An Intraparotid Schwannoma Mimicking A Warthin’s Tumour

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Received date: December 24, 2019; Accepted date: January 09, 2020; published date: January 14, 2020

Citation: Awan Z, Azam A (2020) An Intraparotid Schwannoma Mimicking A Warthin’s Tumour. J Clinical Research and Reports, 2(2); DOI:10.31579/jcrr.2020/013

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Abstract

In this report, we describe the rare presentation of a cystic intraparotid facial nerve schwannoma (IPFNS) in a young female who initially presented with a painful unifocal parotid mass gradually enlarging over 5 years. Preliminary imaging and cytology offered this to be a Warthin’s tumour. Here we describe the limitations of current diagnostic tools in the workup of intraparotid cystic lesions and highlight the importance of keeping rare tumours in mind to optimize pre-surgical management.

Introduction

Schwannomas are very rare, benign, slow-growing, encapsulated tumours of neuro-ectodermal origin from neural sheath Schwann cells [1]. Approximately 30% of schwannomas occur in the head and neck with the majority being in the eighth nerve [2]. There is no clear sex predilection and peak incidence is between the third and sixth decade [3]. The aetiology is still not clear and no identifiable risk factors have been demonstrated thus far for solitary schwannomas however schwannomas may be seen in the autosomal dominant neurofibromatosis type 2 alongside at least one other nervous system tumour such as an acoustic neuroma, meningioma and glioma [4]. A facial nerve schwannoma (FNS) can occur at any point along its course from the cerebello-pontine angle to its peripheral branches [5]. In a review of 700 parotidectomies, Nussbaum observed only 1 case of FNS [6]. The IPFNS is an even rarer entity accounting for only 9% of schwannomas arising from the facial nerve itself [7]. The incidence of IPFNS is about 0.2 to 1.5% [8]. One retrospective study found that only 20 schwannomas out of 3722 involved the facial nerve of which 8 involved the intraparotid segment of the facial nerve [9]. Less than 100 cases of IPFNS have been reported in the literature.

As they are not a prioritized pre-operative diagnosis, they can lead to unintentional treatment errors and secondary intraoperative complications such as facial nerve palsy. Pre-operative diagnostic modalities such as an ultrasound scan (USS), magnetic resonance imaging (MRI) and fine needle aspiration cytology (FNAC) often narrow down the differentials when approaching parotid masses, aiding surgical excision. Most parotid tumours have characteristic cytomorphological features, which can be identified via the aforementioned modalities. Nonetheless because of the very few typical signs of IPFNS, interpretation can be difficult [10].

Case:

A 39 year old pregnant Pakistani female presented with a 5 year history of a painful enlarging right parotid mass. The patient denied any history of facial weakness or constitutional symptoms. She had an unremarkable past medical and family history and was not on any medications. On clinical examination, there was a tender, soft, mobile lump measuring approximately 20mm x 20mm. There were no signs of infection or associated lymphadenopathy and no evidence of facial nerve involvement. Laboratory tests including white cell count, C-reactive protein, viral serology and erythrocyte sedimentation rate were all within normal limits.

An initial USS of the salivary glands demonstrated a hypoechoic, 25mm, lobulated, thick-walled cyst, deeply embedded in the posterior part of the right parotid gland, lying posterior to the mandible (Figure 1).
Figure 1: A markedly hypoechoic, well-circumscribed mass in the right parotid gland with no demonstrable internal vascularity representing a cyst with inspissated contents or a mixed solid and cystic lesion.

For further characterisation, MRI was performed during which period the pain had significantly worsened. This showed a well-defined predominantly cystic lesion measuring 22mm x 22mm x 17mm with low intensity T1 and high intensity T2 signals along the superficial and deep lobes of the right parotid, centering the retromandibular region. The solid components demonstrated contrast enhancement (Figure 2).

Figure 2: A well-defined predominantly cystic lesion. The solid components demonstrate contrast enhancement.
A few normal–appearing intra–parotid lymph nodes were noted with no enlarged or suspicious neck nodes. The conclusion was made of a benign or low–grade predominantly cystic tumour extending into the deep lobe, most likely representing a Warthin’s tumour. The patient subsequently underwent FNAC as she was pregnant and not willing to undergo surgery. Moreover, given the possibility of a low–grade possible malignancy which would have changed the management approach, invasive testing was required. The FNAC showed a benign lesion with cohesive sheets of oncocytic type cells and scattered lymphocytes in keeping with a Warthin’s tumour. As the patient was pregnant, further intervention was postponed. Repeat MRI findings 16 months later were consistent with the previous scan however demonstrated an increase in size to 30 x 22 x 21 mm. The patient subsequently underwent a surgical resection of the mass on the preoperative MRI and FNAC conclusion of a symptomatic Warthin’s tumour. The operation was difficult due to the fact that the cystic tumour appeared to be stuck to the facial nerve, possibly branching into the cyst wall. The cystic lesion burst with spillage into the wound. The surgery was completed with retrograde tracing of the branches of the facial nerve. Postoperatively, the patient sustained a right facial nerve dysfunction. Histology later confirmed an intraparotid gland schwannoma of the facial nerve with cystic areas. On macroscopy, the cut surface showed a well–demarcated, white, firm nodule. Focal cystic changes with haemorrhage were noted with a surrounding rim of adipose tissue. On microscopy, there was an encapsulated lesion with a predominance of Antoni A areas (Figure 3).

**Figure 3:** Hematoxylin & Eosin Stain (x1 magnification). Encapsulated solid tumour composed of sheets of spindle cells forming Antoni A areas.

The solid areas comprised sheets of spindle cells with tapered bland nuclei. Characteristic nuclear palisading forming Verocay bodies and hyalinised blood vessels were also present (Figure 4).
The cystic area showed a rim of macrophages with few lymphocytes, haemorrhage and cellular debris. There was no evidence of malignant changes ruling out the even rarer possibility of a malignant peripheral nerve sheath tumour (MPNST). A few small lymph nodes were also present which showed reactive changes. Completeness of excision was likely.

After discussion at the head and neck MDT, adjuvant treatment was not recommended. The patient was discharged with oral prednisolone, a 2-week outpatient review, a 12-month follow-up MRI, facial nerve rehabilitation and eye care. At the 12 month follow up appointment the patient’s facial nerve function was gradually improving. A plan was made to discuss the option of nerve reanimation with the plastic surgeons.

**Discussion:**

IPFNS was first reported by Ibarz in 1927. It typically presents as a slow-growing, painless mass. Unusually, our patient presented with a painful mass. One literature review shows that more than 75% of IPFNS remain unknown until after surgery as in our case [11]. Nonetheless, the potential for a preoperative diagnosis of FNS is essential for treatment planning [12]. The most common differential diagnoses of cystic parotid masses include retention cysts, post-traumatic sialoceles, Warthin’s tumour, mucoepidermoid carcinoma and necrotic metastases [13].

From an imaging point of view, there is no concrete evidence that any radiological method is superior [10]. In essence, there are no definitive radiological findings for IPFNS (14). In most reports, results are inconclusive or suggestive of a different pathology, as in our case. There have been several reports in which pre–histological diagnostic investigations have mistaken FNS for pleomorphic adenomas [10, 15]. MRI findings suggestive of FNS such as the ‘string sign’ and ‘target sign’ [16,17] were not demonstrated in our case. Nonetheless, these findings are not specific and may be found in other neurogenic tumours such as neurofibroma and MPNST [10]. Moreover, a CT scan was not employed as previous authors [18] have emphasized that CT is not the ideal imaging modality for evaluation of parotid masses, primarily because soft tissue structures and the facial nerve are not well visualized. On the other hand, some authors have reported that a schwannoma that has undergone cystic degeneration will appear as a multilocular cystic structure with multiple, fine septations and solid areas, along with calcifications within the cyst walls on CT [19]. Given our histological findings, this may have been useful to compare. The general conclusion has been that a multitude of parotid tumours can display identical features on imaging thus, the heterogeneity of imaging features and the resulting broad differential diagnosis leads to very low rates of preoperative diagnosis [10].

The literature on the use of FNAC is inconclusive. Some report sensitivity and specificity as 60–100% and 90–100% respectively [20,21] whereas others have found that FNAC is very often not diagnostic, and as in our case, can lend weight to an erroneous diagnosis due to lack of characteristic cytological findings and exact cell type [7,15, 22– 25]. Cho et al overcame this difficulty by employing intra–operative frozen section studies to reach the correct diagnosis [26]. On the one hand, frozen section examination that has been performed intra–operatively is recommended for both diagnosis and exclusion of malignancy [5,27]. On the other hand, occasionally the schwannoma is mistaken for a fibrosarcoma leading to unwarranted radical surgery [28]. Yologlu et al have suggested the option of diffusion weighted images and apparent diffusion coefficients as having a potential role in delineating different parotid masses [29].

Mehta and Deschler agreed with the option of intraoperative conservative biopsy in a nonstimulating portion of the lesion to make a diagnosis [30]. Classification systems have been proposed based on variables such as House– Brackmann grades, size, position and adherence of the mass with

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**Figure 4:** Hematoxylin & Eosin Stain (x4 magnification). Evidence of characteristic Verocay bodies and hyalinised blood vessels. Cells are narrow, elongated and wavy with tapered ends interspersed with collagen fibers.
In summary, cystic intraparotid schwannomas are extremely rare and generally unsuspected. Due to their nonspecific features on imaging and FNAC, they have a tendency to be misdiagnosed preoperatively, highlighting the limitations and pitfalls of the current diagnostic pathway prior to surgery.

Learning Points:

1. Intraparotid facial nerve schwannomas may mimic common neoplastic salivary gland conditions on FNAC and imaging.
2. When approaching a unifocal, unilateral cystic parotid mass, in light of the pre–surgical diagnostic limitations, always keep in mind rare neuroectodermal tumours such as schwannomas due to the subsequent clinical consequences this may have on the patient’s nerve function and recovery.
3. A painful unilateral parotid mass with or without facial nerve involvement may represent a schwannaoma.
4. Further research is required in to new diagnostic tools and surgical options if surgeons encounter IPFNS.

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