First Branchial Cleft Anomalies: Avoiding the Misdiagnosis

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Abstract First branchial cleft anomalies are a very rare entities accounting for less than 1 % of all branchial cleft malformations. They are often misdiagnosed for other cystic lesions occurring in parotid gland and inadequately treated (incision and drainage or incomplete excision) leading to multiple recurrences. We report a series of four patients who were previously operated (incision and drainage) for misdiagnosed first branchial cleft anomalies with subsequent recurrences. All patients underwent superficial parotidectomy with complete tract excision using facial nerve monitoring to prevent iatrogenic injury because of extensive fibrosis. We discuss the literature pertaining to first branchial cleft anomalies, their varied presentations and their relationship to facial nerve in parotid gland and importance of facial nerve monitoring in revision surgery.

Keywords First branchial cleft anomalies · Facial nerve · Superficial parotidectomy · Facial nerve monitoring

Introduction

First branchial cleft anomalies are rare and account for 1 % of all branchial cleft malformations [1, 2]. Presentation can be either in the form of cyst, sinus or fistula in close relation to external auditory canal (EAC), parotid and

anterior neck. Acierno has further classified first cleft anomalies as type I and type II [1]. Type I anomalies run lateral to facial nerve, composed of ectoderm histologically and present as swellings near the ear. Type II anomalies, on the other, run medial to facial nerve, composed of both ectoderm and mesoderm and present between EAC and hyoid bone [1]. Correct diagnosis is often delayed because of their rarity resulting in inadequate treatment and subsequent recurrences. We report a series of four such patients of first branchial cleft anomalies presenting as recurrent parotid swelling along with other clinical presentations of first branchial cleft anomalies, their relationship to facial nerve and management issues.

Materials and Methods

A retrospective chart analysis was performed for first branchial cleft anomalies. Four cases were retrieved. Demographic data in terms of age, sex, duration of symptoms, mode of presentation, pre-operative facial nerve status, number of previous surgeries, type of surgical procedures, intraoperative findings including relationship with facial nerve are listed in Table 1. Male to female ratio was 1:1 with almost all cases presenting in first and second decade of life. Parotid swelling was present in all cases; in addition two cases also had auricular signs and one case had auricular, parotid and cervical signs (Fig. 1a, b). Radiology in the form of computed tomography was done in all cases (Fig. 2). All patients had previously undergone surgery in the form of incision and drainage. Facial nerve was normal in all cases pre-operatively. All patients underwent superficial parotidectomy along with complete excision of tract with facial nerve monitoring. In two cases with auricular signs, cuff of cartilaginous EAC was

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Table 1 Demographic data of cases

S.no	Age (y)	Sex	Duration of symptoms (y)	Presenting symptoms and signs	No. of previous surgeries	Relation to facial nerve
1	21	F	15	Parotid	2	Superficial
2	35	F	Since childhood	Parotid, auricular, cervical	3	Superficial
3	39	M	7	Parotid	1	Superficial
4	23	M	3	Parotid auricular	1	Superficial

Fig. 1 a, b Clinical photographs of the patients 1 and 2 showing parotid lesion with scar marks of previous surgeries

also excised. Post operatively facial nerve was normal in all cases. Two cases had marginal mandibular paresis which recovered completely in the follow-up period. Till date all patients are recurrence free.

Histology

Histopathological examination showed similar features in all cases. A cyst filled with keratinous material was identified adjacent to the salivary gland parenchyma. The fibrocollagenous cyst wall was lined by stratified squamous epithelium, and showed the presence of lymphoid aggregates with germinal centers below the lining epithelium (Fig. 3).

Discussion

Branchial cleft anomalies constitute approximately 30 % of all congenital neck masses. The presentation can be either a sinus, cyst or fistula depending on the incomplete embryological development [2, 3]. However, first branchial cleft anomalies are extremely rare. The first cleft and pouch form the external auditory canal (EAC), eustachian tube, middle ear cavity, and mastoid air cells. First branchial cleft anomalies may commonly involve EAC or rarely middle ear and have a close relation to parotid gland and facial nerve.

Different classification systems have been proposed for first branchial cleft anomalies. Arnot proposed two types: Type I anomalies lie within the parotid gland and lined by squamous epithelium. These type of anomalies present during early or adult life. The theory of origin was postulated to be buried cell rests during obliteration of branchial groove. Type II anomalies are because of incomplete closure of the cleft and found in the anterior triangle of the neck, with a communicating tract to the EAC [4]. Work proposed another classification describing type I anomaly as duplication of membranous EAC and type II anomaly as duplication of both membranous EAC and pinna [5]. Oslen has classified first branchial cleft anomalies into cysts, sinus and fistulas [6].

First branchial cleft anomalies are more common in females and more frequent on left side [7]. They can either have cervical or parotid or auricular signs or combination of them. Cervical signs include pit-like depression near angle of mandible with recurrent watery discharge which can be purulent if infected. Parotid signs included cystic parotid mass and auricular signs include ear discharge with normal tympanic membrane or swelling or fistulous opening in floor of EAC [1]. The sinus or fistulous cutaneous opening is usually found in a triangle bounded superiorly by external auditory canal, inferiorly by hyoid bone and anteriorly by mental region (Poncet's triangle) [8]. Two of our patients presented with parotid signs only as recurrent parotid region swelling and other two presented with auricular signs combined with parotid as well as cervical signs. First branchial cleft anomalies are often misdiagnosed resulting in inadequate treatment like incision and drainage or incomplete excision with subsequent recurrences. Oslen had mentioned that cystic lesions of first branchial cleft anomalies were often misdiagnosed as parotid tumors [6]. The average number of surgeries required for complete excision being 2.4 per patient [8]. In our series the average number of surgeries required for complete excision were 2.2 per patient. Multiple previous surgeries place the facial nerve at higher risk during subsequent surgeries. Three types of relation have been described between first branchial cleft anomaly and facial nerve. It can be either superficial or deep to the nerve or it can run between the branches of facial nerve. Guo et al. [9] in their analysis of 41 patients with first branchial cleft

Fig. 2 a, b Computed tomography (axial and coronal cut) showing well defined cystic lesion in left parotid gland

Fig. 3 Photomicrograph showing a keratin filled cyst lined by stratified squamous epithelium (a, H & E, $\times 100$). Higher magnification view showing focal lymphoid aggregates below the lining epithelium (b, H & E, $\times 200$)

anomalies found that younger patients, lower lesions, lesions on right side and Work type II lesions were associated with anomalies deep to facial nerve. They also found that cysts were superficial to facial nerve in most of the cases, as in two of our cases. Misdiagnosis is common for first branchial cleft anomalies [10]. The differential diagnosis for a cystic lesion in parotid gland includes Warthin's tumor, sialocele, infected lymph nodes, mucoepidermoid carcinoma, benign lymphoepithelial cyst of human immunodeficiency virus (HIV) and first branchial cleft cyst. The correct diagnosis can be achieved by suspicion of first branchial cleft anomaly and meticulous examination looking for external auditory canal sinus or fistula, or cutaneous opening in Poncet's triangle. Imaging in the form of CECT or fistulogram is helpful in diagnosis. The aim of the surgery is to remove the cyst or sinus or fistulous tract completely and to prevent injury to facial nerve. This

requires superficial parotidectomy and excision of the cuff of skin and cartilage of EAC if tract is extending till EAC. We recommend use of facial nerve monitor in recurrent cases as the facial nerve may be difficult to identify in fibrotic tissues.

Conclusion

First branchial cleft anomalies are rare and often misdiagnosed leading to recurrences and high risk of iatrogenic facial nerve injury during subsequent surgery. High index of suspicion is required to diagnose them during first presentation. Complete excision using facial nerve monitoring should be performed in a case with multiple recurrences anticipating extensive fibrosis and distortion of normal tissue planes because of previous surgeries.

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