

## Pediatric Infraorbital Nerve Schwannoma: A Rare Clinical Entity

<sup>1</sup>Raviraj GA, <sup>2</sup>Thomas Rony, <sup>3</sup>Dhanraj GA, <sup>4</sup>Vishal US Rao

<sup>1</sup>Reader, Department of Pedodontics, KGF Dental College, KGF, Karnataka, India

<sup>2</sup>Assistant Professor, Department of ENT, MOSC Medical College, Kolenchery, Kerala, India

<sup>3</sup>Professor, Department of ENT, Annayya ENT Centre, Bengaluru, Karnataka, India

<sup>4</sup>Assistant Professor, Department of Head and Neck Surgery, RR Medical College, Bengaluru, Karnataka, India

**Correspondence:** Raviraj GA, Reader, Department of Pedodontics, #14, 14th Main, HSR Layout, Sector 5, Bengaluru-560105 Karnataka, India, e-mail: raviraj\_mds@yahoo.co.in

### ABSTRACT

This report elaborates a rare case of infraorbital nerve Schwannoma in an 8-year-old boy who presented with the recurrent progressively increasing swelling over the right cheek following initial treatment. The mass was excised surgically via a facial approach. Infraorbital nerve sheath tumors occurring in the pediatric age are rare and can be often mistaken for infectious causes in this age group. Surgery remains the mainstay of treatment in these tumors. Early diagnosis and prompt surgical treatment can serve to minimize the treatment related morbidity arising from multiple interventions while avoiding undue apprehension. Pediatric infraorbital schwannomas, although a rare entity, should be considered in the differential diagnosis of long standing pediatric facial swellings.

**Keywords:** Schwannoma, Infraorbital nerve, Surgery.

### INTRODUCTION

Schwannomas are slow growing benign peripheral nerve sheath tumors arising from the Schwann cells. Although common in the head and neck region, their appearance in the infraorbital area is extremely rare. Schwannomas in this region are most often seen to originate from the nerve sheath surrounding the infraorbital nerve. We elaborate this case of the recurrent infraorbital nerve schwannoma, which was excised using a upper cheek flap.

### CASE REPORT

A 8-year-old patient was referred to the otolaryngology clinic with a history of swelling on the left cheek below the orbit since 4 years. The mass was initially excised at a local hospital using a limited facial incision 4 years back (Fig. 1). One year following the surgery, patient noticed a swelling in this region which was gradually increasing in size. The patient did not have any symptoms except swelling over the left face. Upon clinical examination, sensation over the left cheek was present and ophthalmologic examination did not reveal any abnormality. The swelling was in close proximity to the skin at the infraorbital region but not involving it. FNAC of the swelling revealed a benign tumor of neural origin, possibly schwannoma. CT scan performed revealed a diffuse, minimally enhancing mass in the infraorbital region extending downwards and laterally towards the labial region and reaching up to the buccal mucosa adjacent to the canine (Fig. 2). Fibrosis was noted in the labial region, surrounding the tumors, possibly due to previous surgery.



Fig. 1: Preoperative swelling

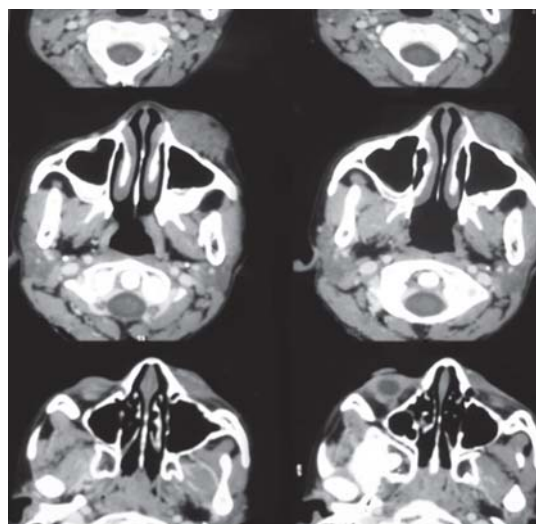


Fig. 2: Preoperative CT scan

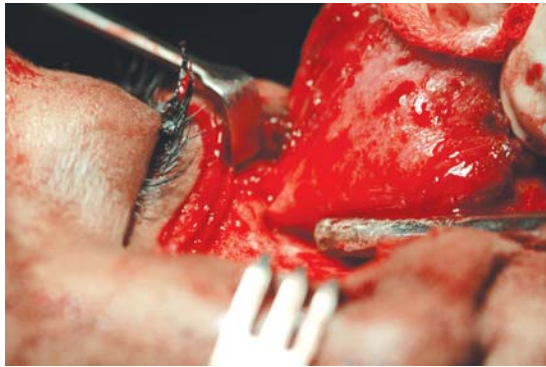


Fig. 3: Tumor arising from infraorbital nerve



Fig. 4: Excised tumor mass

The mass was approached using a Weber-Ferguson incision with a Deffenbach extension, and the upper cheek flap raised. The decision to take this incision was taken primarily due to the diffuse nature of tumor extending laterally, close proximity to the skin at the infraorbital region and surrounding fibrosis from the previous surgery. The mass was found to arise from the infraorbital nerve (Fig. 3). The entire mass was excised with the surrounding fibrosis from previous surgery. A 0.5 cm of the normal stump of the nerve was also excised to ensure liberal margin (Fig. 4).

Postoperatively, there were no complications except paresthesia of the cheek. The patient is disease-free after 6 months postoperatively.

## DISCUSSION

Schwannomas are usually asymptomatic when small and may produce progressive, painless proptosis on enlargement. A variable combination of signs and symptoms may be present due to the variable origin and location of the tumor in the orbit. In the orbit, schwannomas are usually unilateral and may arise from the supraorbital, infraorbital, supratrochlear, ciliary, oculomotor, trochlear or abducent nerves.<sup>1</sup>

Most schwannomas are progressively growing tumors that eventually require treatment. Surgical excision remains

the treatment of choice and the tumor can be excised without compromising the nerve function. Incomplete excision is a common cause of recurrence in these patients. In the present case, the patient underwent previous incomplete excision which led to recurrence. As a result of the previous surgery and the fibrosis therein, we were unable to save the nerve of origin. Therefore, an early complete treatment is indicated to avoid the complications related to progressive growth of the tumor. Schwannomas are slow-growing tumors and do not invade the neighboring bony structures. They may push and erode bony walls by pressure effect. In our case, the orbital floor was intact, and the tumour was found at the point of exit outside the infraorbital foramen.

Schwannomas constitute 1 to 8% of all head and neck tumors and 1 to 4% of the orbital tumors.<sup>2</sup> They most commonly appear between the second and the fourth decades.<sup>2</sup> Trigeminal schwannomas account for almost 0.2% of all intracranial tumors and for approximately 2 to 3% of all intracranial schwannomas with no gender predilection.<sup>3</sup> Although trigeminal schwannomas occur most often in people in their middle decades of life, with the highest incidence occurring between the ages of 38 and 40 years, approximately 10% of schwannomas are diagnosed in patients under the age of 21 years.<sup>3</sup> The oculomotor, ciliary, lacrimal and zygomaticotemporal nerves have been reported as the nerves that the orbital schwannomas most commonly arise. On the other hand, the nerve of origin cannot be identified during surgery in 50% of the cases.<sup>2,4</sup>

Extensive literature search revealed only a few case reports of an orbital schwannoma arising from the infraorbital nerve.<sup>5-11</sup> Since, most of the tumors originate from the sensory nerves, they do not interfere with the eye movements or vision unless they are located in the orbital apex or compress the optic nerve.<sup>12,13</sup>

In conclusion, infraorbital schwannomas are very rare tumors and must be included in the differential diagnosis of the orbital masses inferior to the eyeball.

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