Angioleiomyoma of the inferior turbinate: a rare cause of isolated facial pain

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ABSTRACT

We present the case of a 33-year-old man with right-sided facial pain. Clinical examination revealed an isolated mass attached to the right inferior turbinate. This was confirmed with computed tomography. Excision was achieved endoscopically and histology revealed an angioleiomyoma. Full symptomatic relief was achieved after surgical excision. Less than 1% of angioleiomyoma lesions are found within the sinonasal cavity. We describe the first documented presentation of angioleiomyoma as a cause of isolated, unilateral facial pain; a very common presentation to the otorhinolaryngology clinic. We promote consideration of angioleiomyoma as a different diagnosis in the presence of facial pain and a unilateral sinonasal lesion. Endoscopic resection provides complete symptomatic resolution.

KEYWORDS

Angioleiomyoma – Benign nasal tumour – Facial pain – Inferior turbinate

Accepted 5 May 2019

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Case history

A 33-year-old man presented to the ear, nose and throat outpatient clinic with isolated right-sided facial pain. On endoscopic examination, an exophytic polypoidal lesion was discovered in the right nasal cavity arising from the medial border of the inferior turbinate. Computed tomography (CT) of the nose and paranasal sinuses and an urgent functional endoscopic sinus surgery with biopsy of the polyp were organised. The CT revealed a solitary 15-mm polyp arising within the right middle meatus (figure 1a,b) with no aggressive radiological features. Deep (Keros type III) and asymmetrical descent of the cribriform plates was also noted. Complete macroscopic excision of the lesion was achieved via an endoscopic approach under general anaesthesia.

Histological analysis showed a well-circumscribed lesion composed of fascicles of spindled cells with cigar-shaped nuclei and abundant cytoplasm with a perivascular growth pattern compressing vascular lumina. The lesional cells stained positively for actin (figure 2a) and calponin (figure 2b) and were focally positive for desmin (figure 2c), consistent with a diagnosis of angioleiomyoma. The Ki67 proliferation index was low (figure 2d). Resection margin analysis confirmed complete excision.

At the six-month follow-up, endoscopic examination revealed no signs of recurrence or residual tumour. The patient described complete relief of symptoms.

Discussion

Angioleiomyoma is also termed angiomyoma or vascular leiomyoma. It is a mesenchymal benign tumour of skin and subcutaneous tissue comprising well-differentiated proliferation of smooth muscle with a varying vascular constituent.¹ These rare tumours are predominantly found in women between the ages of 30–50 years of age, most commonly within the uterine wall and lower extremities. Characteristic presentation involves pain exacerbated by cold temperatures. This is thought to be due to nerve involvement or contraction of the tumour's smooth muscle causing ischaemia.

The aetiopathogenesis of angioleiomyoma is poorly understood. Epstein-Barr virus has been found to have a role in the development of smooth-muscle tumours in immunocompromised patients.² This link has not yet been established outside of this patient cohort, however.

Angioleiomyoma within the nasal cavity is a rare occurrence, representing less than 1% of all angioleiomyomas.³ The most common presenting symptoms include epistaxis and nasal obstruction.⁴ Pain is an uncommon presenting symptom of angioleiomyoma in the head and neck. A 2015 review of the literature regarding sinonasal angioleiomyoma revealed only 5 of 54 cases (9%) that reported pain in any form as a presenting symptom. Isolated facial pain is an extremely rare presentation. To our knowledge, this is the only reported case of sinonasal angioleiomyoma presenting exclusively with unilateral facial pain.⁵

nasal cavity. Although radiological features are unable to provide specific diagnostic information in cases of angioleiomyoma, their role is primarily to investigate for signs of an invasive process, which may point towards malignancy. Additionally, the images help the operating surgeon to delineate the patient's anatomy prior to resection of the tumour and allows for surgical planning.

As neither clinical nor radiological examination may offer a definitive diagnosis, accurate diagnosis of angioleiomyoma is therefore made using histopathological techniques. The lesion was well circumscribed, had a low proliferation index and showed no evidence of necrosis or cytological atypia, confirming a benign neoplasm. In our case, the microscopic appearance of spindle cells with 'cigar-shaped' nuclei merging with thick-walled blood vessels was characteristic for angioleiomyoma. The diagnosis

Figure 2 Microscope pictures of lesional staining techniques. (a) Positive staining for actin. (b) Positive staining for calponin. (c) Focally positive staining for desmin. (d) Low Ki67 proliferation index.

The clinical diagnosis of angioleiomyoma is extremely difficult, as this pathology does not present with characteristic signs or symptoms that are distinct from other benign sinonasal diagnoses. Therefore differential diagnosis in the clinical setting should be broad.

Our case advocates CT as the primary radiological investigation for unilateral polypoidal disease within the

was confirmed with strongly positive stains for actin and calponin, and focal positive staining for desmin. S-100 staining was negative, excluding neurogenic aetiology.

Although rare, there is potential for malignant transformation from angioleiomyoma to leiomyosarcoma. It is thus essential to achieve complete excision of these lesions. We advocate a wide excision technique under endoscopic

guidance to achieve this excision. This technique is consistent with the methods of other sinonasal angioleiomyoma reports within the literature.^{2,4}

Conclusion

Sinonasal angioleiomyoma is a rare diagnosis with malignant transformation potential. This unique presentation of isolated facial pain highlights the importance of the consideration of angioleiomyoma as a differential diagnosis in patients with this presentation. We advocate radiological investigation with CT and resection using an endoscopic approach to achieve total excision.

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