Case Report

DOI: 10.7860/IJARS/2022/52705.2781

Ear, Nose and Throat Section

Parapharyngeal Schwannoma Mimicking Peritonsillar Abscess in a Young Female: A Rare Case Report

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ABSTRACT

Parapharyngeal Space (PPS) tumours are responsible for 0.5% of head and neck cancers. Presenting features can mimic Peritonsillar Abscess (PTA). A differential diagnosis of PPS tumour should therefore be considered in young, systemically well patients with peritonsillar masses that are refractory to antibiotic therapy. This report focuses on a rare case of parapharyngeal schwannoma in an otherwise fit 29-year-old female, who presented with a three month history of a right sided oropharyngeal swelling and a sensation of 'something' in her throat. Blood tests, specifically full blood count and C-Reactive Protein (CRP) were normal and attempts at aspiration under local anaesthetic yielded no pus. As the patient showed no signs of improvement despite Intravenous (i.v.) antibiotics and steroid therapy; further investigations were initiated including Flexible Nasoendoscopy (FNE) and contrast enhanced Magnetic Resonance Imaging (MRI) of the neck. The FNE revealed a right sided parapharyngeal mass which was later confirmed on MRI scanning, measuring 7 cm in craniocaudal dimension arising from the deep lobe of the parotid gland. After a fine needle aspiration yielded inconclusive histological evidence, Multidisciplinary Team (MDT) discussion advised surgical excision. The patient underwent excision of the right parapharyngeal tumour and right partial parotidectomy via a transparotid and transcervical approach utilising a modified blair incision and a facial nerve monitor. Histology confirmed a parapharyngeal schwannoma. Postoperative outcomes have been reassuring as the patient has been disease-free four years postexcision.

Keywords: Flexible nasoendoscopy, Head and neck tumours, Oropharyngeal swelling

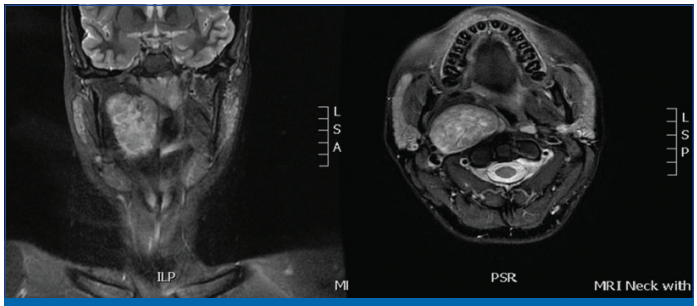
CASE REPORT

A 29-year-old otherwise fit and well caucasian female presented with a three month history of right sided oropharyngeal swelling, referred otalgia and hoarse voice to the Otolaryngology Department. She was an ex-smoker with no relevant medical history and was not on any regular medication.

Oropharyngeal examination highlighted an erythematous, non exudative right sided peritonsillar mass. No clinically palpable cervical lymphadenopathy was noted. A working diagnosis of right PTA was made. Blood tests, specifically full blood count and CRP was unremarkable. Multiple attempts at needle aspiration, incision and drainage under topical anaesthesia were made but no pus was yielded. Intravenous (i.v.) benzylpenicillin 1.2 grams four times daily and i.v. metronidazole 500 milligrams three times daily was administered along with i.v. dexamethasone (6.6 milligrams once daily) and analgesic therapy (paracetamol 1 gram four times daily, ibuprofen 400 milligrams three times daily, codeine 60 milligrams as required upto a maximum of four times daily and benzydamine oral rinse 15 millilitres four times daily).

Flexible nasopharyngolaryngoscopy revealed a large right sided parapharyngeal mass with associated swelling in the right level 2 region, as well as mobile vocal cords and normal appearances of the glottis and hypopharynx. Contrast enhanced MRI of the neck confirmed a right parapharyngeal mass measuring 7 cm in cranio-caudal dimension arising from the deep lobe of the parotid gland. The mass displayed high T2 and low T1 signals with enhancement of the solid component [Table/Fig-1].

Radiological findings pointed that the mass arising from the deep lobe of the parotid gland was most likely to reflect a pleomorphic adenoma of the parotid gland. However, fine needle aspiration yielded inconclusive histological evidence therefore the patient was discussed in a Multidisciplinary Team (MDT) meeting.

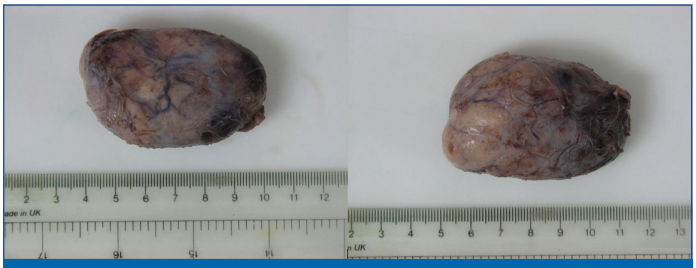


[Table/Fig-1]: Contrast MRI scan of the neck demonstrating an enhancing right parapharyngeal mass (coronal view on the left and axial view at the level of C2 vertebral body on the right).

Following MDT discussion, the definitive plan was for surgical excision. The patient was not initiated on any medication prior to surgery. The patient underwent excision of the right parapharyngeal tumour and right partial parotidectomy via a transparotid and transcervical approach utilising a modified Blair incision and a facial nerve monitor. The main facial nerve trunk and branches were identified and preserved (confirmed via stimulation at the end of the procedure). To facilitate access to the tumour, a facial flap was raised anteriorly and the parotid gland was separated from the sternomastoid and the posterior belly of digastric. The greater auricular and facial nerves were identified and preserved.

A partial parotidectomy was performed and the posterior belly of digastric was divided to facilitate access to the tumour. The tumour was identified as a firm mass in the right PPS medial to the internal and external carotid arteries and hypoglossal and lingual nerves. The hypoglossal and lingual nerves were identified, mobilised and preserved. Bimanual palpation via the oropharynx was used to assist in identifying the tumour location. The internal and external carotid arteries were identified and preserved, with a vascular loop placed on the external carotid for control. Surgicell absorbable haemostatic dressing was placed around the muscular PPS. The tumour was excised from soft tissue attachments and sent for histopathology.

A size 14 French gauge suction drain was placed, which was removed three days postoperatively. Immunohistochemistry (IHC) demonstrated strong positivity for S100 and only patchy/minimal positivity for Smooth Muscle Actin (SMA). Negative staining was seen for CD34 and Desmin. These features were in keeping with a schwannoma, which was completely excised [Table/Fig-2,3].



[Table/Fig-2]: Macroscopic description: a 65×40×28 mm well-defined firm lobulated piece of tissue. The external surface has been inked black. Serial sectioning shows solid white and cystic haemorrhage areas.

[Table/Fig-3]: Macroscopic description: irregular fibrofatty tissue and muscle, measuring 27×25×13 mm. The external surface has been inked black. Serial sectioning shows fibrofatty tissue and muscle with a 15×4×3 mm component comprising salivary tissue and a few 2-3 mm lymph nodes.

Day one post following surgical excision, the patient underwent a flexible nasopharyngolaryngoscopy which confirmed bilaterally mobile and symmetrical vocal cords and normal laryngeal appearances.

During the immediate postoperative period the patient remained on regular analgesia. The patient had a right hypoglossal nerve weakness and right marginal mandibular weakness which was evident on movement of facial muscles but not noticeable at rest. The patient was successfully discharged three days postoperatively. Postoperative outcomes have been positive with the patient being disease-free four years postexcision.

DISCUSSION

Peritonsillar Abscess (PTA) or quinsy is a common presentation to both primary and secondary care with an incidence of 7589 cases in the UK [1]. The PPS tumours are rare however, and account for less than 0.5% of head and neck neoplasms [2,3]. Schwannoma is reported to account for one-third of all PPS tumours making it an even more unusual diagnosis [3]. Common presenting features of both PTA and PPS schwannoma include dysphagia, dysphonia, odynophagia and a unilateral oropharyngeal swelling. Given the overlap in clinical features, combined with the fact that PTA is significantly more common than PPS, it is possible for delays in the diagnosis of PPS tumours to occur. In such cases, clinicians need to be aware of PPS tumours as an important differential diagnosis. This is particularly relevant in cases where there is persistent unilateral oropharyngeal swelling and/or ipsilateral medialisation of the palatine tonsil in patients who are otherwise systemically well, usually with normal or minimally raised inflammatory markers not suggestive of an acute infective process. Cases of PPS tumours mimicking PTA are extremely rare [3].

A case report by Siupsinskiene N et al., reports a young male who presented with a two month history of a left sided oropharyngeal swelling. This was initially managed as a PTA for four weeks with no improvement [3]. The FNE revealed a narrow left nasopharynx due to a large mass and further CT and MRI scans confirmed a left PPS lesion, confirmed to be a schwannoma after histological examination [3]. This case report further reinforces the importance of having a wide range of differentials when faced with an oropharyngeal swelling, to ensure correct diagnosis and ensure further management can be initiated. Another rare case study by Mallik KC et al., highlighted a parapharyngeal schwannoma arising from the lingual nerve [4]. The patient presented differently to this case report, being asymptomatic for two years and eventually presenting with a progressive right sided neck swelling [4].

Mallik KC et al., explained the tumour could not be dissected away from the lingual nerve and therefore the nerve had to be sacrificed however, all nerves were identified and preserved macroscopically in our case report [4]. This further demonstrates the variation in anatomical involvement with parapharyngeal schwannoma and thus, the importance of preoperative imaging. Mra Z et al., reported a case of a 26-year-old male who presented with pyrexia, dysphagia and left PTA to the Emergency Department [5]. Despite needle aspiration, an hour later the swelling had reaccumulated which prompted MRI investigation which revealed a cystic lesion of the left PPS [5]. This was removed and sent for histology which was diagnostic of a lymphangioma [5]. This case further corroborates the findings in our case report of PPS lesions mimicking PTA.

Radiological imaging plays a central role in determining the dimensions and nerve of origin of a PPS schwannoma. This is vital in minimising the risk of postoperative neural deficit [6]. Although various authors have proposed initial evaluation with contrast enhanced Computerised Tomography (CT), MRI remains the gold standard [7]. An important polish study by Czerniewicz-Kamińska A et al., focused on CT in establishing the diagnosis and origin of neurogenic tumours [7]. It summarised that radiological findings can often be ambiguous in neurilemmomas and to make an accurate and definitive diagnosis a combination of clinical, radiological and histological evidence is required [7]. Saito DM et al., presents a retrospective review to determine if preoperative imaging can determine whether a parapharyngeal schwannoma originates from the vagus nerve of cervical sympathetic chain [8]. Over 90% of patients in the study underwent MRI studies and revealed vagal schwannomas splaying the carotid artery and jugular vein apart, while sympathetic chain schwannomas displace these vessels together [8]. This is very important as it allows the surgeon to predict with precision vessel displacement and subsequently, counsel the patient on specific anticipated neurological deficits [8].

Miller F et al., reviewed 51 patients to determine which imaging modality (CT or MRI) is more accurate in delineating schwannomas [9]. The MRI was able to locate the tumour in 95% of patients, while CT was able to localise the tumour in 84% [9]. MRI, was therefore far superior in comparison to CT and therefore allows the surgeon to decide the surgical approach with the least morbidity [9]. Ijichi K and Murakami S reviewed 29 cases of parapharyngeal schwannomas and determined MRI scans are crucial in determining the tumour localisation and distinguishing the tumour origin [10]. Furthermore, it highlighted that if a paraganglioma is suspected, an angiography or CT angiography is more appropriate as the tumour is located around the bifurcation of the carotid artery [10]. Another noteworthy case was reported by Gilardi A et al., describing an extracranial schwannoma of the hypoglossal nerve presenting as a deep neck space abscess [11]. MRI incorrectly identified this mass as a paraganglioma due to a “salt and pepper” pattern [11]. Due to the nature of the mass, a team of Ear, Nose and Throat (ENT), vascular and oral maxillofacial surgeons were consulted in order to plan for the best surgical approach for the patient [11]. However,

immunostaining was diagnostic for peripheral nerve sheath tumour. This case importantly highlights the potential pitfalls of MRI and the vital need for a holistic approach to these tumours.

The differential diagnoses for parapharyngeal masses are extensive, but more than half are salivary gland tumours [12]. The second most common are neurogenic tumours, closely followed by paragangliomas [12]. Other subgroups include lesions such as branchial cleft cysts, lymph nodes and haematological tumours [12]. Most of the lesions (80%) are benign [12]. Although a large proportion is benign, 20% of PPS schwannomas display malignant characteristics [12]. Histopathological examination displays a well-circumscribed, encapsulated lesion with a mixture of growth patterns [13]. Antoni A pattern displays high cellularity in an ordered architectural pattern. Antoni B shows a loose meshwork of less dense cellularity resulting in a disordered architectural pattern [13]. Verocay bodies, as seen in this reported case, consist of palisades of Schwann cell nuclei, first described by Uruguayan neuro-pathologist Jose Verocay [13]. Schwannomas are uniformly reactive for the S100 protein. Although the S100 protein exists in a wide variety of tissues and cell types, negativity for CD34 as in this case, ensures differential diagnoses of sarcoma, meningioma or neurofibroma are less likely [13]. Negativity for desmin may also exclude leiomyosarcoma or soft tissue sarcoma [13].

There are several surgical approaches for excision of PPS masses. A number of factors determine the approach to be undertaken. These include site and size of the tumour, adequate exposure, minimising the risk of neurovascular injury and surgeon's preference [14]. The trans-parotid approach is traditionally utilised for deep lobe parotid masses and it allows early exposure of the facial nerve [14]. Larger salivary gland masses may requires this approach to be combined with the more common transcervical approach [14]. This is often utilised in resecting PPS masses, as it allows adequate exposure of neurovascular structures whilst minimising the risk of oral flora contamination. The transmandibular approach is an alternative and offers the widest access to the PPS. This allows for excision of large and highly vascular PPS masses [15].

Ultimately, the direct transoral approach allows for excision of large, well circumscribed lesions extending into the oropharynx. This is undertaken at increased surgical risk however, as there is limited exposure of neurovascular structures [6]. Schwannomas are largely resistant to neoadjuvant radiotherapy, which is therefore not indicated in management [16]. There is an absence of national guidance on management of PPS schwannomas, which may be due to the fact that it is a rare and benign tumour. As expected, UK National Institute for Health and Care Excellence (NICE) guidance is focused on presentation, investigation and management of head and neck malignancy [17]. Encapsulation and rarity of recurrence ensures that prognosis for schwannoma is excellent and surgical resection is the mainstay of treatment [18]. The management of PPS masses must be a patient-centered, multidisciplinary approach, which factors in relevant co-morbidities, tumour size, suspected diagnosis and patient preference.

CONCLUSION(S)

The Parapharyngeal Space (PPS) schwannomas are exceptionally rare and the majority of patients are normally asymptomatic or present acutely with similar symptoms to a PTA. The MRI scans remain the gold standard in the diagnostic process and should be performed preoperatively in order to plan radical removal of the tumour with its capsule. This should always be done using a multidisciplinary approach to ensure all of the patients' needs are met.

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PLAGIARISM CHECKING METHODS: [Jain H et al.]

- Plagiarism X-checker: Oct 05, 2021
- Manual Googling: Dec 28, 2021
- iThenticate Software: Feb 15, 2022 (8%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:

- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? No
- For any images presented appropriate consent has been obtained from the subjects. No

Date of Submission: **Oct 03, 2021**
Date of Peer Review: **Nov 10, 2021**
Date of Acceptance: **Dec 29, 2021**
Date of Publishing: **Apr 01, 2022**