An unusual finding of schwannoma in the columellar area—A case report

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ABSTRACT

INTRODUCTION: Schwannomas are benign tumours of the nerve sheath that originate from Schwann cells. Less than 4% of these tumours arise in the sinonasal tract. Columellar involvement is extremely rare – three other cases involving the columella have been reported since 1967.

PRESENTATION OF CASE: A 25-year-old woman presented with a swelling of the nasal columella from eight months into pregnancy. She presented with right nasal obstruction and discomfort over the nasal bridge. Pre-operative MRI and ultrasound were performed. The mass was surgically excised using an external septorhinoplasty approach giving a good cosmetic outcome. Histopathologic examination demonstrated schwannoma.

DISCUSSION OF CASE: Nasal schwannoma may present with variable symptoms. We discuss the MRI and histological features of schwannoma. A literature review suggests that schwannomas may have accelerated growth in pregnancy. The open rhinoplasty approach is the favoured method for excision of schwannomas near the columellar region.

CONCLUSION: Nasal schwannomas are rare in the sinonasal tract; however they need to be considered part of the differential diagnosis for nasal masses. The treatment of choice for these lesions is surgical excision.

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1. Introduction

Schwannomas, also called neurilemmomas, are benign tumours of the nerve sheath. They are relatively common tumours with approximately 25–45% arising from the head and neck region [1]. Sinonasal involvement is uncommon with only 4% of these tumours involving the nasal and paranasal cavity [2]. Columellar involvement is extremely rare. A literature search found only three other published cases of schwannoma involving the columella [3,4]. We therefore present the 4th documented case of a columellar schwannoma. This case report has been reported in line with the SCARE criteria [5].

2. Case presentation

A 25-year-old Middle Eastern woman presented to our ENT department in a tertiary teaching hospital with a two-month history of a swelling of the nasal columella. She first noticed this eight months into pregnancy, and she attended our clinic about one month after uneventful delivery of twins. The lesion gradually increased in size during her pregnancy, and was associated with

Fig. 1. Columellar mass immediately before operation.
right nasal obstruction and discomfort over the nasal bridge. Her sense of smell and taste remained intact. There was no history of epistaxis, rhinorrhea or pain. She was otherwise fit and well.

On examination there was a soft, smooth, non-tender expansive lesion over the columnella, with mild telangiectasia of the overlying skin (Fig. 1). It was felt to be separate from the anterior nasal septum. Flexible nasoendoscopy revealed no abnormality within the nasal cavity. A non-contrast MRI revealed a 1.9 × 1.4 × 1.1 cm homogeneous mass in the right nasal columella, of high T2 and intermediate T1 signal, abutting the nasal septum with no deeper extension (Fig. 2).

The differential diagnosis at the time she was seen included a nasal dermoid cyst. The decision to surgically excise this was made based on the recent enlargement and cosmetic impact. Pre-operatively, there was concern regarding vascularity of this lesion therefore an ultrasound scan was performed. It revealed a solid lesion with significant vascularity and multiple feeding vessels.

She underwent her operation after a 13-month clinical investigation. The size of the lesion had remained stable since the end of her pregnancy. An open rhinoplasty approach was used to excise the mass, with use of a columnellar chevron incision followed by raising superior and inferior skin flaps and dissection of the lower lateral cartilages. The mass was found to be well-encapsulated. It was bluntly dissected from the skin and cartilage. The upper lateral cartilages were not encountered and the lesion was completely excised with the capsule intact (Fig. 3). The medial crura of the lower lateral cartilages were apposed with an absorbable suture before skin closure. Nasal splints were not used.

Post-operative histological examination of the specimen revealed an encapsulated spindle cell neoplasm measuring 23 × 18 × 12 mm (larger than initially suggested on MRI). There were foci of peripheral palisading of the lesional cells, with formation of Verocay bodies. Admixed with the spindle cells were frequent small to medium diameter blood vessels with hyalinised walls. There was little cytological atypia, no atypical forms and no necrosis found. Immunohistochemistry was performed, showing strong and diffuse nuclear and cytoplasmic staining for S100 and strong nuclear staining for red Sox–10. Although only very occasional mitotic figures were found, the Ki67 (MIB-1) stain showed a higher proliferation fraction than is typically found in schwannoma (5–8% as opposed to >1% in most examples). CD34, CD31 and ERG highlighted the prominent vascular background. Staining for oestrogen receptor (ER) and progesterone receptor (PGR) were both negative (at the threshold used to evaluate ER and PGR).

Fig. 2. Appearance of columnellar lesion on MRI T2 sequencing (axial and sagittal views).

Fig. 3. Intra-operative view: dissection of lesion with intact capsule.
Fig. 4. Immediate post-operative appearance.

Fig. 5. a. Haematoxylin and eosin stained section showing Antoni A (top) and Antoni B (bottom) areas of the Schwannoma. Verocay bodies composed of palisaded spindle cells are present in the Antoni A areas. b. Immunohistochemistry for S100 showing strong and diffuse staining of the tumour cells. c. Immunohistochemistry for Ki67 (MIB-1) demonstrated a proliferation fraction of 5-8% overall.

over-expression in breast cancer). The overall picture was that of a benign peripheral nerve sheath tumour, most in keeping with a cellular schwannoma.

Recovery following the operation was uneventful with satisfactory cosmetic outcome (Figs. 4 and 5). Follow-up at six months showed no signs of recurrence.

3. Discussion

Schwannomas were first described by Verocay in 1908 [6,7]. Symptoms of sinonasal schwannoma vary with the location of the tumour, whether in the external nose or presenting in the nasal cavity. Most common symptoms are nasal obstruction, but they may also present with epistaxis, mucopurulent rhinorrhea, anosmia, facial swelling, ptosis, headache, or pain [2]. Apart from columellar involvement, schwannomas have also been reported to occur in the nasal tip or the anterior nasal septum, where the treatment approaches may be similar [8–11].

A definitive diagnosis is made based on the histologic findings. In our case, MRI features of homogeneity and high T2 signal are consistent with what is reported in the literature [2,9,10]. Imaging can be useful to delineate the extent of the lesion if a broad differential is being considered (e.g. dermoid cyst, lipomas, leiomyoma, leiomyosarcoma); however these features of schwannoma on MRI are not diagnostic [10]. Neurofibromas should be considered in the differential diagnosis of schwannoma. Both of these tumors originate from Schwann cells. Schwannomas are occasionally associated with neurofibromatosis type 2, whereas cutaneous neurofibromas are classically associated with neurofibromatosis type 1.

Microscopically, schwannomas are usually encapsulated and biphasic (highly cellular Antoni A, hypocellular Antoni B areas);
they also follow a palisading pattern (Verocay bodies – 2 rows of elongated palisading nuclei alternating with acellular zones) [6]. Two case series with a total of 11 patients show that sinonasal schwannomas have an unusual feature of lack of encapsulation and possible ulceration of the epithelial covering, yet without local recurrence or metastasis on long-term follow-up [12,13]. This was not the case in our patient, and one other case of columellar schwannoma was also reported to be well-encapsulated [3].

There are several subtypes of schwannoma: cellular, pigmented, plexiform and epithelioid. In our case the patient had a cellular schwannoma. Schwannomas in general have a low recurrence rate of only 2%, and that is usually associated with neurofibromatosis type 2 [9].

Given that our patient presented with an increase in size of the lesion during pregnancy (final histological size > initial size on imaging), we searched the literature and found reports of accelerated growth of schwannomas in pregnancy [14,15], with one case of an orbital schwannoma shown histologically to have the presence of progesterone receptors [12]. The effects of pregnancy on vestibular schwannomas (acoustic neuromas) have been studied – acceleration of tumour signs and symptoms in late pregnancy were attributed to either direct hormonal stimulation of growth, or to hormonally-induced vascular dilatation [16]; however the exact role these hormones play is unclear as studies of frequency of oestrogen and progesterone receptor expression have conflicting results [17]. In our case, although there was no over-expression of oestrogen or progesterone receptors histologically, there remains the possibility that there could be receptors below the threshold for detection.

The nerve of origin for schwannomas of the columella is difficult to determine, but it is likely to be a branch of the ophthalamic or maxillary divisions of the trigeminal nerve, or from autonomic nerves supplying the vasculature [8].

The treatment of choice for schwannoma is resection. Different approaches are used depending on the exact subsite of the tumour. As it is a benign, usually well-encapsulated lesion, disfiguring excisions are unnecessary. For lesions in the columella (or subsites adjacent to the columella e.g. nasal tip), excision with the open rhinoplasty approach (collumellar incision) seems to be the ideal surgical method as reported in various cases [3,4,7,8,17] – it offers good exposure to the nasal cartilage framework, making it easier to perform modification of nasal tip deformities and asymmetry [18].

4. Conclusion

To summarise, nasal schwannomas are rare but may be considered in the differential diagnosis of soft tissue masses of the external nose. MRI can be useful to evaluate the lesion and its effect on surrounding structures. Pregnancy may contribute to enlargement of schwannomas with resulting increase in symptoms. These tumours are benign and treatment with excision is usually complete in most cases. We recommend the external rhinoplasty approach to surgical excision.

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Ethical approval

We don’t require ethical approval. Has been exempted by our hospital.

Consent

Consent obtained.

Author contribution

Sarah Bouayyad: Collected data, literature review, wrote article. Jen Ong: Reviewed and edited article. Matthew Ellis: Advice on article content, support collecting patient data.

Guarantor

Sarah Bouayyad.

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