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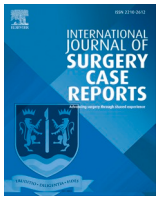

Case report

Pterygopalatine fossa schwannoma–case report and literature review

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ABSTRACT

Introduction: Schwannomas are well-differentiated, benign tumours that originate from the Schwann cells of nerve sheaths. They constitute 25-45% of all the head and neck tumours and can cause significant morbidity depending on the site of origin. The pterygopalatine fossa is the rarest site of involvement, with only a few cases reported in the literature.

Case presentation: This is the case of a 46-year-old male who presented with a twelve [12] month history of left-sided facial pain and progressive swelling. Contrast-enhanced computed tomography (CT) and magnetic resonance imaging (MRI) revealed a large soft tissue lesion located in the left pterygopalatine fossa (PPF). Surgical management is presented, and technical details of the repair are discussed. At one year follow-up, there were no signs of recurrence, and the cosmetic outcome was satisfactory.

Discussion: This case is one of a handful of reported cases of PPF schwannomas in the English literature. Although a significant percentage of schwannomas arise in the head and neck region, the pterygopalatine fossa is the rarest site of involvement. Due to its clinically inaccessible location and complex connections, the pterygopalatine fossa can act as a natural conduit for the spread of inflammatory and neoplastic diseases in the head and neck.

Conclusion: Currently, the endoscopic endonasal approach (EEA) is preferred due to its safety and good oncologic outcome. There is also decreased morbidity as it is minimally invasive. Furthermore, surgeons embarking on the EEA should be equipped with an image guidance system and be trained in advanced endoscopic techniques. However, the open approach remains a reliable and proven surgical method to treat large tumours located within this intricate and inaccessible area.

1. Introduction

Schwannomas are benign tumours originating from the Schwann cells of nerve sheath. These tumours are rare and can be found in any part of the body. However, 25–35% of cases are found in the head and neck region. Fewer than 4% of these tumours involve the nasal cavity, paranasal sinuses or even the pterygopalatine fossa (PPF) [1].

The pterygopalatine fossa (PPF) is a small, clinically inaccessible space located in the deep face. This area serves as a major neurovascular crossroad between the oral cavity, nasal cavity, nasopharynx, orbit, masticator space, and the middle cranial fossa. Due to its complex location and connections, it can potentially act as a natural conduit for the spread of inflammatory and neoplastic diseases in the head and neck [2].

2. Case report

Mr. M.G, a 46-year-old man, presented to an ENT specialist with an eight-month history of pain and swelling over the left cheek. There was no history of associated trauma, and he had not experience any sinonasal or dental symptoms. His past medical history, psychological and drug history was non-contributory to his management. He was a non-smoker and did not consume alcohol.

On examination, there was a noticeable left-sided facial swelling that was firm, smooth and non-tender (see Fig. 1). Examination of the oral cavity revealed an underlying mass at the left canine fossa. Flexible nasoendoscopy was unremarkable. There was no focal neurological deficit.

A contrast-enhanced computed tomography (CT) scan revealed a 6.1 cm × 5.1 cm × 4.6 cm expansile soft tissue mass occupying the left infraorbital region and was located posterior to the left maxillary sinus (see Fig. 2). The exact origin was not positively identified by CT.

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Magnetic resonance imaging (MRI) showed the mass centered in the left PPF. It measured 6.2 cm × 6.1 cm × 5.5 cm (see Fig. 3). This mass was noted to be expanding superiorly, laterally and anteriorly with the remodeling of the maxillary antrum. The mandibular division of the trigeminal nerve was seen and appeared to be intact. The appearances were consistent with that of a neuroma or nerve sheath tumour.

An incisional biopsy was performed, and histopathological examination confirmed the mass to be a schwannoma. On immunohistochemical (IHC) analysis, S-100 staining was positive. The patient was then counselled on the surgical options available. This surgery was performed by a senior Consultant Head and Neck surgeon who is also a lecturer at the University of the West Indies. The preferred approach was an open transmaxillary utilizing a Weber-Ferguson incision (see Fig. 4). This approach was chosen because it facilitated broad exposure of the maxillary sinus thereby minimizing damage to surrounding structures. Due to the numerous feeding vessels from the internal maxillary artery coupled with the inaccessible nature of the PPF, the external carotid artery was identified and secured via a neck incision as a preliminary procedure. This technique would help control intraoperative haemorrhage in the event of damage to the proximal part of the left internal maxillary artery. The use of haemostatic energy devices in the PPF is particularly risky. This is because of its deep, inaccessible nature and proximity to neurovascular structures. Post-operatively he was admitted to a regular observation ward with close monitoring.

Final histology confirmed the mass to be a schwannoma; however, the nerve of origin was not identified. This patient's post-operative recovery was uneventful, and he was discharged two days later. There was no neurological deficit. At one year follow-up, the patient was satisfied and there was a satisfactory functional and cosmetic outcome (see Fig. 5). There were no signs of recurrence. This work has been reported in line with the SCARE 2020 Criteria [3].

3. Discussion

Verocay first described schwannomas in 1910 as slow-growing solitary tumours which originate from the neuroectodermal Schwann cells of cranial, peripheral, and autonomic nerve sheaths. Occurrence in the nasal cavity, the paranasal sinuses, and the adjacent pterygopalatine fossa is rare, accounting for less than 4% of all head and neck schwannomas [4]. The peak incidence is between the third and fourth decades of life but can occur at any age, including young children [5] and they have no gender or race predilection. Because of the relatively indolent and typically benign course, most schwannomas are asymptomatic in their early-stage or tend to cause local symptoms due to compression.

A Pubmed review of the literature from 1980 revealed only 27 cases of benign primary sheath nerve tumours of the PPF (see Table 1) [6–10].

These tumours may present with varying signs and symptoms depending on the nerve of origin and extent of the mass. Symptoms are

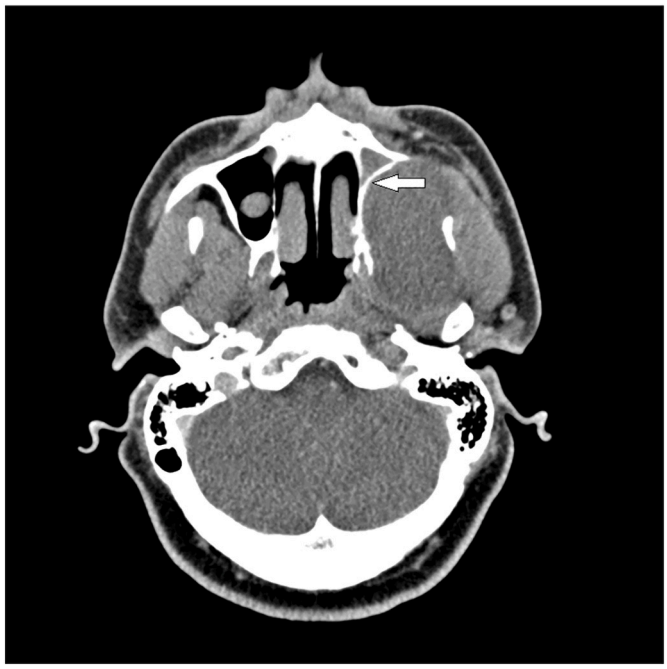

Fig. 2. CT scan showing 'bowing' posterior wall of maxillary sinus (white arrow).

Fig. 1. Clinical presentation showing facial asymmetry.

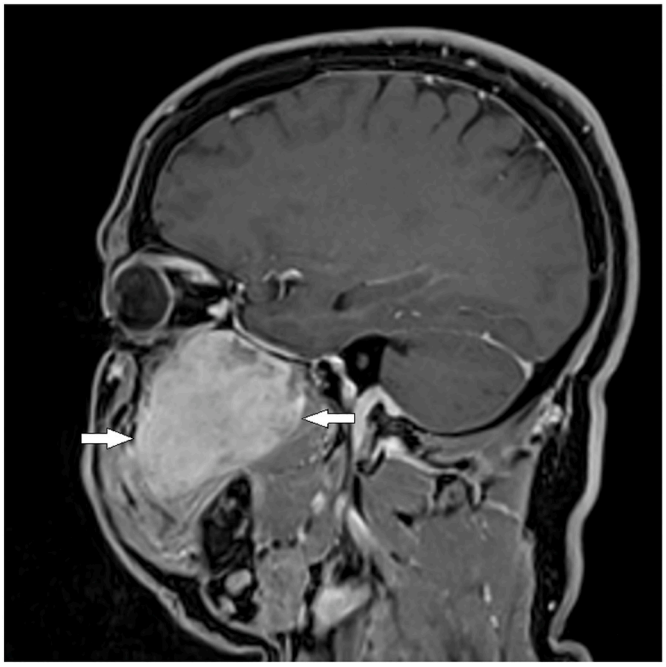

Fig. 3. Sagittal MRI showing antero-posterior extent of the tumour (white arrows).

typically insidious and non-specific symptoms that often delay the diagnosis [6]. Furthermore, despite the availability of CT/MRI, a few patients still present with large tumours in this location, as seen with this index case.

Diagnosis is usually established by a combination of imaging and histological evaluation. Computed tomography (CT) scans help to localize the disease and determine the extent of bony resorption or erosion. Magnetic resonance imaging (MRI) is considered complementary to CT and is helpful to evaluate intracranial involvement. MRI shows low-intensity signals on T1-weighted images and intermediate to high intensity on T2-weighted images [11]. Angiography helps determine the lesion's vascularity and is useful if planning for pre-operative embolization. The diagnosis of a schwannoma is confirmed by biopsy and histopathological examination. Histology typically shows alternating regions of compact spindle cells (Antoni A areas) arranged in short, interlacing fascicles or hypocellular myxoid zones (Antoni B areas). Areas of nuclear palisading with nuclear alignment in rows are known as Verocay bodies. Schwannomas demonstrate S-100 positivity on immunohistochemistry [12].

The definitive treatment for these benign tumours is complete surgical excision. Various approaches have been described, and the method chosen is determined by the location, extent of the lesion and surgical expertise [13]. Broadly speaking, these may be classified as open or minimally invasive endoscopic.

Open approaches include:

- Anterior: Caldwell-Luc, Weber-Fergusson
- Lateral: Infratemporal fossa, Transcervical

Endoscopic approaches include:

- Tranmaxillary
- Transnasal
- Transpalatal

Historically, excision of tumours within this region has been performed through a lateral or anterior surgical corridor. Although initially designed for approaching the infratemporal fossa, lateral approaches also give access to the PPF [14]. However, this approach is highly invasive, employing extensive facial incisions, craniotomies or osteotomies. The anterior route is transmaxillary using either a Weber Fergusson or Caldwell-Luc incision. This approach has a disadvantage of scar formation at the incision site, facial oedema and maxillectomy.

Endoscopic endonasal approaches (EEA) with image guidance have become part of the armamentarium of skull base surgeons globally. This has shown to be safe, effective and results in better cosmetics and quality of life [6]. For tumours centred in the PPF, the EEA provides direct access to the lesion while decreasing manipulation of neural and vascular

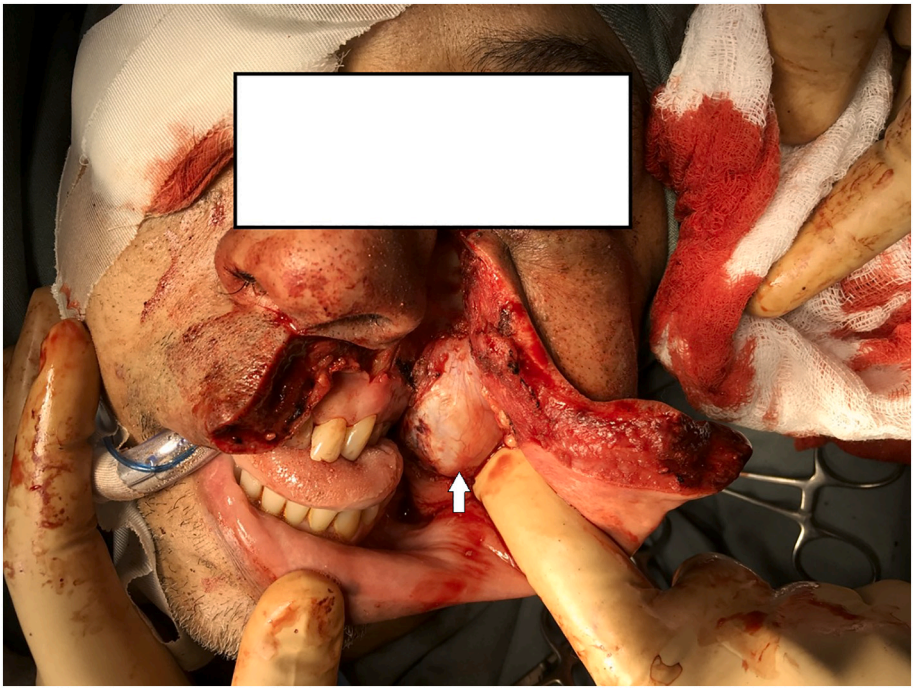

Fig. 4. Gingivo-buccal skin flap elevated to expose the tumour (white arrow).

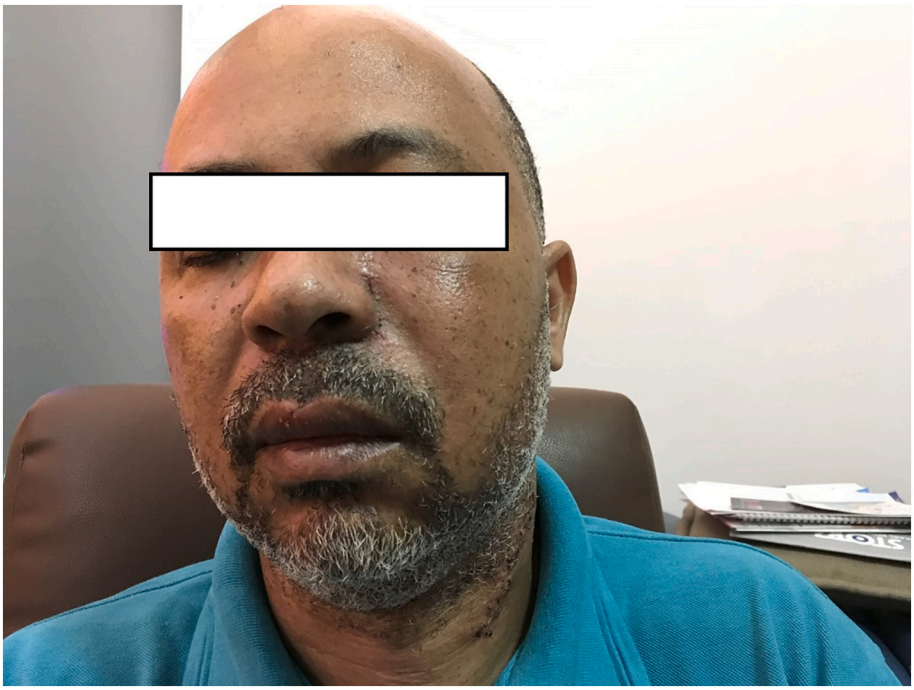

Fig. 5. Post-operative clinical photograph revealing satisfactory cosmetic outcome.

structures, with less overall risk than the other approaches.

The following basic steps are essential for the success of this approach [15,16]:

- Performing an extended medial maxillectomy on the side of the lesion to obtain full access (for visualization and instrumentation) to the posterior wall of the maxillary sinus,
- Control of the vascular structures (internal maxillary artery and its branches); and identifying the second genu of the internal carotid artery by following the vidian canal posteriorly;
- Obtain complete visualization and control of the vidian nerve and V2.

EEA may be associated with post-operative nasal crusting, nasal obstruction, hyposmia and rarely secondary sinusitis or mucocele. These are usually transient symptoms that will improve within 3 to 6 months.

PPF is one of the most anatomically complex regions of the human body, and the lack of image guidance made the endoscopic approach particularly challenging in this case. Sasindra et al. in 2008 [17] reported a case of a giant schwannoma arising from the infraorbital nerve in the PPF. This tumour was treated via a 2-stage approach. After embolizing the main feeding vessels, open surgical excision was performed via a transmaxillary approach, utilizing a Weber-Ferguson incision. Reconstruction of the orbital floor was delayed in this case until the final histopathological report was available and negative margins were confirmed [17].

Plzak et al. [18] reported on a case series of 12 benign tumours of the PPF between 2014 and 2016. Ten of these tumours were juvenile nasopharyngeal angiofibromas, and two were schwannomas. All surgeries were successfully performed via the endoscopic approach with minimal complications. Therefore, the authors of this study suggested that the EEA be employed for the management of masses in the PPF once fundamental prerequisites are met:

- Thorough pre-operative evaluation of CT/MRI
- The availability of adequate technical equipment (endoscopes, endoscopic skull base instruments, a navigation system)
- Adequate experience and skill of the surgeon in the use of advanced endoscopic techniques.

Generally, recurrences of head and neck schwannomas are rare after complete excision and as such, radiation therapy is not indicated for these benign tumours.

4. Conclusion

Masses of the PPF can be effectively treated by either open or endoscopic approaches. Currently, the endoscopic approach is preferred due to its safety and good oncologic outcome. There is also decreased morbidity as it is minimally invasive. Furthermore, surgeons embarking on the EEA should be equipped with an image guidance system and be trained in advanced endoscopic techniques. However, the open approach remains a reliable and proven surgical method to treat large tumours located within this intricate and inaccessible area.

Consent statement

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Ethics approval

Not Applicable.

Guarantor

Wendell Dwarika
Shivanand Maharaj

Table 1
Published cases of PPF schwannomas from 1980 to our present case.

	Year	Author	Age	Sex	Tumour Size (mm)	Approach
1	1980	Gooder et al	55	M	n/a	Open (Weber-Ferguson)
2	1981	Puterman at al	24	M	50	Open (Weber-Ferguson)
3	1983	Bitoh et al	38	F	40	Open (Infratemporal)
4	1987	Suenaga et al	18	F	n/a	Open (Weber-Ferguson)
5	1988	Iwai et al	7	F	50	Open (Infratemporal)
6	1993	Klossek et al	57	n/a	29	Endoscopic
7	1993	Klossek et al	n/a	n/a	n/a	Endoscopic
8	1994	Chen et al	13	F	n/a	Open (Infratemporal)
9	2002	Pasquini et al	50	M	40	Endoscopic
10	2003	Del Gaudio et al	33	F	<25	Endoscopic
11	2005	Boedeker et al	25	M	n/a	Open (Caldwel-Luc)
12	2005	Robinson et al	23	n/a	n/a	Endoscopic
13	2005	Martinez Ferreras et al	40	n/a	n/a	Endoscopic
14	2007	Keh et al	54	F	20	Endoscopic
15	2008	Sasindran et al	22	M	>50	Open (Weber-Ferguson)
16	2009	G Rose et al	33	F	50	Endoscopic
17	2010	Rosique-López et al	52	F	50	Endoscopic
18	2011	Xu et al	57	F	n/a	Endoscopic
19	2011	Xu et al	37	M	n/a	Endoscopic
20	2011	Xu et al	40	F	n/a	Endoscopic
21	2011	Suh et al	35	F	14	Endoscopic
22	2011	Suh et al	45	F	27	Endoscopic
23	2014	Bresson et al	41	F	30	Endoscopic
24	2015	Zhou et al	23	F	n/a	Open (Caldwel-Luc)
25	2015	Newaskar et al	10	M	54	Open (Infratemporal)
26	2015	Siew Min Keh et al	54	F	20	n/a
27	Present Case	Dwarika et al	46	M	61	Open (Weber-Ferguson)

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CRediT authorship contribution statement

Maharaj S: Writing - Original Draft Preparation, Writing - Review & Editing Preparation

Dwarika W: Conceptualization, Supervision, Review & Editing Preparation

Declaration of competing interest

The authors have no financial and/or personal relationships to declare as conflicts of interest that could inappropriately influence their work. This work was completed without any external source of funding and/or study sponsors.

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