Classification of first branchial cleft anomalies: is it clinically relevant?

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Background **There are three classification systems for first branchial cleft anomalies currently in use. The Arnot, Work and Olsen classifications describe these lesions on the basis of morphology, tissue of origin and clinical appearance. However, the clinical relevance of these classifications is debated, as they may not be readily applicable in all cases and may provide no additional information on how the lesion should be managed.**

Objective **We seek to investigate this issue by applying these classification systems to cases from our centre and evaluating the information gained.**

Patients and methods **A retrospective case note review of all first branchial cleft anomalies excised at our institution between 2004 and 2014 was carried out, recording patient demographics, information on the anomalies and how they were investigated and managed.**

Results **This search identified eight unilateral cases and one bilateral case of first branchial cleft anomalies. These were a heterogenous group of lesions, which were variably investigated and managed. Categorization of these cases into Arnot, Work and Olsen subtypes did not correlate with the lesion's relation with the facial nerve or the outcome of excision.**

Conclusion **The current classification systems used for first branchial cleft anomalies have little clinical relevance apart from providing extensive descriptions to aid in diagnosis. We advise instead that clinicians use imaging techniques to gain as much information as possible about these lesions before excision and be aware of the risk to the facial nerve at the time of excision. A description of the lesion's relation with the facial nerve at the time of excision may provide more information on the likely outcomes compared with the classifications currently in use.** ***Ann Pediatr Surg*** **13:8–13 © 2017 Annals of Pediatric Surgery.**

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Introduction

Anomalies of the first branchial cleft are rare, accounting for less than 8% of all branchial anomalies [1,2], with an annual incidence of ~1/1 000 000 [3], and are more common in the female population compared with the male population [4]. These anomalies form because of abnormal development of the first branchial cleft, found between the first and second branchial arches during the fourth and fifth weeks of gestation.

First branchial cleft lesions arise specifically from the ventral portion of the first cleft [5], which like the other branchial clefts generally obliterates by week 8. However, the other constituent parts of the first cleft do persist, forming the external auditory meatus, cavum conchae and external tympanic membrane [5]. Therefore, a persisting anomaly of the ventral portion will develop in this region. Furthermore, the later development of the parotid gland and migration of the facial nerve to the same area mean that first branchial cleft anomalies have unpredictable relations with these structures. Because of the nature of their development first branchial cleft anomalies are a heterogeneous group of defects [6]. Consequently, these anomalies may present in different ways and are commonly misdiagnosed and inadequately managed [2,3,7].

Despite their heterogeneity, several classification systems have been proposed to categorize these anomalies.

The first classification was proposed by Arnot in 1971 [8], who described two types based on morphology:

(1) Type I includes a painful cyst or discharging sinus in the area of the parotid gland, which is often closely associated with the lower branches of the facial nerve and may extend deeply into the infratemporal fossa. It presents during early or middle adult life.
(2) Type II includes a sinus or superficial cyst in the anterior triangle of the neck, which may have an external opening below the angle of the mandible, and a track, which extends to and may communicate with the external auditory canal. There is a variable relationship with the facial nerve. It presents during infancy or early childhood.

Work, in 1972 [9], also described two types of first branchial cleft anomaly. However, this classification is based on tissue of origin:

(1) Type I are generally cysts of ectodermal origin, considered to be a duplication of the membranous external auditory canal. Classically, these occur medial to the concha and frequently extend to the postauricular crease, running superior to facial nerve. Histologically, they have a squamous epithelium lining.

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(2) Type II are cysts or sinuses of ectodermal and mesodermal origin, considered to be a duplication of the membranous external auditory canal and pinna. They commonly present with an abscess below the angle of the mandible, with a track running superiorly to the external auditory canal, which has a variable relationship with the facial nerve. Histologically, they contain squamous epithelium and cartilage.

Finally, Olsen in 1980 [10] proposed a simpler classification system based on the clinical presentation of the lesion:

(1) Cyst.
(2) Sinus.
(3) Fistula.

There is debate as to the clinical relevance of these classification systems [1,10–12]. Lesions may not fit the descriptions easily, and often anomalies are difficult to categorize based on examination or even imaging investigations. In particular, the Work classification can only be determined retrospectively [13] once the lesion has been excised and examined. Furthermore, the information ascertained may make no difference to the proposed management plan.

To assess the clinical relevance of the classification systems described, we investigated all cases of first branchial cleft anomalies dealt-with at our tertiary referral centre for Paediatric Otorhinolaryngology over the last decade.

Patients and methods

All first branchial cleft anomalies excised at the Royal Hospital for Sick Children, Glasgow, between 2004 and 2014 were sought using a computer-based theatre records system. A retrospective review of the relevant case notes sought information on patient demographics and nature of the lesion from clinical descriptions and imaging. Data on the management of the anomaly, including its relation with the facial nerve, histological findings and any complications, were also recorded.

Results

Nine patients (eight female, one male) who received treatment for first branchial cleft anomalies were identified during this time period. Eight patients had a unilateral first branchial cleft anomaly (four right-sided and four left-sided), and one patient had bilateral first branchial cleft anomalies.

The mean age at the time of excision was 5 years (range 1–13 years). Five patients had been managed conservatively in the past with oral antibiotics, and three patients had undergone previous incision and drainage of an abscess secondary to the anomaly.

Four patients underwent imaging in the form of ultrasound and MRI before excision. Excision of the lesion required the addition of superficial parotidectomy and dissection of the facial nerve in three cases. A facial nerve monitor was used intraoperatively in nine of the 10 operations, and there were no instances of postoperative facial nerve paralysis. Wound infection affected three patients, although none of the patients developed a recurrence of their first branchial cleft lesion.

Classification of the anomalies according to the Arnot, Work and Olsen systems is shown in Table 1 in comparison with the lesion's relationship with the facial nerve.

Cases

Case 1

A 1-year-old girl presented with a cystic swelling behind her left ear, connected to an intermittently discharging sinus in the left submental region.

MRI showed that this fluid collection extended from the postauricular region around the sternomastoid muscle to the submental sinus (Fig. 1).

This first branchial cleft anomaly was excised with a partial parotidectomy, showing that the lesion was inferior to and separate from the main trunk of the facial nerve. More anteriorly, the lesion was deep to the mandibular and cervical branches of the facial nerve (Fig. 2).

Case 2

A 2-year-old girl developed a cystic swelling behind the left ear, which became infected and discharged pus both from its surface and from a sinus at the inferior attachment of the left lobule, which had been present since birth.

On operation, this cyst and the associated sinus were found to be part of a duplication of the ear canal. The lesion was in superficial tissue planes; therefore, the surgeon was confident that the lesion was distant from the facial nerve and that formal dissection of the facial nerve was not required.

Case 3

A 1-year-old boy with a cyst in the left submandibular region, which became infected and discharged through a sinus below the mandible, was managed conservatively. After 3 years the cyst became infected again and surgical excision was planned.

At operation a tract was followed between the discharging sinus and a large cyst containing pus. This cyst extended from the external and internal carotid arteries to the tip of the styloid process. The facial nerve was not formally dissected, and no facial nerve monitor was used intraoperatively; however, given the course described, it can be inferred that the lesion ran deep to the trunk of CN VII.

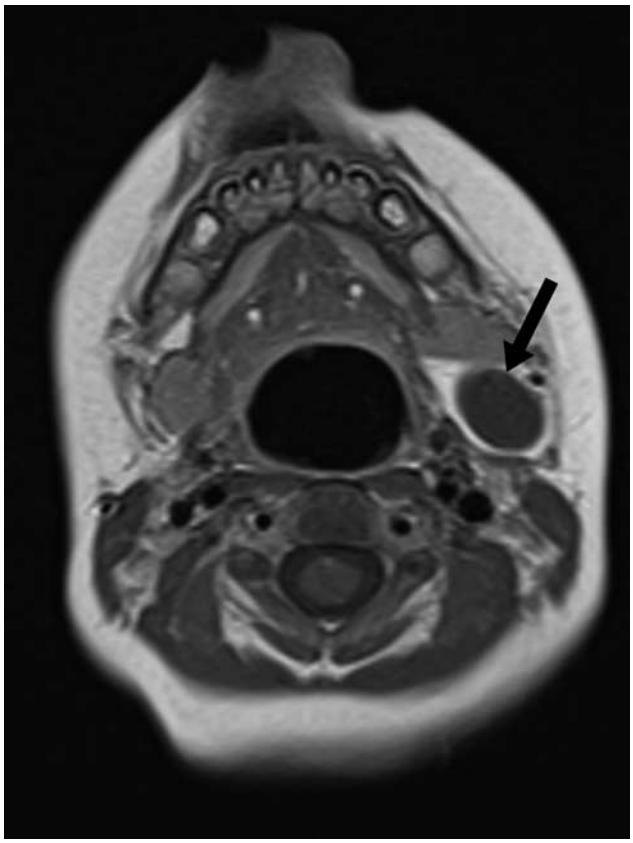
Case 4

A 4-year-old girl with a history of profound bilateral sensorineural hearing loss secondary to Pendred's syndrome presented with a recurrent swelling in the right preauricular area. This area had been incised and drained on one occasion, but otherwise had been managed with oral antibiotics.

Table 1 Describing first branchial cleft lesion(s) in each case, comparing Arnot, Work and Olsen subtypes to the relationship to the facial nerve at excision

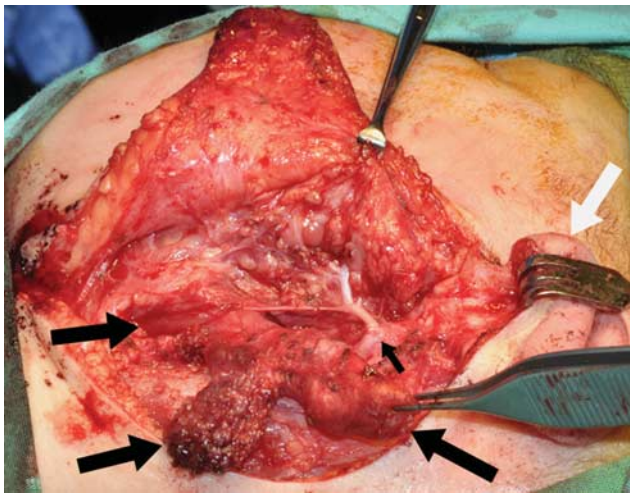
Case	Description	Arnot	Work	Olsen	Relationship with the facial nerve
1	Postauricular cyst, discharging sinus submental area	II	II	Sinus	Deep to mandibular and cervical branches
2	Postauricular cyst with communicating sinus at the inferior attachment of the lobule	I	II	Sinus	Distant
3	Cyst and sinus submandibular region	II	I	Sinus	Deep to trunk
4	Preauricular cyst	I	I	Cyst	Distant
5	Postauricular cyst, sinus on lobule	I	II	Sinus	Distant
6	Submandibular cyst and sinus	II	II	Sinus	Directly superficial to trunk, branches pass around lesion
7	Submandibular cyst and sinus	II	II	Sinus	Upper and lower divisions pass around lesion
8	Bilateral sinuses in external auditory canals	I	II	Sinus	Distant
		I	II	Sinus	Distant
9	Cyst within parotid gland	I	II	Cyst	Directly superficial to main trunk

Fig. 1



T1 axial image from case 1 showing left-sided first branchial cleft anomaly identified by a black arrow.

Fig. 2



Intraoperative image from excision of left branchial cleft anomaly in case 1. A white arrow indicates the position of the left ear for orientation, with right and left black arrows demonstrating the extent of the lesion and the black arrow in the centre indicating the parotid tail, which has been reflected away from the facial nerve, indicated by a smaller black arrow.

MRI showed a superficial lesion in the region of the right tragus, which was bulging into the superficial right external auditory meatus (Fig. 3).

This lesion was excised showing that it involved a tract, which extended from the tragal cartilage to the posterior aspect of the right ear canal and was distant from the facial nerve.

Case 5

A 4-year-old girl presented with a left postauricular abscess, which was found to connect to a sinus on the left lobule. This description was confirmed when the lesion was formally excised 1 month later, showing that it was distant from the facial nerve. Once again the superficial nature of the lesion meant that formal facial nerve dissection was not required.

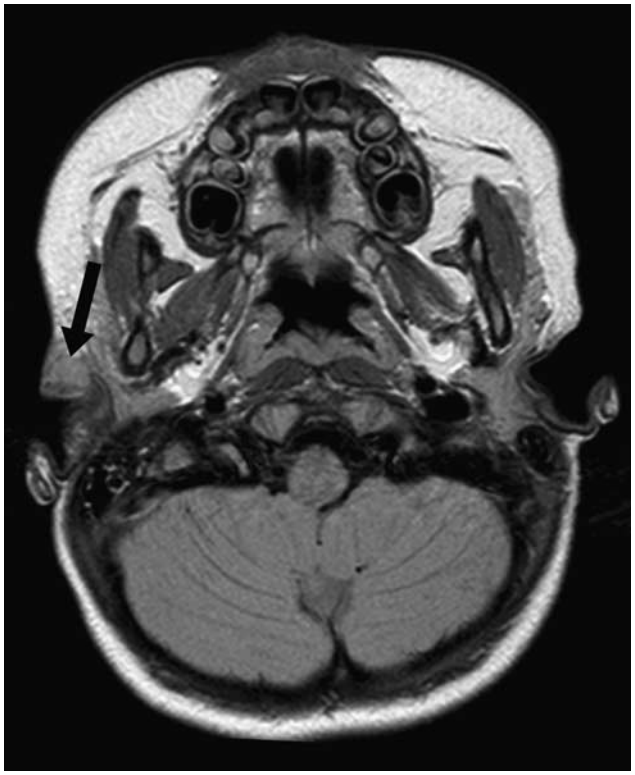
Postoperatively, she developed a wound infection, which settled with oral antibiotic therapy.

Case 6

A 5-year-old girl was known to have a sinus below the angle of the right mandible since birth, which had been intermittently discharging for 2 years. Excision was planned when an associated tender swelling developed deep to this sinus.

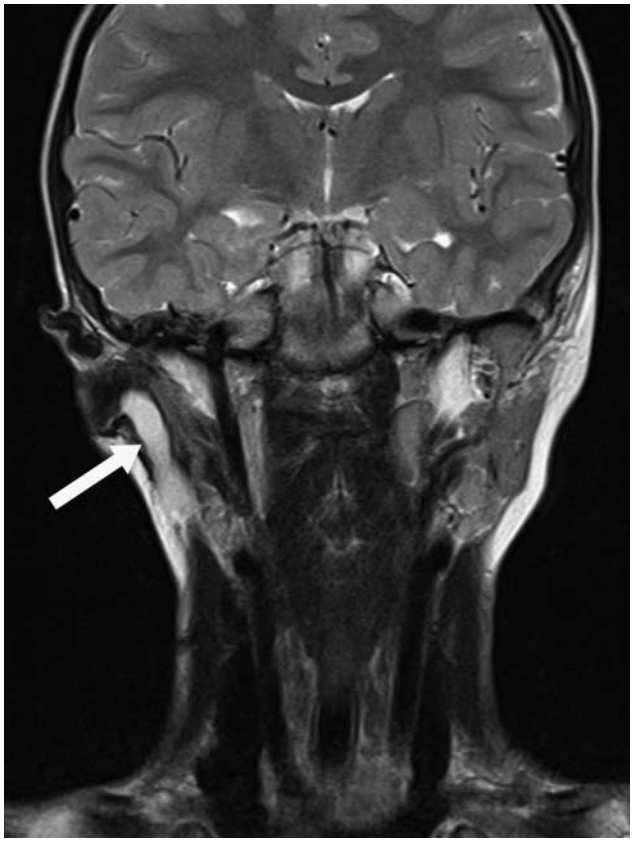
On operation, a duplication of the cartilaginous ear canal was found extending from the junction of the right cartilaginous and bony canals to the superficial sinus. The lesion was directly superficial to the trunk of the facial nerve; therefore, the upper and lower divisions arose deep to the lesion. The zygomatic branch, however, was seen coming off at the stylomastoid foramen and running over

Fig. 3



T1 axial image with a black arrow indicating first branchial cleft anomaly anterior to right external auditory meatus.

Fig. 4



T2 coronal image from case 7 with a white arrow indicating first branchial cleft anomaly.

the surface of the lesion, initially anteroinferiorly before looping posterosuperiorly to cross superficial to the lesion and upper division of the facial nerve.

At 1 week postoperatively, the wound became red, swollen and tender, discharging yellowish fluid. This was managed with a further course of intravenous antibiotics switched to oral after 3 days.

Case 7

A 1-year-old girl presented with a tender swelling inferior to the right mandible. A punctum was noted in the overlying skin, but there was no history of discharge. This was managed with incision and drainage.

This patient presented 5 years later with continuing swelling and occasional discharge from the punctum. MRI showed a cystic lesion extending from the inferior aspect of the right external auditory canal through the parotid gland, with a sinus opening inferior to the right mandible (Fig. 4).

On operation, this lesion was found superficial to the lower division of the facial nerve, yet deeper to the upper division of the facial nerve.

Case 8

A 9-year-old girl presented with an intermittently discharging sinus and associated swelling within the left external auditory canal.

The cyst and sinus were excised, showing an associated tract extending down but not into the substance of the left parotid gland. This was, however, distant from the facial nerve.

A year later she presented with similar symptoms from a pit in the floor of the right external auditory canal. On excision, this was shown to be a less extensive first branchial cleft anomaly, which again was distant from the facial nerve.

Case 9

A 13-year-old girl presented with a swelling anterior to the right ear. On MRI, a lobulated lesion within the right parotid gland was demonstrated (Fig. 5).

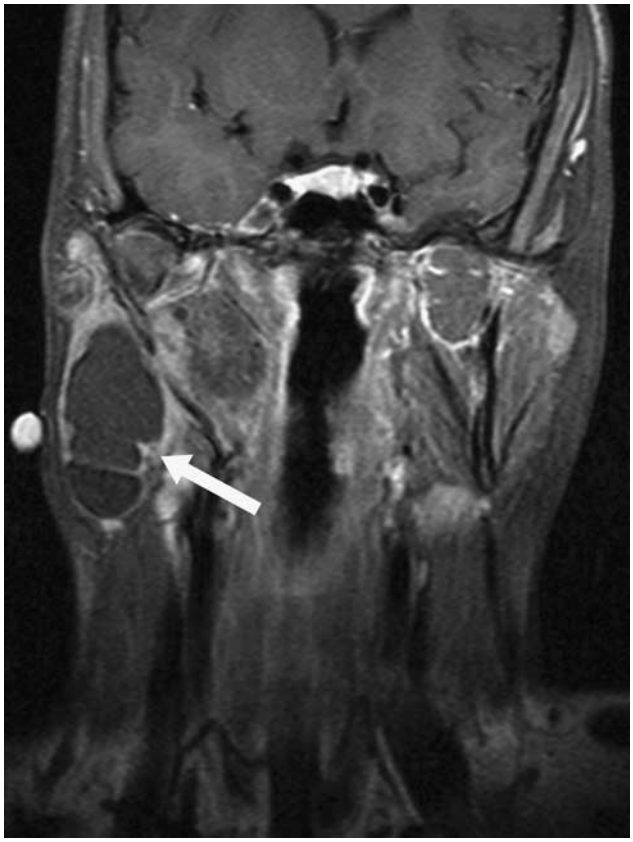
On excision this appeared as a duplication of the right external auditory canal, which was directly superficial to the main trunk of the facial nerve. Retrograde dissection from the facial nerve branches to its main trunk was required to separate the lesion from the facial nerve.

Discussion

The difficulties presented by the current classification systems for branchial cleft anomalies are made apparent in this review of recent cases at our centre.

The lesions described are indeed varied. They range from extensive sinus tracts, which are intimately associated

Fig. 5



T1 fat-saturated postcontrast coronal image from case 9. First branchial cleft anomaly is indicated by a white arrow. The area of clinically apparent swelling has been marked with a cod liver oil capsule, acting as a skin marker.

with the facial nerve, to a sinus directly overlying a small cyst deep to the floor of the external auditory canal. This variation presents as a result of the many disordered processes, which may occur during the complex development of the branchial clefts. Any attempt to separate these lesions into well-defined subtypes will therefore be challenging.

In the first instance, the Arnot classification presents several issues. Classically, an Arnot type I lesion presents during early or middle adult life [8]; however, five of our nine cases in children less than 13 years presented with the same description of a cyst and/or sinus around the parotid gland. Furthermore, the Arnot type I lesions described were distant from the facial nerve in all but one case, which contrasts with the standard presentation of a lesion, which is often closely associated with the lower branches of the facial nerve.

As regards the Work classification, clinical description of these lesions often does not fit the histological results. For example, in cases 1, 2 and 5 the anomalies described fit better with the clinical description of a type I duplication of the external auditory canal. However, cartilage was found on histological investigation of these lesions and therefore they were designated as Work type II. Nonetheless, cartilage found within the specimen is not necessarily part of the anomaly. To excise the lesion completely, cartilage may be removed from the surrounding structures, thereby negating the defining factor in this classification system and rendering it useless clinically. This is in addition to the issue with delay in classification caused by the need for histological examination of the excised specimen.

The Olsen classification is simpler, noting only the morphology of the lesion as a cyst, sinus or fistula. However, this description of the lesion's clinical appearance cannot really be considered as a classification system, despite attempts in the original article to describe separate disordered embryological processes for each subtype. In this case series even the nature of the anomaly is not always immediately apparent without imaging or direct visualization at the time of excision. For example, in case 8, bilateral external auditory canal pits were discovered to represent the end of a sinus tract extending from the parotid region.

Noting these issues, it is important to realize that the ease with which these classification systems are implemented is important only if the information they provide is actually useful. Classification of the anomaly otherwise becomes an academic exercise that provides no benefit to either the patient or the clinician.

An important factor that these classifications could provide an insight into is the lesion's relationship with the facial nerve. This information could provide useful assistance at the time of excision to help prevent the serious complication of facial nerve injury. However, reviewing the relation of each anomaly with the facial nerve in combination with the Arnot, Work and Olsen subtypes showed no consistent correlation. The only exception was the group of Arnot type I lesions, in which four of the five anomalies described were distant to the facial nerve. Conversely, Arnot himself described these lesions as being closely associated with the lower branches of the facial nerve. Therefore, this trend is in direct contrast to the classification's original description. This tendency is also not replicated in other published case series. Del Pero *et al.* [12], in his paper describing the Sheffield experience with branchial cleft anomalies, presented 18 cases in total; of which, 11 were classified as Arnot type I. Of these 11 Arnot type I lesions, only two were distant from the facial nerve, as we have found. Solares *et al.* [11] described 10 cases; of which, seven were of Arnot type 1. In this case series none of these lesions were distant from the facial nerve.

A further possible use of these classification subtypes is to compare outcomes between similar first branchial cleft anomalies. However, it must be noted that these cases represent 10 anomalies excised, with no postoperative facial nerve weakness or recurrence of the lesion despite huge variation in the type of lesion. The standard rate of recurrence overall for first branchial cleft anomalies was 3% after primary excision [6]. However, recurrence rates of up to 22% [6] were reported in those cases complicated by preoperative infection, a situation seen in almost half of the cases we presented. The rate of

facial nerve injury also varies between published reports. Solares *et al*. [11] reported facial nerve injury in 10 patients and Magdy and Ashram [14] reported two episodes of temporary paresis in 18 cases, whereas five cases of temporary paresis and one of permanent facial nerve damage out of 39 patients were reported by Triglia *et al*. [15]. D'Souza *et al*. [1] in his extensive literature review showed a statistically significant ($P = 0.05$) higher rate of facial nerve complications in cases in which the facial nerve had not been identified at the time of excision.

Perhaps, then, a more useful factor to consider when comparing outcomes for excision of first branchial cleft anomalies is the relation of the lesion with the facial nerve, as seen at the time of excision. It makes sense that a lesion that is intimately associated with the facial nerve will have a much higher risk of injury compared with a lesion distant to it. In addition, excision of such a lesion will also pose a higher risk of postoperative recurrence, as the margins taken around the lesion may be limited due to proximity to the nerve. Therefore, as much as considering the position of the facial nerve in relation with the anomaly at the time of excision is important to prevent complications, its position is also important in providing some way to differentiate between these lesions.

Several means of describing the relation of the nerve with the lesion have been used in the literature, although the most common is to describe whether the lesion is superficial or deep to the facial nerve [2]. Our cases also show instances in which the lesion runs between divisions or branches of the nerve and several cases in which the anomaly is distant to the nerve. This information allows a closer comparison of differing types of lesions based on the likelihood of the two most important postoperative complications: facial nerve injury and recurrence.

Conclusion

The cases presented support the premise that the current classification systems used for first branchial cleft anomalies have little clinical relevance. Lesions within the same subtype vary widely, and therefore no useful prognostic information can be inferred from this description. However, the classifications presented do provide extensive descriptions of how these lesions may present, and may be useful in the recognition of these commonly misdiagnosed lesions.

Once the diagnosis of first branchial cleft anomaly has been suggested, the authors recommend that instead of considering these rare lesions in terms of Arnot, Work or Olsen classifications, clinicians should instead focus on collecting as much information as possible before excision. Preoperative imaging, preferably using MRI for its superior soft tissue definition, is recommended. Determining the important relation of the facial nerve will likely still be impossible at this stage, although knowledge of the extent of the lesion can provide useful information. Intraoperatively, a facial nerve monitor should be used to aid recognition and protection of the nerve and its branches. Nonetheless, the surgeon should be prepared to formally dissect out the facial nerve by superficial parotidectomy if required, and should be alert to the possibility of finding the facial nerve in an abnormal position. Finally, we recommend that future reports of these uncommon anomalies provide information on the relative position of the facial nerve, so as to allow comparison between similar lesions in a varied group.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 D'Souza AR, Uppal HS, De R, Zeitoun H. Updating concepts of first branchial cleft defects: a literature review. *Int J Pediatr Otorhinolaryngol* 2002; **62**:103–109.
2 Guo YX, Guo CB. Relation between a first branchial cleft anomaly and the facial nerve. *Br J Oral Maxillofac Surg* 2012; **50**:259–263.
3 Liu Y, Li T, Xue J, Jia J, Xiao S, Zhao E. First branchial cleft fistula presenting with internal opening on the Eustachian tube: illustrated cases and literature review. *Int J Pediatr Otorhinolaryngol* 2012; **76**:642–645.
4 Al Mahdi AH, Al Khurri LE, Atto GZ, Dhaher A. Type II first branchial cleft anomaly. *J Craniofac Surg* 2013; **24**:1832–1835.
5 Ertas B, Gunaydin RO, Unal OF. The relationship between the fistula tract and the facial nerve in type II first branchial cleft anomalies. *Auris Nasus Larynx* 2015; **42**:119–122.
6 Bajaj Y, Ifeacho S, Tweedie D, Jephson CG, Albert DM, Cochrane LA, *et al*. Branchial anomalies in children. *Int J Pediatr Otorhinolaryngol* 2011; **75**:1020–1023.
7 Tham YS, Low WK. First branchial cleft anomalies have relevance in otology and more. *Ann Acad Med Singapore* 2005; **34**:335–338.
8 Arnot RS. Defects of the first branchial cleft. *S Afr J Surg* 1971; **9**: 93–98.
9 Work WP. Newer concepts of first branchial cleft defects. *Laryngoscope* 1972; **82**:1581–1593.
10 Olsen KD, Maragos NE, Weiland LH. First branchial cleft anomalies. *Laryngoscope* 1980; **90**:423–436.
11 Solares CA, Chan J, Koltai PJ. Anatomical variations of the facial nerve in first branchial cleft anomalies. *Arch Otolaryngol Head Neck Surg* 2003; **129**:351–355.
12 Martinez Del Pero M, Majumdar S, Bateman N, Bull PD. Presentation of first branchial cleft anomalies: the Sheffield experience. *J Laryngol Otol* 2007; **121**:455–459.
13 Hickey SA, Scott GA, Traub P. Defects of the first branchial cleft. *J Laryngol Otol* 1994; **108**:240–243.
14 Magdy EA, Ashram YA. First branchial cleft anomalies: presentation, variability and safe surgical management. *Eur Arch Otorhinolaryngol* 2013; **270**:1917–1925.
15 Triglia JM, Nicollas R, Ducroz V, Koltai PJ, Garabedian EN. First branchial cleft anomalies: a study of 39 cases and a review of the literature. *Arch Otolaryngol Head Neck Surg* 1998; **124**:291–295.