recurrences or infections. On subsequent follow up at six

of the sternocleidomastoid muscles (SCM). Three cases had this very close to the medial aspect of clavicle. The sixth case developed the opening later on and had it in the middle third of SCM (Table 2). The discharges from the openings were as shown in table 2. One patient drained milk on feeding. Case no 6 presented as an OPD patient with a swelling on the left side of the neck at 1½ months of age. On palpation there was crackling sound on it. The baby was brought again with stridor and mild respiratory distress. Symptoms were relieved after aspiration of whitish liquid from the cyst. The patient went home but the swelling recurred and ruptured after one month or so. The discharge was whitish liquid initially and later on it became clear mucoid.

weeks and in some later on also none had any problem and scars were nominal.



**Fig. 1: Contrast fistulogram revealing communication and delineation of oesophagus**



**Fig. 2: MRI scan shows third branchial cyst with gas fluid level**



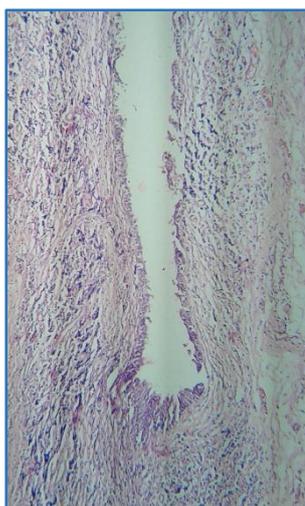
**Fig. 3:** Tract with inner opening in the left PFS



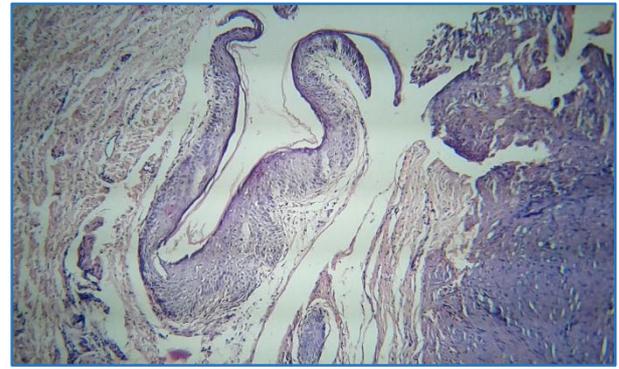
**Fig. 4:** On probing tip of the bougie came out from the mouth, indicating a relatively straight tract



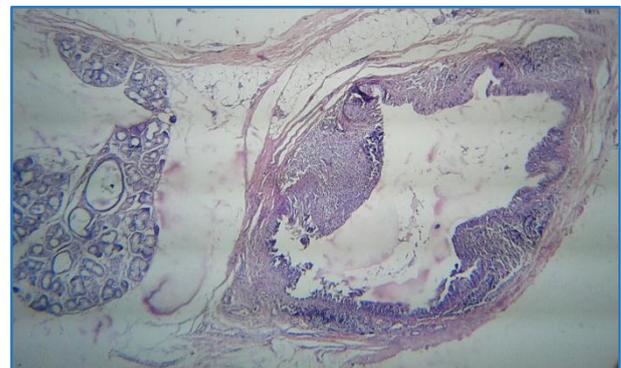
**Fig. 5:** Fistula dissected up to the pharyngeal wall



**Fig. 6:** HPE shows pseudo stratified columnar epithelium with lymphocytic infiltration on the wall



**Fig. 7:** HPE shows hyperplastic stratified squamous epithelium with lymphocytic infiltration on the wall



**Fig. 8:** HPE shows metaplastic squamous stratified epithelium with some exocrine mucus secreting gland on the wall

**DISCUSSION:** Embryologically, third and fourth arch anomalies have close association with developmental changes of aortic arches. Hence these fistulae course posterior to internal carotid artery rather than through the bifurcation of carotids. Complete fourth branchial fistula has not been demonstrated clinically. Theoretically its courses differ on two sides. On the left it should go to superior mediastinum and arch around the aortic arch to reach the PFS. But on the right it should remain on the neck and arch around the right subclavian artery.<sup>3</sup> Other workers have also supported this view and opined that instead of the theoretical long tract, these fistulae take a straight course to PFS.<sup>2</sup>

As in our series some workers also observed the incidence of these fistulae exclusively on the left side.<sup>2,4,5</sup> Whereas some others have found few cases on the right side, but in most of these series left side outnumbered the right side.<sup>6,7,8</sup> Only in one series out of five cases four were on the right side.<sup>9</sup>

Common presenting feature in the fistulae were the presence of an opening on the anterior aspect of the SCM muscle since birth. Some are in the opinion that these can be anywhere of the anterior aspect of SCM.<sup>3</sup> Mucoïd discharge or milk discharge on sucking is also encountered. Inflammation of left lobe of thyroid gland was encountered in many series.<sup>2,4,10</sup> Cervical mass was the symptom in those case with cyst. Where infected cyst was encountered commonly.<sup>4,8,11</sup> As in one of our case Liberman and

colleague.<sup>4</sup> Encountered a newborn who presented with respiratory distress.

In our series we had four internal fistulae out of six cases. Liberman et al<sup>4</sup> found two out of seven.

Apart from the clinical finding, laryngoscopy, ultra sound scan, CT scan, MRI scan along with contrast fistulogram were the investigations done in most of the series.<sup>4,11</sup> Ueda et al<sup>9</sup> claims that Barium swallow x-ray can detect the fistula. We relied mostly on the clinical and operative finding. Contrast fistulogram was done in three patients and MRI scan in one patient. Out of four patients Pereira et al<sup>12</sup> diagnosed all cases by laryngoscopy only.

Though we did not get any, thyroid tissue attached to the excised tissue was encountered in HPE in some cases.<sup>4,13</sup>

Most of the authors are in opinion that simple excision of the tract by defining adjoining structures is the required treatment.<sup>3,7</sup> Liberman M et al<sup>4</sup> have advocated excision of left lobe of thyroid gland along with the tract. Whereas Pereira et al<sup>12</sup> described an operative technique of oblique thyrotomy above the cricothyroid joint after detaching the inferior constrictor and have claimed good result in it. We employed simple excision of the tract with a small incision around the external opening. Only in one case we had to make a bigger incision to define the surrounding structures. Some others have advocated excision starting from pharyngeal side and removal of the posterior border of the thyroid ala to prevent recurrence.<sup>6</sup> Cibliano et al<sup>5</sup> have advocated in injecting Fibrin glue to obliterate the fistula.

In our series there was no recurrence. But in some series recurrences were found and re-explorations were required.<sup>4</sup> Lin et al had advocated extensive dissection of the tract especially to the pharyngeal wall to prevent its recurrence.<sup>14</sup>

**CONCLUSION:** Rare branchial arch anomalies especially 3rd arch anomalies are not very rare.

They do not take courses described theoretically basing on its embryogenesis. But they may have a variable course. Treatment does not require an extensive dissection demonstrating the vital neck structures. It can be done with a small incision around the external opening and dissecting close to the tract as done in second arch fistulae.

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