Case Report
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Facial Rehabilitation after Surgical Excision of Intra-Parotid Facial Nerve Schwannoma

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Abstract

Background: Plexiform schwannomas of the intra-parotid facial nerve are rare slow growing benign tumours, usually painless and sometime present with facial disability. The aim of this case discussion is to raise awareness of this rare clinical entity, highlight the difficulty to diagnose this condition pre-operatively, discuss the management options and the role of speech and language therapist in facial rehabilitation post-operatively.

Method: Case report and literature review.

Case Report: We present an unusual case of a 21-year-old Caucasian male who presented with a painless lump in his right parotid gland which clinically appeared to be a benign salivary gland tumour. MRI scan revealed an intra-parotid mass but the Fine Needle Aspiration Cytology (FNAC) was inconclusive. The patient underwent right extra capsular parotidectomy *via* face lift approach. Intra-operatively part of the facial nerve was identified and multiple nodules were excised. Unfortunately, the patient woke up with a complete paralysis of his right face. In the immediate post-operative period, the facial paralysis was managed conservatively. Subsequently, the patient underwent intense therapy by our speech and language therapist. One year later, the facial palsy improved significantly to HB grade III. The formal pathology revealed plexiform schwannoma.

Conclusions: Plexiform schwannoma of the facial nerve is an extremely rare benign tumour. Every effort should be made to diagnose this condition pre-operatively if serious outcome is to be avoided.

Keywords: Plexiform schwannoma; Peripheral nerve sheath tumour; Neurofibroma; Salivary gland; Magnetic resonance imaging; Histopathology; Facial palsy; Rehabilitation

Introduction

Plexiform schwannoma is a rare distinct entity of schwannoma, accounting for 2-5% of all schwannomas [1-4]. In the head and neck region, the parotid gland is the preponderant site for mesenchymal neoplasms and schwannoma [4]. Apart from the parotid gland, rare cases have been described in the submandibular gland [5] and in the sublingual gland [6].

Clinically, patients usually present with a painless lump but can also present with symptoms related to mass effect caused by the tumour including lower motor neuron facial nerve palsy and dysphagia [4,7]. Gross et al., found that 87.1% (n=108) of intra-parotid schwannomas within the literature presented as painless masses, while 18.5% (n=23) caused some form of facial nerve dysfunction [8].

In this article, we present an unusual case of an intra-parotid plexiform schwannoma. The aim of this case discussion is to raise awareness of this rare clinical entity, highlight the difficulty to diagnose this condition pre-operatively, discuss the management options and the role of speech and language therapist in facial rehabilitation post-operatively.

Case Report

A 21-year-old Caucasian male presented with a painless lump in his right infra-auricular region which had been present for several years. Apart from history of smoking, he was fit and well. Physical examination revealed a 2x2 cm lump in his right parotid gland with normal overlying skin. The rest of the ENT examination was normal.

Magnetic Resonance Imaging (MRI) scan showed a well-defined intra-parotid lesion measuring 34x20x30 mm. The lump was located in the superficial lobe extending into the deep lobe of the

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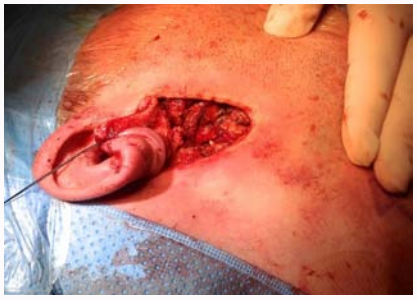

Figure 1: An example of face lift incision for right partial partotidectomy.

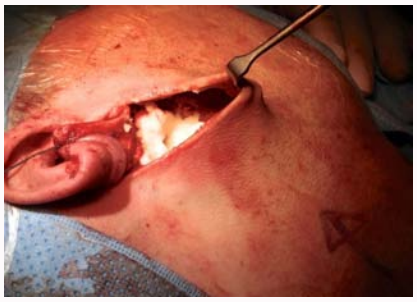

Figure 2: The haemostasis is achieved using the bipolar diathermy and absorbable haemostatic agent. The patient was provided with a head bandage and no drain was inserted after the surgery.

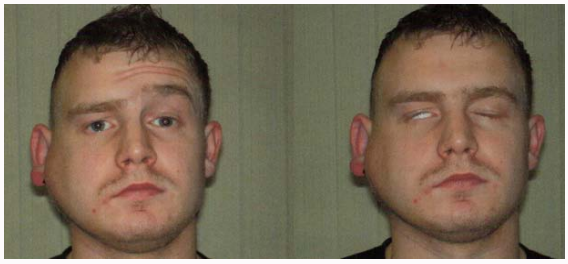

Figure 3: Right sided facial weakness 2 weeks after the operation.

right parotid gland. The lump had a heterogeneous intermediate appearance on T2 and STIR and there was an isointense signal on T1. The lump demonstrated mass effect with medial displacement of the retromandibular vein. The scan also showed bilateral level II lymph nodes which were not considered to be significant. Clinically, the lesion appeared to be a pleomorphic adenoma but the Fine Needle Aspiration Cytology (FNAC) was inconclusive.

After careful counselling, decision was taken to intervene surgically. The patient underwent extracapsular right partial parotidectomy through a small face lift incision (Figure 1). Facial nerve monitor was not used but the surgery was assisted with surgical loupe magnification. Intra-operatively, multiple nodules were found scattered within the parotid gland. Part of the facial nerve was identified and preserved. Three discrete lumps were excised which were mostly in the superficial lobe but were also extending into the deep lobe. Another lump was palpable in the anterior part of the parotid gland but was not removed in order to avoid damage to the terminal branches of the facial nerve. Haemostasis was achieved and no drain was inserted (Figure 2).

In the recovery room, the patient was found to have House-Brackmann (HB) grade VI facial nerve palsy affecting his right face. In the immediate post-operative period, the facial paralysis was managed conservatively. The patient was discharged home but was readmitted 2 days later with post-operative wound infection requiring intravenous antibiotics to which he responded well. The surgical exploration with a view to reconstruct the facial nerve was offered but declined by the patient.

The histopathological examination of the parotid lumps confirmed a diagnosis of plexiform schwannoma with classic Antoni A component and Verocay bodies were seen; there was absence of Antoni B component. Immunohistochemistry for S-100 revealed strong and diffuse positivity in the spindle cells. Neurofilament Protein (NFP) immunohistochemistry was negative and Epithelial Membrane Antigen (EMA) immunostaining showed positivity in the perineural cells in the capsule of the nodules. There was no evidence of dysplasia or malignancy.

The patient was managed in a multidisciplinary skull base clinic. He underwent intense facial rehabilitation therapy with our speech and language therapist. On his follow-up visits, the patient continued to make good clinical progress. Two-week post-operatively, he had acquired reasonably good facial motor function related to the cervical and marginal mandibular branches. However, there was absence of movement in the buccal and upper divisions of the facial nerve (Figure 3). He continued with the facial nerve trophic stimulation and biofeedback. At 2-month review, the ophthalmic branch appeared to show some recovery and he was able to close his right eye completely (Figure 4). A year post-operatively, his facial palsy improved to HB grade III (Figure 5). The patient has continued to attend our department for clinical and radiological surveillance of the residual lumps in his right parotid gland for the last 9 years.

Discussion

Plexiform Schwannoma (PS) is a rare, benign, nerve sheath tumour; it is typically very slow growing and has no malignant potential. It represents approximately 4.3% of all schwannomas. They occur in two types, either conventional or cellular type. It is characterised by an intraneural, plexiform and often multinodular pattern of growth. There is no association between plexiform

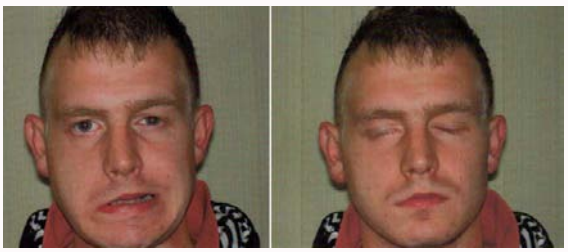

Figure 4: Right sided facial weakness 2 months after the operation.

Figure 5: Right sided facial weakness 1 year after the operation.

schwannomas and neurofibromatosis type 1, it can however occur in the setting of neurofibromatosis type 2 [3,9]. These tumours tend to arise in superficial soft tissue and show a predilection for the head and neck region, this is thought to be due to the abundance of superficial peripheral nerves in this area [10].

Plexiform Schwannoma (PS) may occur in association with neurofibromatosis such as NF2 but not with NF1 [3,9]. One study reported a rate of 11% of PS being associated with NF2 [2]. Tumours associated with a syndrome will generally occur at a younger age group. For patients within this age group who present with PS, consideration must be given to whether there is an underlying syndrome present. If so then it is likely that multiple lesions will be present. The histopathological features of syndrome associated lesions do not vary from other lesions [2]. Our patient did not display any features suggestive of this.

Plexiform Schwannoma (PS) can be very large and they can show a macroscopic or microscopic plexiform pattern often with multi-nodularity. Lesions are usually encapsulated by residual perineurium. PS usually occurs in the conventional subtype although less frequently it can be of the cellular type. Most conventional type lesions will show Antoni A tissue, Verocay bodies are often identified, and Antoni B tissue may also be present. These lesions will generally stain diffusely positive for S-100 protein [3,7,9,11]. Fine Needle Aspiration Cytology (FNAC) is helpful if the Verocay body is observed, but in most cases similar to our case, the diagnosis is inconclusive or mistaken as pleomorphic adenoma [12-14].

Pre-operative diagnosis of intraparotid schwannomas is often challenging radiologically and pathologically. Ultrasound and Computed Tomogram (CT) can assist in the diagnosis. However, magnetic resonance imaging with gadolinium can provide more specific characteristics of the schwannoma especially when used in conjunction with cytology analysis. This is more helpful in diagnosing intra-parotid schwannomas. Shimizu et al., suggested that the presence of a target sign (increased peripheral signal intensity and decreased central signal intensity on T2 weighted images) and a growth toward the facial canal are distinguishing features of this type of tumour [15]. Nevertheless, radiological findings can be non-specific, and the presence of bland spindle cells in biopsies does not exclude pleomorphic adenomas [7,14]. Therefore, greater awareness of intra-parotid schwannomas may help to increase the diagnostic accuracy, thus avoiding unnecessary facial nerve morbidity.

The most common differential diagnosis for a parotid lump is a pleomorphic adenoma, which is a benign tumour that can occasionally undergo malignant transformation into carcinoma ex-pleomorphic adenoma with distant metastasis. Other common parotid lumps include haemangioma, lipomas and Warthin's tumour also known as lymphamatous papillary cystadenomas. The latter is an entirely benign tumour usually in the tail of the parotid gland. Malignant parotid tumours are rare and among others include mucoepidermoid and adenoid cystic carcinomas. Neurogenic tumours such as neurofibromas and malignant peripheral nerve sheath tumours should also be considered. Neurofibromas typically occur with NF 1, it is important that these are identified as they have the potential to become malignant. A traumatic neuroma may also present in a similar manner.

Plexiform Schwannomas (PS) are generally managed with surgery alone [12,15-18]. Given their slow rate of growth PS do not tend to respond to chemotherapy or radiotherapy. Surgical excision should be aimed at complete excision of the tumour with preservation of the facial nerve but this can be very challenging when these tumours can be multi-focal as seen in our case. Complete excision of the tumour is usually carried out either with a partial or total parotidectomy [4]. The latter often carries a significant risk of permanent facial nerve palsy. Alicandri-Ciufelli et al., had proposed a decision-making algorithm in managing intra-parotid facial nerve schwannoma [16]. In the case of type A or B neoplasms, or if the patient had a pre-operative facial nerve HB IV or worse, the authors would favour a resection of the intra-parotid facial nerve schwannoma and to do a reconstruction of the nerve where necessary. In the case of pre-operative HB grade III or better and type C or D neoplasms, patients would undergo an intra-operative biopsy to rule out malignancy, and a possible conservative management could be adopted [16].

Literature search showed a variety of techniques of PS excision and relevant outcomes. Lee et al., report that it is possible to remove intra-temporal Facial Nerve Schwannoma (FNS) and preserving a reasonable facial nerve function, by performing stripping surgery [17]. Ma et al., had also reported that in most intra-parotid FNS, it is possible to dissect the schwannoma off the nerve trunk and preserve the facial nerve function [18]. Lee et al., had found similar findings with their cohort of 14 intra-parotid FNS achieving normal facial nerve function in all their patients post-operatively [19]. Contrary to this, Li et al., proposed only to proceed with tumor removal if it can be resected easily off the nerve. They did not recommend dissecting the tumor from the facial nerve in cases where the tumor had intra-temporal lesions [20]. Cho et al., found in their cohort of 18 patients with mesenchymal tumours of the major salivary glands, neurogenic tumors were the most common benign neoplasms. Five of six schwannomas occurred in the parotid gland. At the time of surgery, these schwannomas were found arising from temporal and zygomatic branches of the facial nerve [4]. In our patient, it is likely his schwannoma probably arose from the buccal branch and the upper division of the facial nerve. Consequently, the most severe loss of function was observed in those areas. Facial nerve repairs using sural nerve grafts and hypoglossal-facial nerve cross-over can be considered to restore facial nerve function and assist in facial rehabilitation [13]. Our patient had declined re-exploration and facial nerve repair.

Management of facial palsy may include medical therapy, surgical decompression, physical therapy, injectable fillers and surgical reanimation procedures. When the integrity of the facial nerve is preserved in its entirety or in parts whereby some terminal branches were sacrificed, facial rehabilitation can assist in regaining a reasonable degree of facial function. The HB grading system [21] has been criticized for its lack of zonal facial paralysis; hence, eFACE [22] has been popularized as the better and more accurate scale for facial paralysis assessment to assist rehabilitation. Facial paralysis is not only debilitating physically to patients but its effect on speech, communication, oral functions, emotional expression and psychological upset are well recognized [23-25]. In 2011, the Cochrane library published an updated systematic review on physical therapy which includes exercise, biofeedback, laser treatment, electrotherapy, massage and thermotherapy, used to accelerate recovery, improve facial function and minimize sequelae for Bell's palsy [26]. No statistically significant inter-group differences were found. The authors concluded that there was low quality evidence that facial exercises reduce long term sequelae in acute cases. The effects of tailored facial exercises need to be further confirmed with

good quality randomised controlled trials [26]. In the same year, Pereira et al., only found one study presenting sufficient data to perform a meta-analysis [27]. Randomised controlled trial on mime therapy had shown significant improvements in functionality in facial paresis [28]. Our patient had beneficial effect with facial nerve trophic electromyographic (EMG) stimulation and mirror feedback training.

Finally, PS do not metastasize but if excision is incomplete then there is a risk of local recurrence. Our patient had 4 known PS in the superficial and deep lobes of his parotid gland. One of the 4 PS was not fully excised to avoid damaging one of the terminal branches of the facial nerve. In view of this, he is on long-term clinical and radiological surveillance.

Conclusions

This case demonstrates the difficulty of pre-operative diagnosis of this tumour and indeed of all intra parotid neurogenic tumours. The diagnosis of a neurogenic tumour should be considered for all head and neck masses. The mainstay of treatment is surgical excision as these tumours do not respond to radiotherapy but this does carry a significant morbidity to the patient and risk of permanent facial paralysis. Although the use of facial nerve monitoring is a standard practice in parotid surgery these days, excision of multi-focal PS still carries a high risk of facial nerve palsy. It is therefore vital to include this in the informed consent. Surgery has to be tailored to each individual case and risk assessment of each case has to be made. Long-term surveillance with parotid MRI scans is necessary particularly in patients with incomplete tumour excision or when the complete excision of multi-focal PS is doubtful. Timing and selection of therapeutic intervention in facial palsy is critical. Facial rehabilitation can provide satisfactory patient reported outcome. We recommend managing these patients in a multidisciplinary team setting involving a speech and language therapist with a special interest in facial palsy rehabilitation. This can lead to favourable outcome as is evident in our patient.

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