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Conservative management as a viable alternative in the management of paediatric first branchial arch fistula: a case series

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Abstract

Objective

First branchial arch abnormalities are rare. Surgical excision is the mainstay of treatment and described in the literature. Excision can be associated with significant complications. We describe factors influencing operative and non-operative management of patients.

Methods

Case review was conducted between January 2012- September 2022 of patients with first brachial arch abnormalities at Alder Hey Children's Hospital, UK. Analysis of electronic patient records, operation notes and extraction of clinical outcomes were obtained.

Results

4 patients were identified with an average age of 2 years and 4 months. 3 were female. 3 underwent operative intervention, one of which had a recurrence postoperatively and was manged conservatively. The non-operatively managed patient remains conservatively managed with no complications.

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Conclusion

First brachial arch abnormalities can be managed operatively and non-operatively depending on patient factors. Steps to improve surgical planning such as CISS MRI may help decision making and risk stratification of operative management.

MeSH keywords:

Branchial Clefts, Branchial Arch, Branchial Cleft Cyst, Conservative Treatment, Facial Nerve Injuries

Introduction

First branchial arch/cleft abnormalities are rare congenital abnormalities with an estimated incidence of approximately 1 case per 1 million people [1]. First arch abnormalities account for 1-10% of all branchial abnormalities [2, 3]. These involve the development of an abnormal cyst, sinus, cartilaginous remnant, or fistula between the skin just inferior to the mandible or in the preauricular region. These pass through the parotid region and opening in and around the external auditory meatus [3].

Symptoms associated with first branchial arch fistulas may include recurrent infections, mucopurulent discharge from openings, pain or painless swelling [4]. Treatment typically involves surgical excision of the tract to prevent recurrence with all literature reporting excision once diagnosis is made [4 - 10]. However, due to the serious potential for concomitant facial nerve paralysis, surgery can be challenging and requires meticulous planning. This can often be challenging given the difficulties in locating the extracranial facial nerve on MRI imaging in children[11]. Some patients may benefit from more conservative management when different surgical and patient circumstances are considered.

We present 10 years of experience managing first brachial arch abnormalities describing both successfully surgically managed and non-surgically managed paediatric patients in-order to highlight the different management strategies and when these may be used, in contrast with previously published literature.

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Materials and Methods

Clinical records were retrospectively reviewed from Alder Hey Children's Hospital, a large UK paediatric tertiary ear nose and throat (ENT) centre from January 2012 until September 2022. All patients with the ICD-10 code Q-180, Q181 or Q182 "Sinus, fistula and cyst of brachial cleft" were identified and included.

Patient records and imaging were reviewed and only patients with confirmed first brachial arch abnormalities were included. Patient demographics, histories, examinations, investigations, and intervention data were extracted and included. Management is discussed.

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Results and Analysis

157 patients were identified meeting the search criteria, with a total of 4 patients meeting inclusion criteria. 153 patients were excluded as they had diagnoses other than first brachial arch abnormalities such as preauricular sinuses or other branchial arch abnormalities. All remaining patients had a first brachial arch fistula.

The average age of first presentation to ENT services was 2 years 4 months (10 months, 1 year 2months, 5 years 5 months, and 1 year 11 months). 3 (75%) were female, all abnormalities were on the left side. 3 patients underwent operative management at some point, 1 managed conservatively. 1 patient who was operatively managed had an initial operative excision by another team and had recurrence which was managed conservatively subsequently [Table 1].

Patient A:

A 1 year 11-month-old male patient presented to ENT outpatients with a left sided neck swelling since birth with oily discharge. There was no history of infection, purulent discharge erythema. The child was otherwise healthy. Examination findings showed 2cm firm swelling at the angle of the mandible with discharge, but no infective signs.

A sebaceous cyst was suspected. Ultrasound imaging of the lump showing the opening had a 4mm tract leading a complex cyst at the inferior lower pole of the

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parotid gland and extending anteriorly toward the skin. Subsequent MRI scanning showed the tract from the mandibular lump passing posterior-superiorly into the deep lobe of the parotid and terminated in the external auditory meatus. [Figure 1]

The patient was discussed in the local ENT radiology meeting and the child's parents elected for excision of the tract which occurred 8 months after initial presentation. Post-operatively the patient had no facial nerve deficits but had a wound infection which was treated with inpatient intravenous antibiotics and analgesia. They had no further recurrence and they were discharged just under 16 months years following excision.

Patient B:

A 5 year and 5 month old female patient presented to ENT outpatients with left sided swelling in the preauricular region and discharge. The child had a prior history of excision of a first brachial arch sinus under the paediatric general surgeons at age 2 years and 5 months. The previous operation identified and excised a tract but was seen to taper out before the parotid gland. They developed swelling and discharge over the same left site 6 months later. The reason for delayed re-presentation was not known. Examination identified they had a thickened swelling under the angle of the left mandible. Recurrence of a first branchial arch abnormality was suspected.

Ultrasound showed a 2mm tract extending from the mandibular swelling passing superiorly across the parotid toward the external auditory canal. MRI showed a tract with 5x10mm expansion lateral to the parotid extending across and deep to

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the parotid. The distal segment lay immediately inferior and parallel to the left external acoustic canal. The location of the abnormality was close to the usual tract of the facial nerve, however its relationship to the facial nerve could not be determined due to limitations of the MRI imaging [Figure 2].

Operative options were discussed with the parents including the option of total parotidectomy for complete excision of the tract and significant subsequent risk of facial nerve damage. The parents elected for nonoperative treatment.

The patient was discharged to local surveillance 3 years 5 months after initial excision.

Patient C:

A 1 year and 2 month old female patient presented to ENT outpatients with a history of two recurrently discharging left sided puncta. One punctum was located in the preauricular area and the other on the left ear lobe. Examination showed no signs of active infection.

Ultrasound showed a 3mm tract between the two puncta passing inferior to the opening of the external auditory canal and which did not pass through the parotid parenchyma. The lobular element showed a 3x7mm expansion. MRI scanning demonstrated a serpiginous cystic space extending from the left earlobe passing deep coursing beneath the external auditory meatus to preauricular region. [Figure 3]

At this point, the patient had ongoing discharge and therefore the decision was made to proceed with excision. This was completed 2 years and 2 months following

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initial presentation and histology confirmed a first brachial cleft fistula. Post operatively the patient had no complications or recurrence and was discharged 8 months after excision.

Patient D:

A 10 month old female patient presented to ENT outpatients with a history of pit in the left mandibular region with thick discharge since birth. There was no prior history of infection. Examination showed no signs of infection, but thick discharge was expressed. No external auditory canal opening was found on otoscopic examination.

Ultrasound scanning demonstrated a tract leading from the punctum to a 1cm cyst just deep to the ear lobe. MRI scanning demonstrated a tract from the posterior aspect of the cheek toward the parotid gland communicating with a 10 x 7 x 9mm cyst. The tract was found to extend medially passing interior to the external acoustic canal and tapered in the left para-pharyngeal soft tissue [Figure 4].

Management strategies were discussed with the parents and given the minimal symptom burden, risks of possible marginal mandibular nerve damage and potential need for parotidectomy, a watchful waiting strategy was used. They were last seen 4 years after initial presentation with no significant complications.

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Discussion

We present a series of 4 patient with differing management strategies. Within the literature, the mainstay of treatment for first branchial arch anomalies is excision of the lesion [4 - 10]. In these studies 414 patients are described with 0-11% and 0-9% facial nerve palsy and recurrence rates respectively [4 - 10]. There is no previously published non-surgical management strategies for first branchial arch anomalies in children. We present the first case series to offer conservative alternative management strategies for these children to avoid high risk complications which can result in significant long-term morbidity. The children presented in this series demonstrate a range of clinical manifestations, aberrant anatomy and clinical journeys with a tailored management approach.

In this series, patient D had very little troublesome symptoms whereas patient C suffered recurrent infections with concomitant morbidity. In the former patient, undertaking an operation with associated significant morbidity such as facial nerve paralysis would be difficult to justify with low symptom burden. In such cases, conservative measures, namely, close observation for changes in symptoms, antibiotic therapy with microbiology support, wound care including water precautions, and analgesia will help optimise care. Counselling parents is key to aid them in recognising signs of infection and when to seek medical attention should complications arise.

Further to this point, patient B had more significant symptomology than patient D, however from the result of the MRI scan, this patient presented greater operative challenges and therefore a higher risk of complication.

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Identification of relational anatomy pre-operatively is incredibly important in stratifying risk. Identifying the course of the extracranial facial nerve on imaging is challenging making surgical risk predication harder. Conventional MRI sequencing was used in the assessment of the patients in this series which has a limited ability to track then course of the extracranial facial nerve. Constructive Interference in Steady State (CISS) T2 MRI sequencing may offer superior facial nerve imaging. CISS MRI has been used to investigate and map the facial nerve in adults and has been described in the detection of paediatric facial nerve aplasia and Moebius syndrome [12, 13]. Its use in imaging the extracranial course is of particular interest in first branchial arch abnormalities given the often-close relationship of tracts with the facial nerve. In adults CISS sequences show potential in fine visualisation of the extracranial course and are superior to double-echo steady state with water excitation sequences [14, 15]. CISS MRI sequency for facial nerve identification has not been described in the literature for surgical planning of paediatric brachial arch abnormalities. This provides an opportunity for utilisation in the planning of excisions and in the risk stratification for patient and parents, however this is yet to be investigated

Research covering larger patient groups needs to be collected to better understand the long term outcomes of conservative management. Additionally, the role of CISS MRI in first branchial arch abnormalities should be explored with primary research and its impact on operative management.

Ultimately, clinicians need to support discussions with parents over the benefits and risks of each management strategy to allow tailored management decisions for

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each individualised case. This will provide superior outcomes whilst minimising potentially life changing morbidity.

Summary:

- First brachial arch abnormalities are rare causing pain, swelling, discharge and recurrently get infected.
- Surgical excision is the only described technique in the literature which not without complication including facial nerve palsy and recurrence.
- This series adds that some patients may be more suitable for conservative measures when complication risk are higher.

Conclusion

First brachial arch abnormalities are rare and result in aberrant tracts in and around the external auditory canal and angle of the mandible. These present with a range of symptomology. Excision is the mainstay of treatment and has dominated the research literature. This case series proposes a unique alternative use of conservative measures in selected patients. Patient and parent counselling supported by specialist input is vital to optimise outcomes and minimise morbidity.

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Competing interests:

The authors declare none

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Figure 1

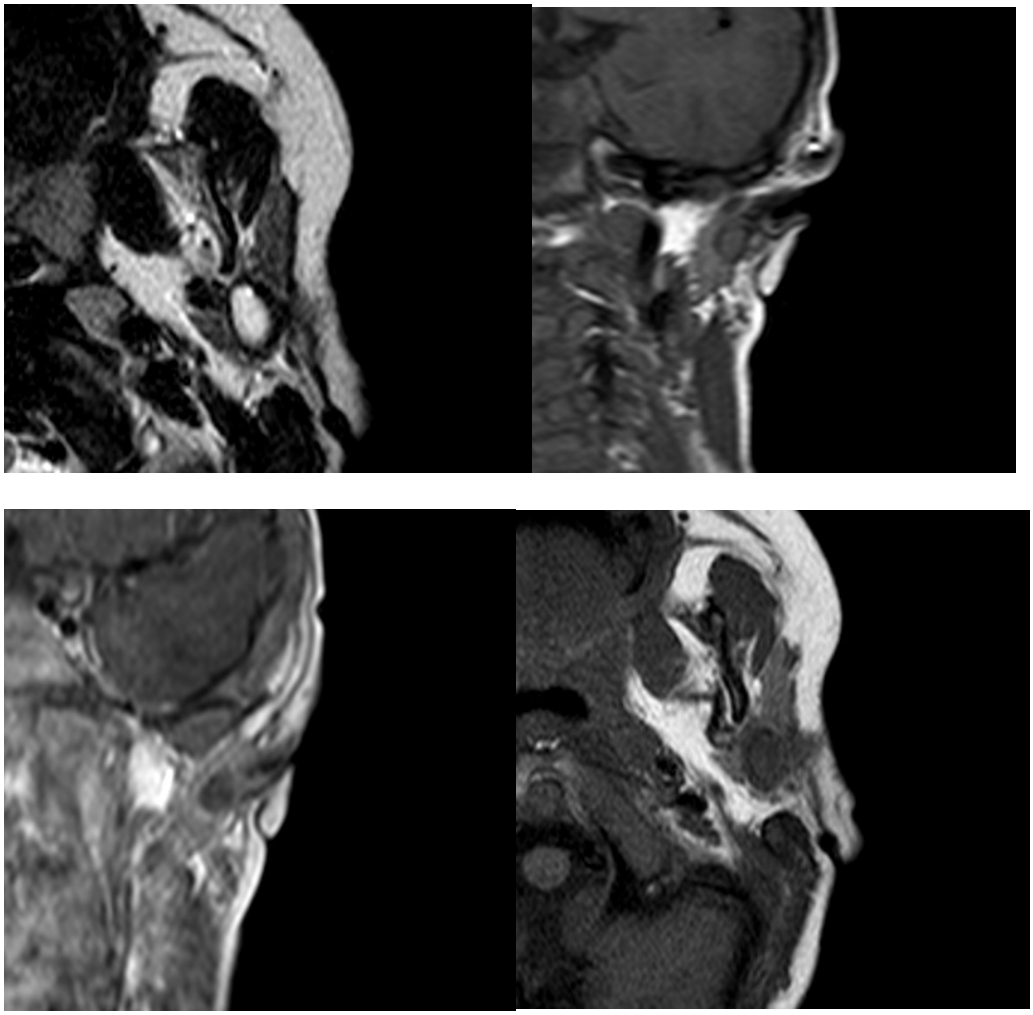

Figure 1: Axial T2W TSE (top left), coronal T1W TSE (top right), coronal T1 post gadolinium (bottom left), axial T1W TSE (bottom right) MRI images of patient A. MRI displays a tract (white arrow) passing posterior-superiorly into the deep lobe of the parotid and terminating in the external auditory meatus.

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Figure 2

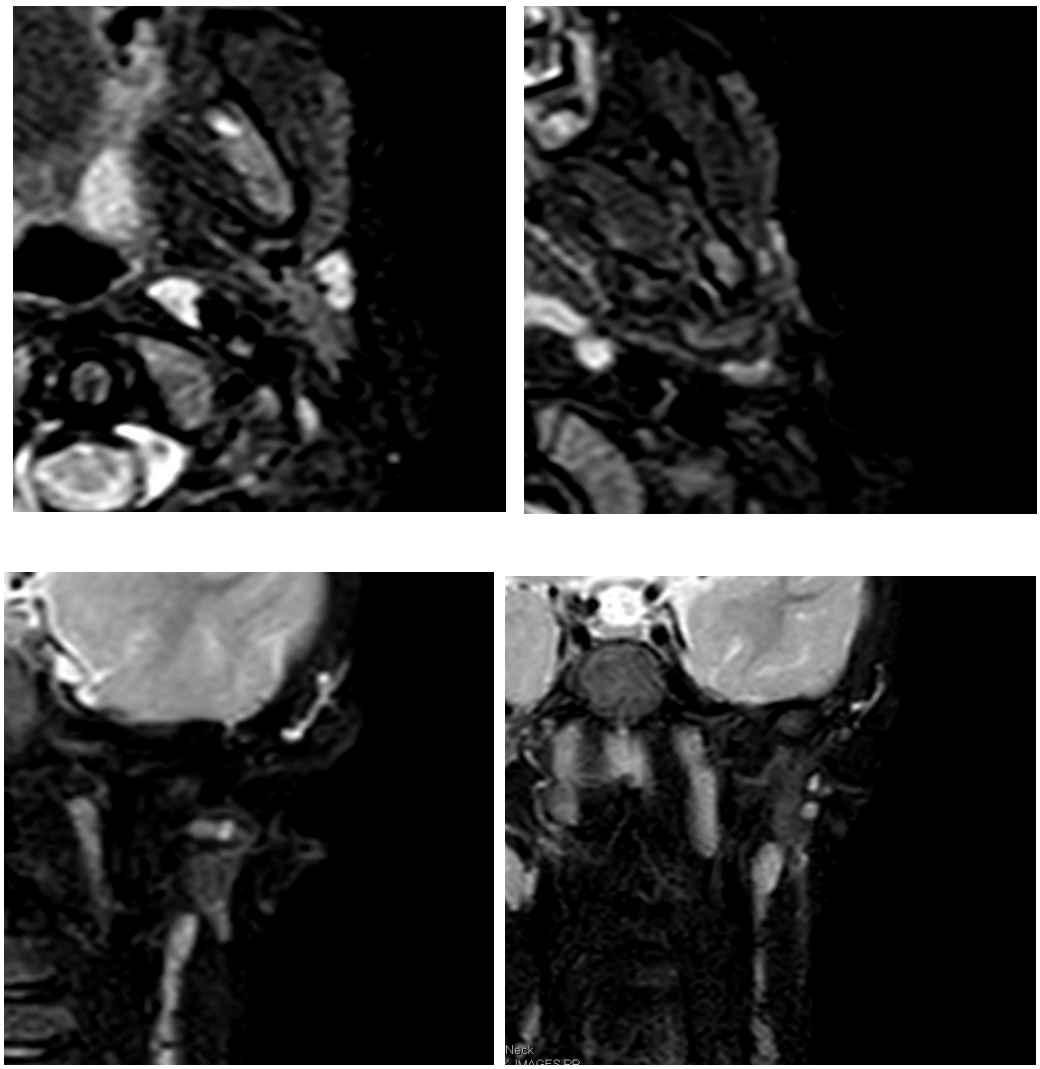

Figure 2: Axial STIR (top left and right) and coronal T2W TSE (bottom left and right) MRI images of patient B. MRI displays 5x10mm expansion (white arrow) lateral to the parotid extending across and deep to the parotid (white triangle). The distal segment lay immediately inferior and parallel to the left external acoustic canal.

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Figure 3

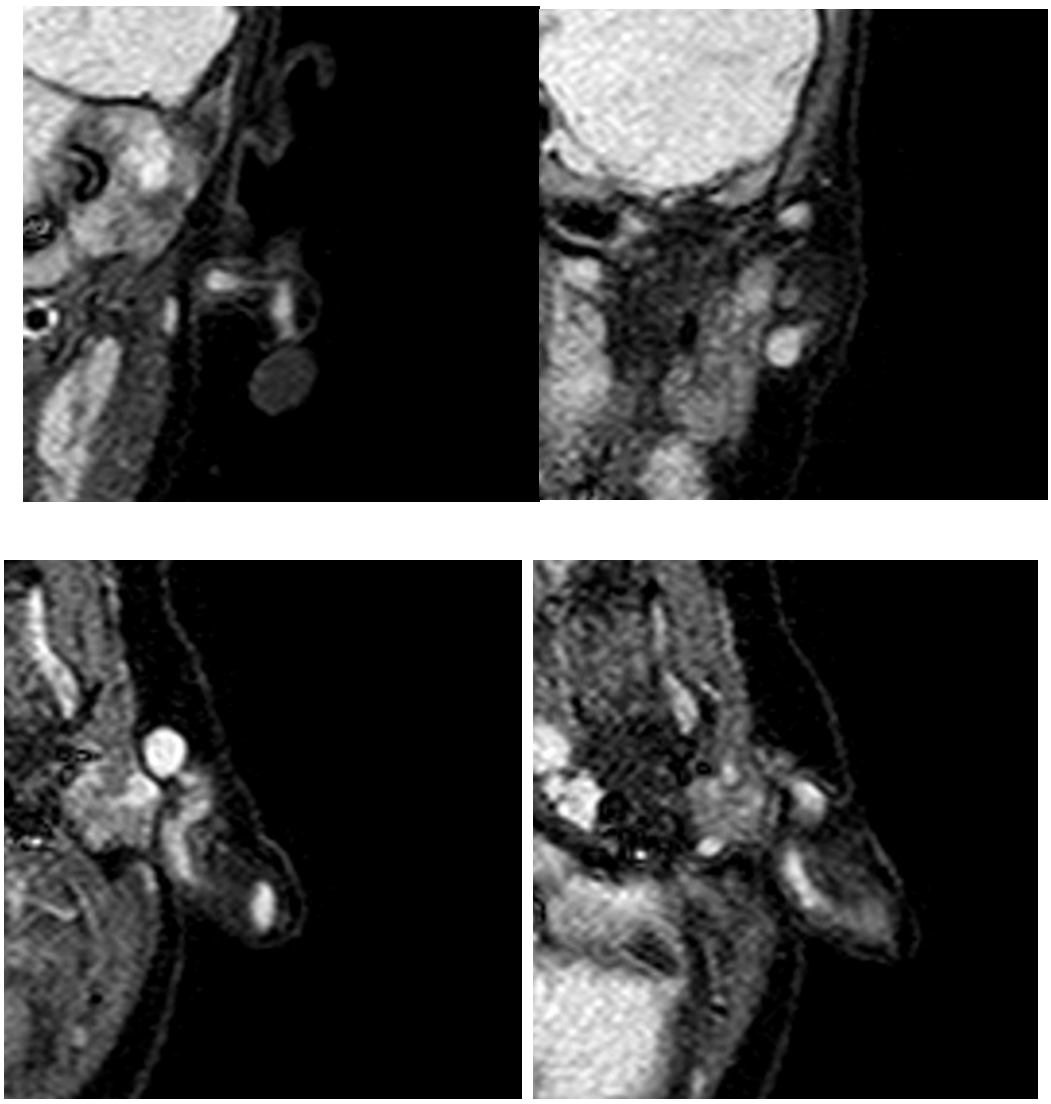

Figure 3: Coronal STIR (top left and right) and Axial STIR (bottom left and right) MRI images of patient C. MRI showed a serpiginous cystic space extending from the left earlobe (white arrow) passing deep coursing beneath the external auditory meatus to preauricular region (white triangle)

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Figure 4

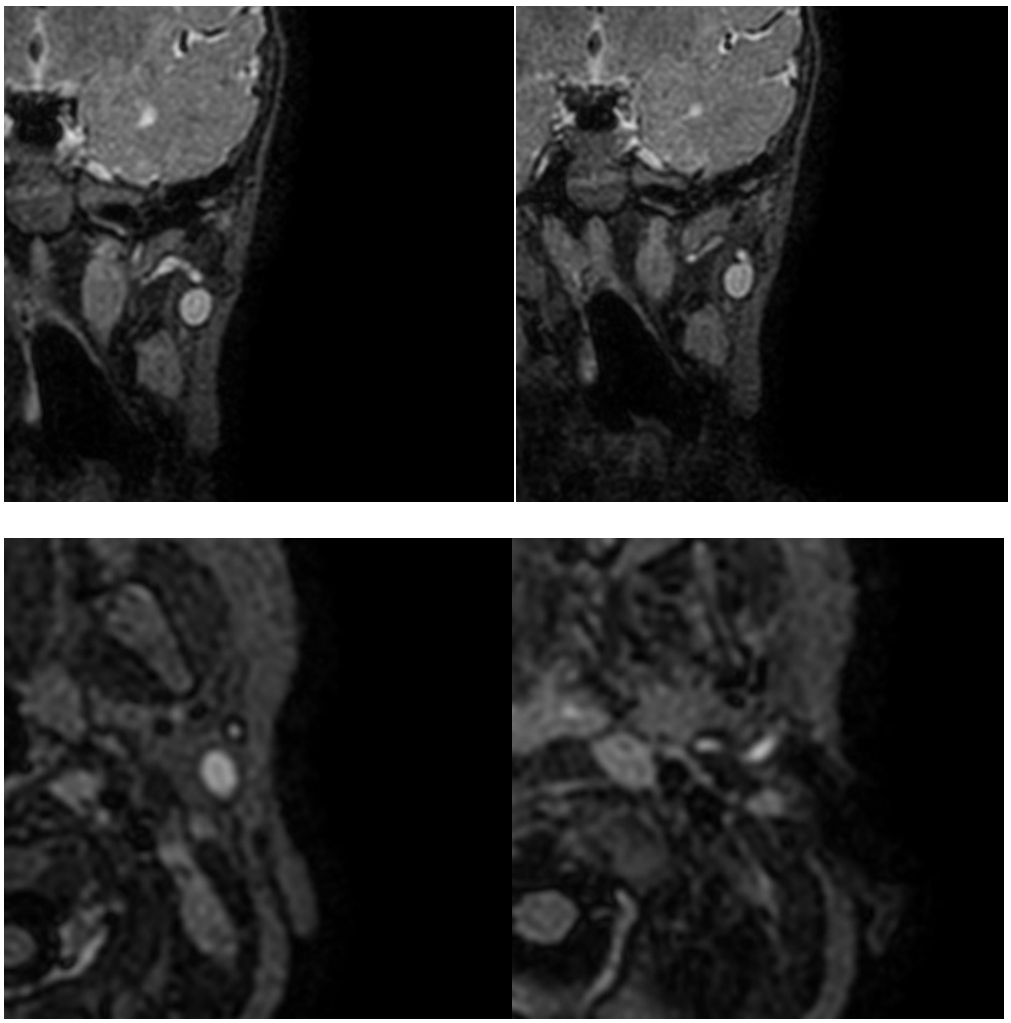

Figure 4: Coronal MPR (top left and right), Axial MPR (bottom left and right) MRI images for patient D. MRI showed a tract from the posterior aspect of the cheek toward the parotid gland communicating with a 10 x 7 x 9mm cyst (white arrow). Tract extended medially (white triangle) passing interior to the external acoustic canal and tapered in the left para-pharyngeal soft tissue.

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Figure 5

Table 1:

Patient Case	Age at presentation (Years, Months)	Sex	Side	Management
A	1,11	M	Left	Excision
B	5,5	F	Left	Excision + recurrence managed conservatively
C	1,2	F	Left	Excision
D	0,10	F	Left	Conservative

Table 1: Summary table of demographic details and general management principles of patients.

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